

Reoperative Pediatric Surgery

Edited by

Steven Teich, MD

Donna A. Caniano, MD



 Humana Press

REOPERATIVE PEDIATRIC SURGERY

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Edited by

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Cover illustration: The picture on the front cover depicts the two authors performing surgery together. We wish to thank Dr. Jon Groner for his photographic expertise.

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We are inspired by our patients and their families, who place their trust in our surgical expertise and wisdom. During our training in pediatric surgery we were fortunate to have witnessed commitment to long term patient care by our esteemed teachers, Drs. H. William Clatworthy, Jr., E. Thomas Boles, Jr., and Marc I. Rowe.

We dedicate this book to our respective parents, Pauline and Abraham Teich and Mary and James Caniano, who taught us that we could accomplish anything through hard work and perseverance.

We also dedicate this book to our respective spouses, Esther Chipps and Richard Flores, who are our closest friends, wisest advisors, and sources of daily strength.

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PREFACE

“Good judgment comes from experience, and often experience comes from poor judgment.”

Rita Mae Brown

Reoperative surgery is a challenge that is confronted by every surgeon. Although a particular operation may be initially performed with technical skill and followed by appropriate postoperative care, functional and/or anatomic problems may require further surgical attention.

The unique circumstances of pediatric patients may predispose them to a greater likelihood of requiring reoperation after a major procedure.

- A bowel resection in a neonate may develop a stricture if the anastomosis does not grow at the same rate as the adjacent bowel. The reoperative anastomotic technique is critical, as is the decision whether to resect or taper dilated bowel.
- The cancer survival rate has increased dramatically for many pediatric tumors. These patients often require reoperation for treatment of recurrences, as well as for treatment of complications of chemotherapy, such as second malignancies.
- Pediatric surgical patients often require lifelong follow-up that is obviously much longer than for adults. This increases the chances of requiring reoperation for many conditions, including gastroesophageal reflux disease and inguinal hernia.
- Even a “simple” gastrostomy may develop complications related to growth. With linear growth, the skin of the abdominal wall often migrates towards the chest wall. Therefore, the gastrostomy becomes angulated with leakage of gastric contents onto the abdominal wall, necessitating repositioning of the gastrostomy away from the costal margin.
- Pediatric patients with congenital diseases, such as cystic fibrosis, often require multiple reoperations for complications related to their underlying condition.

It is important to mention that not every pediatric surgery reoperative problem has a wealth of contemporary literature. Often reoperative surgery requires seldom used and more complex operative techniques. Frequently, these techniques are too new or too specialized to be found in current pediatric surgery textbooks. For this reason, we have enlisted a group of authors who are recognized experts for their respective topics to provide the most up-to-date information on reoperations for their pediatric surgical colleagues.

The pediatric surgery literature on reoperations is fragmented and sketchy. The need for a pediatric surgery textbook that critically analyzes and consolidates all the available literature on reoperations is obvious. For this reason, we have compiled a detailed source of information on reoperations for all areas of the body, all parts of the gastrointestinal tract, all types of pediatric solid tumors, and many common but perplexing problems that we co-manage with other pediatric specialists.

This book has been a labor of love. Now, we hope that it will become a valuable reference for pediatric surgeons, pediatric anesthesiologists, general surgeons performing pediatric surgery, and all pediatric physicians.

We wish to thank our secretaries, Cathy Rings and Teresa Rodich, for their invaluable assistance in the preparation of this book.

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1

Radiology of the Postoperative Patient

Diagnosis and Intervention

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ABSCESS OF THE CHEST AND ABDOMEN
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INTRODUCTION

The integrated and effective use of radiological diagnostic modalities and interventional techniques and therapies provides the surgeon with the opportunity to make accurate diagnoses of reoperative issues and complications and to provide timely intervention. A close functional relationship between surgeons and radiologists allows the surgeon the full advantage of surgical therapies or radiological interventional techniques and therapies, as best suits the individual patient needs. Interventional radiology offers a multimodality image-guided and minimally invasive management approach to a multitude of reoperative issues and complications. Consultation between

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the primary surgeon and the radiology team provides discussion of techniques, interventional therapeutic options, expected outcomes, and contingency plans. The surgeon must have a clear understanding of the contrast media used, anatomic approaches of interventional procedures, and associated potential complications, should operative intervention be required following radiological diagnosis and/or intervention.

ABSCESS OF THE CHEST AND ABDOMEN

Abdominal abscesses are the most common indication for image-guided drainage, and appendicitis is the most common etiology (1–5). Appendicitis is more common in children than adults, and children are more likely to have perforated appendicitis and abscesses (1–3,5–7). Percutaneous drainage combined with antibiotics may allow for delayed less-invasive surgery (laparoscopic or small right lower quadrant [RLQ] incision) in children who present with rupture and abscess (7–14). Alternatively, the drainage of postoperative abscesses can eliminate a second surgery (2–7). Other causes of intraabdominal abscesses are less common, but include infected cerebrospinal fluid (CSF) and pancreatic pseudocysts, necrotizing enterocolitis (NEC), Crohn’s disease, and postoperative abscesses of any cause.

The imaging techniques are dependent on the suspected site of abscess. Although CT scanning is most commonly used for abdominal sepsis (Fig. 1), magnetic resonance imaging (MRI) or ultrasound may be more useful in the musculoskeletal system or with superficial lesions (Fig. 2).

After obtaining appropriate history, physical examination, laboratory tests, and abdominal plain radiographs, abdominal computed tomography (CT) scan has become the gold standard for the diagnosis of abdominal abscess (1,3,4). If the patient’s

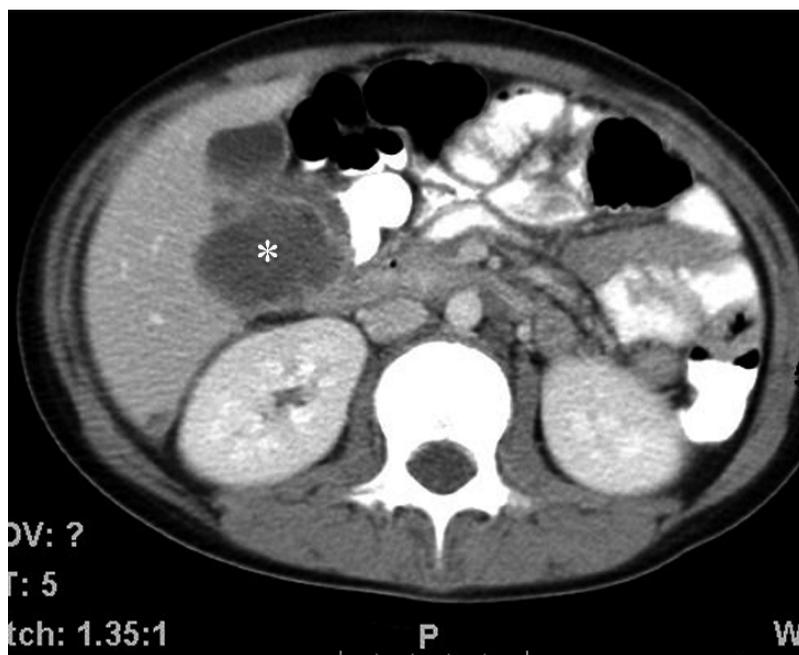


Fig. 1. CT scan of a febrile patient after surgery for ruptured appendicitis shows an abscess (*) near the gallbladder fossa.

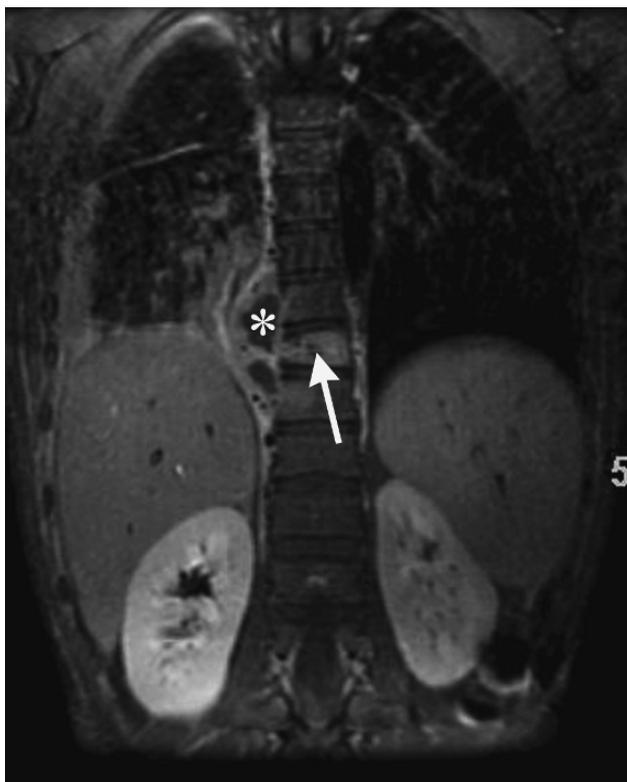


Fig. 2. Coronal image from an MRI shows osteomyelitis of a thoracic vertebral body (arrow), and an adjacent paraspinal abscess (*), drained with ultrasound guidance.

symptoms last longer than 48 hours or if the surgeon has a high clinical suspicion of abdominal abscess, a CT scan with both oral and IV contrast should be performed. The oral contrast improves the recognition of abscesses, as children have less intraabdominal fat than adults and oral contrast differentiates intestinal loops from abscess (Fig. 3). Although CT scanning for abdominal pain has proliferated rapidly, this increased utilization coincides with new information regarding radiation risks in children. Standardized CT dosing techniques are available to decrease exposure as much as possible. Ultrasound can sometimes substitute for CT. This is particularly true in infants with NEC.

Musculoskeletal abscesses are usually diagnosed either with MRI or ultrasound (Fig. 2). MRI is the best imaging tool to evaluate for abscesses related to osteomyelitis. Ultrasound can identify fluid collections within areas of cellulitis.

Neck infections are typically first imaged with CT (Fig. 4A,B). However, after an area of necrosis is identified, ultrasound is more reliable in determining if the lesion is fluid-filled and drainable (15). Chest abscesses are best imaged with CT, and will be discussed in the section on pulmonary infections (16,17).

Ultrasound guidance is ideal in children (1). Children are usually smaller, which may allow sonographic visualization of abscesses. Ultrasound is portable, multiplanar, and provides real-time imaging without the deleterious effects of ionizing radiation. Freehand techniques allow the maximum flexibility during the procedure and increase



Fig. 3. Patient is postappendectomy for perforated appendix. Oral contrast on the CT allows for differentiation between the bowel loops and the abscess (*).

the access site choices. High-quality ultrasound equipment is required with multiple transducers ranging from high-frequency probes for excellent near-field visualization to larger lower frequency probes for deeper abscesses. Endocavitary probes are also needed for selective drainage procedures. CT and CT fluoroscopy are often the preferred guidance modalities in adults. These techniques are also useful in children when ultrasound cannot visualize the abscess because of overlying gas or bone; however, the radiation exposure must be minimized (18).

Most abdominal abscesses are accessible from a transabdominal approach with ultrasonographic guidance (Fig. 5A,B), although deep pelvic abscesses may require transrectal or transgluteal techniques (8,9,12–14,19,20). Transrectal drainage is guided with ultrasound, using either a transabdominal transducer and imaging through the bladder, or an endocavitary probe in the rectum (Figs. 6 and 7). Transgluteal drainage is usually performed with CT guidance (14,20), although transgluteal ultrasound guidance has been described (13). Transgluteal drainage has a reported higher complication rate because of vascular injury and is considered more painful; however, even with the smaller sciatic notch in children, some have excellent success with this technique (14, 20). Abscesses in other sites are accessed through the most direct approach, avoiding vascular and other important structures.

Needles, wires, and catheters are similar to those used in adults. Because of sedation concerns and the lack of cooperation in most children, we choose equipment that provides the greatest procedural efficiency. Ultrasound guidance allows the use of larger needles in smaller patients as the advancement is performed in real time, and the best access window is chosen. This decreases the number of steps and simplifies the procedure. If the access window is small, or if CT guidance is necessary, smaller access sets are available. Standard drainage catheters are available from multiple manufacturers, with most patients receiving 8–12 Fr drains, although that is dependent on the thickness of the fluid. Smaller catheters (5 or 6 Fr) are often helpful in neck or

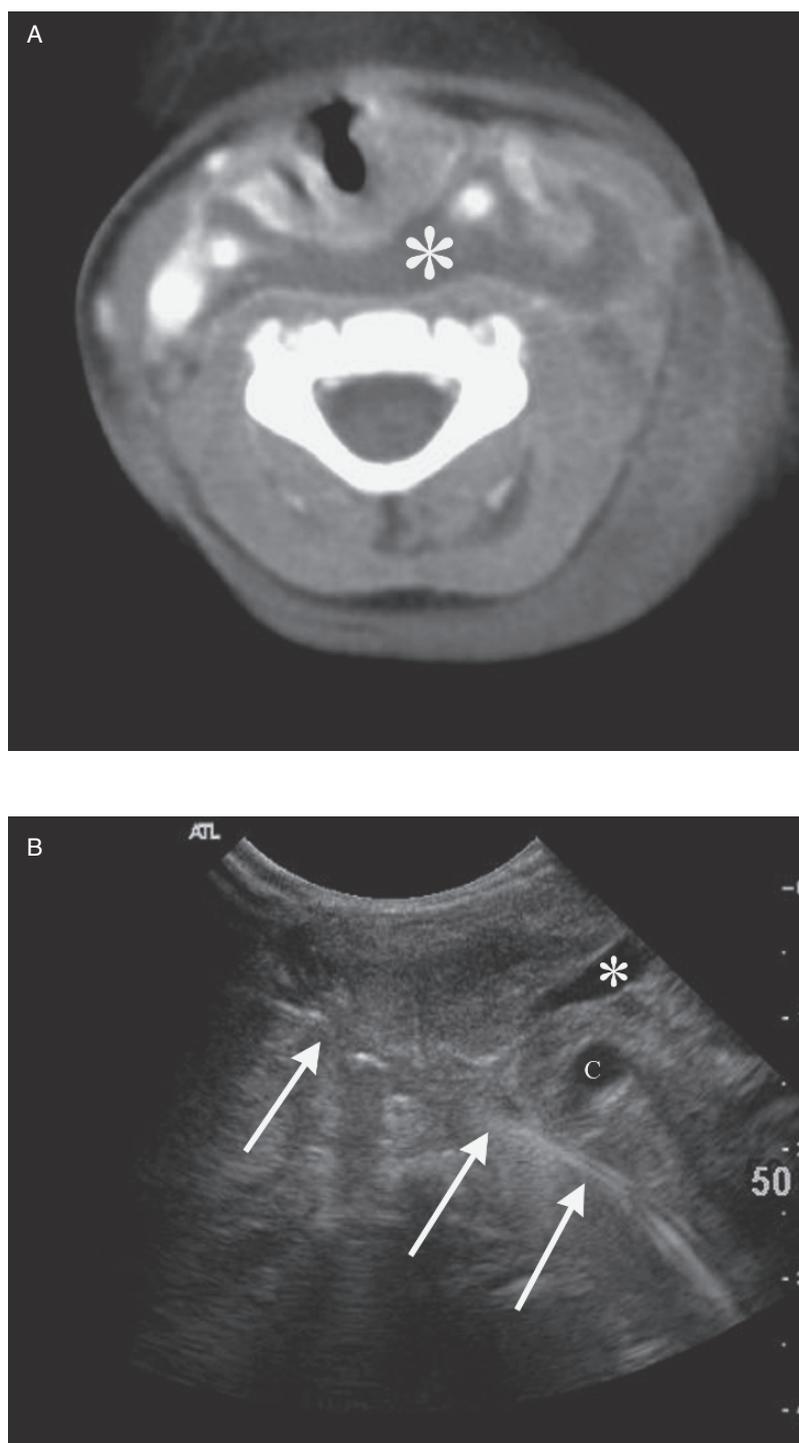


Fig. 4. Patient with fever after tonsillectomy. (A) There is a retropharyngeal abscess (*). (B) Demonstrates ultrasound guidance to place a drainage tube (arrows) into the abscess avoiding the carotid artery (C) and the jugular vein (*).

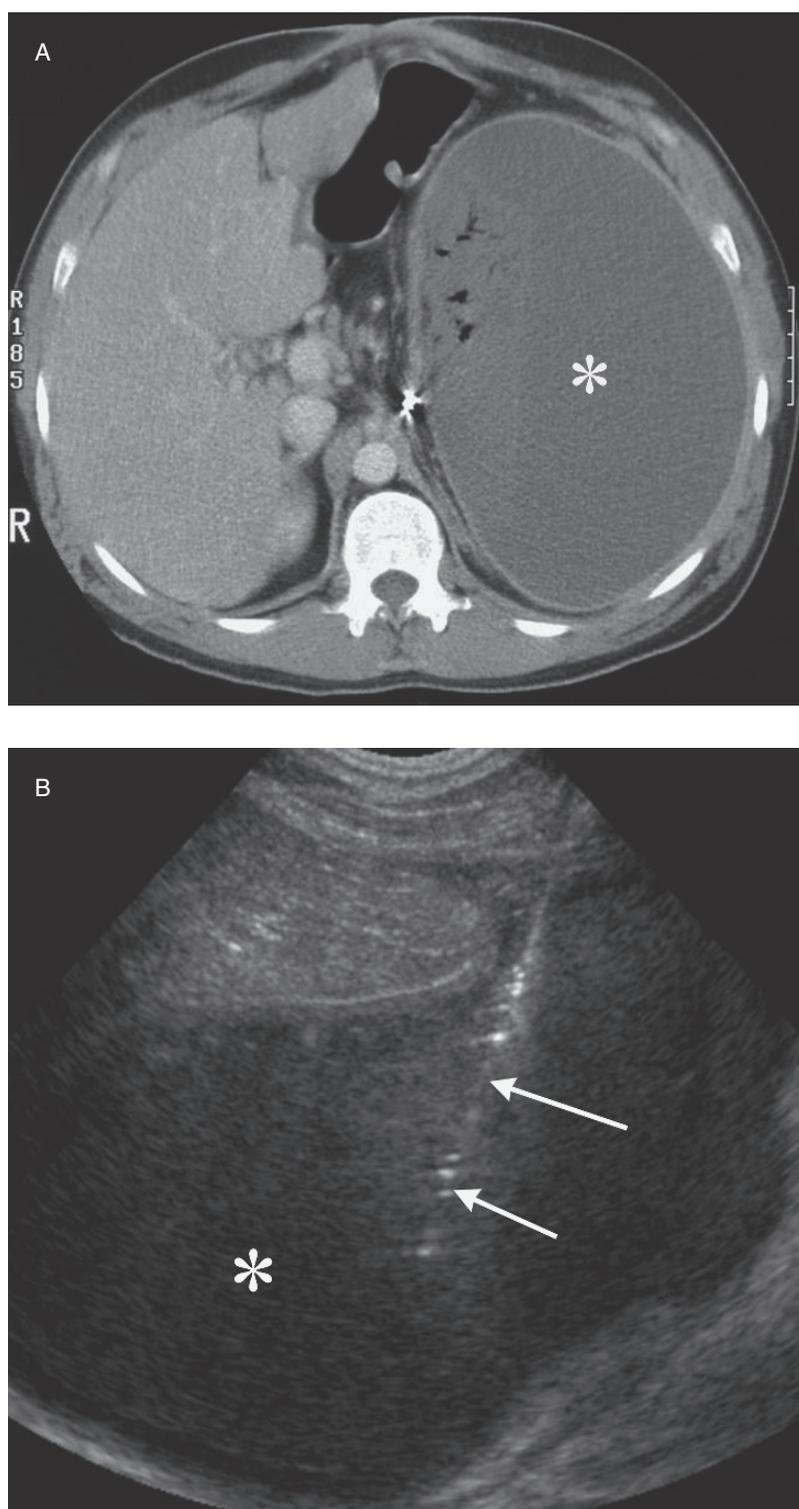


Fig. 5. Splenic abscess. (A) CT shows a large abscess (*) in the splenic bed after splenectomy. (B) Needle (arrows) placement into the abscess (*) with ultrasound guidance.

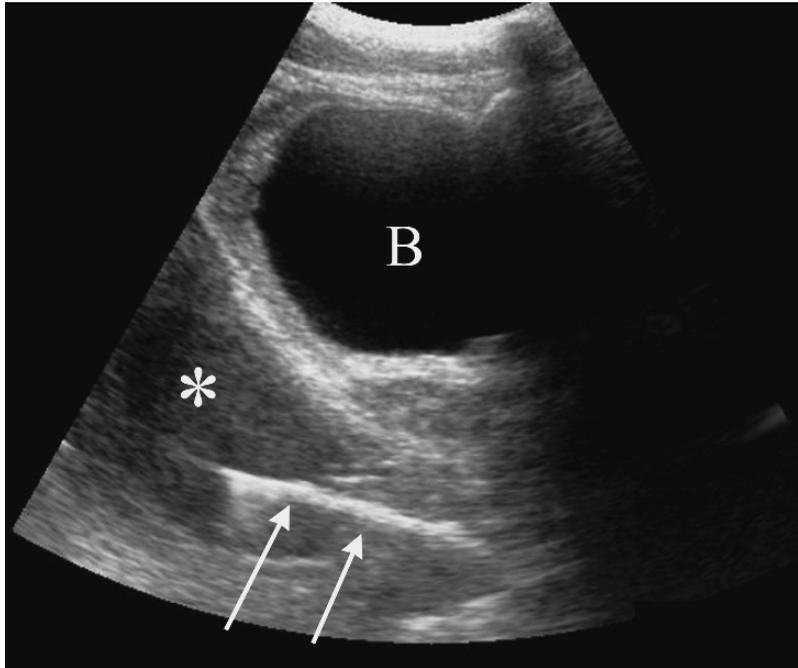


Fig. 6. Pelvic abscess (*) is seen behind the bladder (B). The drain (arrows) is advanced with ultrasound guidance into the abscess.

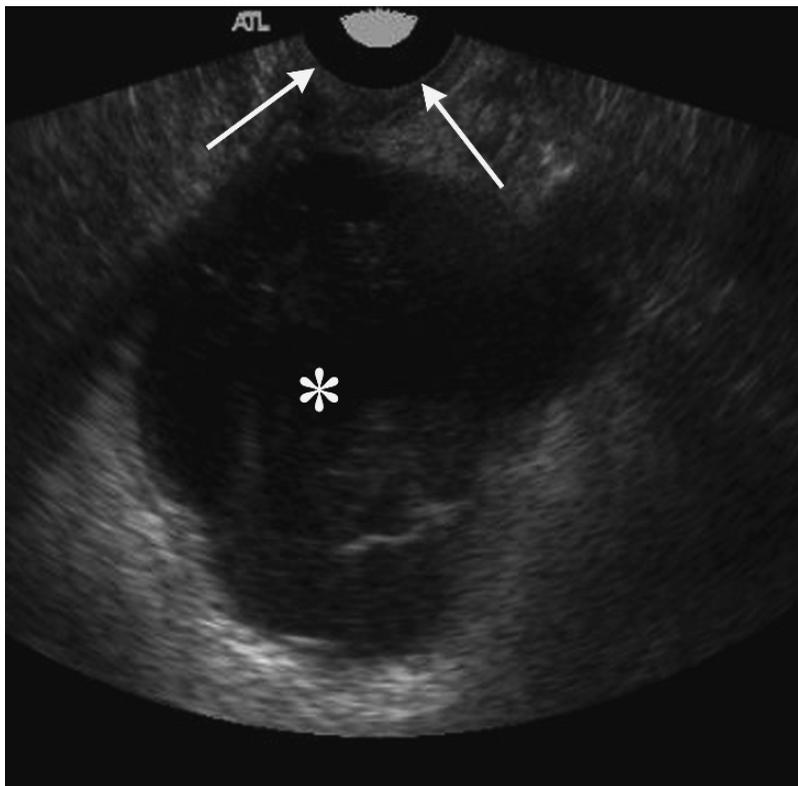


Fig. 7. Transrectal abscess drainage. The ultrasound probe (arrows) will guide placement of a drain into the pelvic abscess (*).

superficial abscesses; however, most abdominal abscesses require larger tubes because of increased fluid viscosity. For abscesses from NEC, 18–20 G intravenous catheters can be used for aspiration with ultrasound guidance (Fig. 8). This is often done portably with the premature infant remaining in an incubator.

The abscess is evacuated as much as possible immediately, and lab samples are sent. Currently, all catheters are placed to bulb suction, as the patients are not compliant with keeping the bag dependent for gravity drainage. The catheter is secured with an adhesive device unless the patient is too small to allow adequate fixation, in which case the catheter should be sutured to the skin. Saline flush with 10 cc is performed every shift. This amount should be subtracted from the tube output, although this is a common charting error.

Assessment is performed at least daily. The patient usually becomes afebrile with drainage less than 10 mL/day within 2 days, and almost always within 4 days; after which time the drain is removed. If tube output continues after 48 hours, we perform a tube injection to evaluate for a fistula.

Image-guided drainage procedures in both pre- and postoperative appendiceal abscesses are successful in 81–100% of patients (7,8,10–13,20). Complications from abscess drainage are uncommon, occurring in up to 11% (9–11), with catheter migration the most common (8,11). Bloody pus is almost universal, but significant hemorrhage

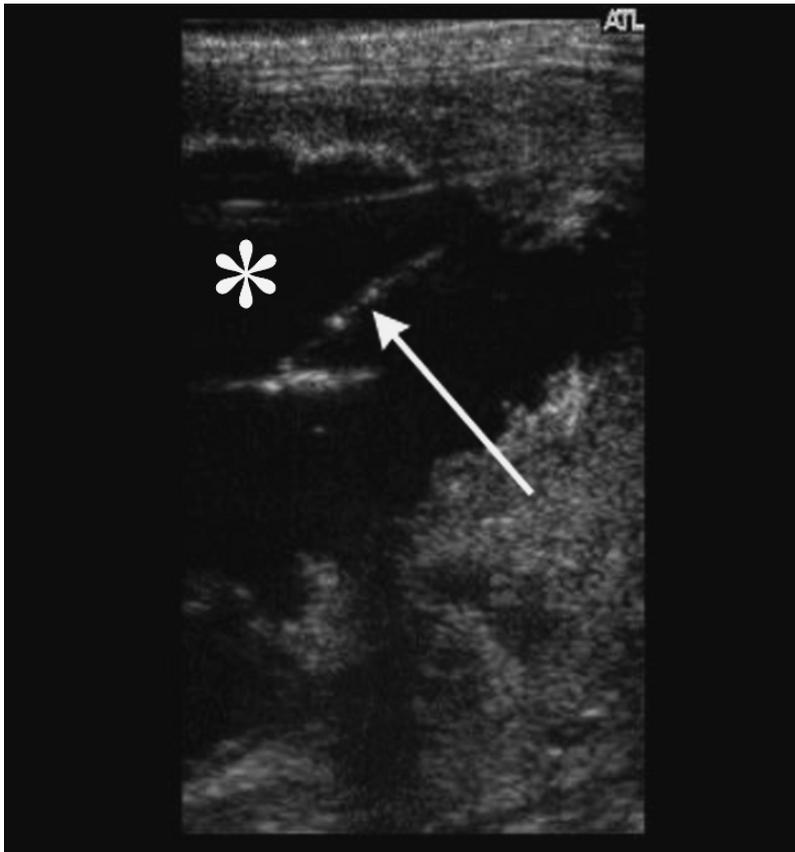


Fig. 8. An IV cannula (arrow) is advanced into abscess (*) after surgery for NEC.

is rare. Vascular injury can occur from any approach, but is probably more common with the transgluteal technique owing to the proximity of the gluteal vessels (9,13,14). The inferior epigastric artery can be injured during transabdominal drainage, but can usually be identified and avoided with ultrasound during guidance for the procedure. Bowel perforation is another risk, but some authors traverse bowel when necessary without significant consequences (7). Inadvertent injury to other organs and the female reproductive tract are possible but rare.

INTESTINAL FISTULA

Intestinal fistulae can originate from both normal and abnormal bowel. Postoperative fistulae may result from anastomotic leak or disruption of bowel, as well as from inadvertent injury during surgery. Spontaneous fistulae tend to originate from diseased bowel (i.e., Crohn's disease, radiation injury, malignancy, and ischemia (21,22)). There is approximately 30% incidence of fistula formation in patients with Crohn's disease (23). Enterocutaneous, enteroenteric or enterocolic, entero- or colovaginal, and entero- or colovesical fistulous communications may be seen, depending on the location and etiology of the underlying disorder. Fistulae from intrinsically normal bowel tend to spontaneously close with conservative management. Those arising from diseased bowel often require surgical intervention (23). The radiologic evaluation in patients with fistulizing disease continues to evolve with advances in imaging technology.

Plain film radiographs are commonly utilized as an initial screening evaluation in the patient with an acute abdomen, but are of limited utility for evaluation of fistulizing disease. On occasion, extraluminal gas may be seen in fistula or abscess cavity, but is often difficult to recognize (24). Positive contrast (barium or water-soluble contrast) examinations utilizing the small bowel follow-through (SBFT), small bowel enteroclysis (SBE), or double-contrast barium enema remain valuable tools for evaluating internal fistulas between loops of bowel or between bowel and other organs. Fistulous tracts may be outlined by contrast or air, and demonstrate direct intercommunication between structures or end blindly in the soft tissues (24) (Fig. 9). Cutaneous fistulas may be studied by direct contrast injection following canalization of the cutaneous opening with an angiocatheter, feeding tube or Foley catheter (fistulogram).

Abdominal and pelvic CT combined with oral and intravenous contrast, or CT enteroclysis, are effective in demonstrating some forms of fistulae, predominately enteroenteric, enterocolic, enterocutaneous, and enterovesical forms (25). Enhancing extraluminal tracts, when associated with inflammatory disease, may be identified containing air or fluid. Enterovesical and colovesical fistulas may demonstrate air within the bladder and focal bladder wall thickening at the site of fistulous communication. Scanning of the pelvis should be obtained prior to intravenous contrast administration so as not to obscure contrast within the bladder lumen originating from the fistulous tract (24). Complications related to fistula formation (i.e., abscess formation), are also readily recognized.

Recent advances in MRI techniques, shortened scan times, and improved tissue contrast have led to increased utility in imaging of the gastrointestinal tract. Contrast enhanced MRI is highly sensitive for depicting active bowel inflammation, distinguishing acutely inflamed bowel with luminal narrowing from fibrotic strictures, defining extraintestinal complications, and in colorectal disease. MRI is now the imaging study of choice for evaluating complex perianal fistulae as are commonly present in Crohn's disease. MRI can accurately determine a fistulous tract's relationship



Fig. 9. Enterocolonic fistulae. Anterior radiograph of the abdomen during SBFT. Multiple inflamed segments of bowel are demonstrated consistent with Crohn's disease. Contrast containing enterocolonic fistulae (arrowheads) are demonstrated with contrast extending into the rectum. Blind-ending sinus tracts are noted centrally.

to the sphincters and levator ani, which is crucial for therapy. Fistulae and sinus tracts are hypointense (dark) on T1-weighted sequences and hyperintense (bright) on T2-weighted sequences, depending on the amount of fluid, edema, and inflammation (24).

The interventional radiological management of enteric fistulae centers on percutaneous drainage of associated abscesses (e.g., intraabdominal or psoas abscesses). In this setting, the catheter may drain longer than in more common abscess settings; however, when combined with medical therapy and bowel rest, will most often support fistula closure.

SMALL BOWEL OBSTRUCTION (SBO) AND ILEUS

Mechanical bowel obstruction results from an anatomic obstruction to flow of intestinal contents. In adynamic or paralytic ileus, there is reduced or absent peristalsis in all or a portion of the intestinal tract without actual mechanical obstruction. Differentiation between obstruction and ileus in the postoperative patient is difficult because the clinical presentation is clouded by incisional pain, narcotics, abdominal distention, and normal adynamic ileus (26). The radiologic evaluation relies on direct communication between the radiologist and surgeon to avoid unnecessary delays in treatment. Prompt and precise imaging diagnosis allows triage of patients into a surgical or non-surgical management category.

Plain film radiography remains an important and frequently requested initial examination in patients with suspected obstruction. Supine and erect plain films of the abdomen should be obtained. In patients who are too sick to stand or in the young child, lateral decubitus views should be obtained (27,28). Additional views include the horizontal beam decubitus films as well as the prone and crossfire prone views. These views assist in redistributing air (negative contrast) within the gastrointestinal tract, allowing more accurate

assessment of the presence or absence of air in the more distal part of the bowel (29). On the supine and prone radiograph, gas is normally present in the stomach, colon, and rectum. The normal small bowel gas pattern includes absence of small bowel gas or presence of small amounts of gas within up to four nondistended loops (<2.5 cm in diameter). On the normal upright or decubitus film, air–fluid levels are almost always seen in the stomach and occasionally in the cecum, ascending colon, and terminal ileum. There is normal distribution of gas and stool within a non-distended colon. A nonspecific bowel gas results when at least one loop of borderline or mildly distended small bowel (2.5–3 cm in diameter) with three or more air–fluid levels is present on upright or decubitus radiographs. The colonic and fecal distributions are normal or with mild distention. A pattern of probable SBO occurs when multiple gas- or fluid–fluid loops of dilated small bowel loops are seen with a relatively small or moderate amount of colonic gas. An unequivocal SBO pattern is defined as dilated gas or fluid-filled small bowel loops in the setting of a gasless colon (26) (Fig. 10). There should be no difficulty in distinguishing between unequivocal SBO

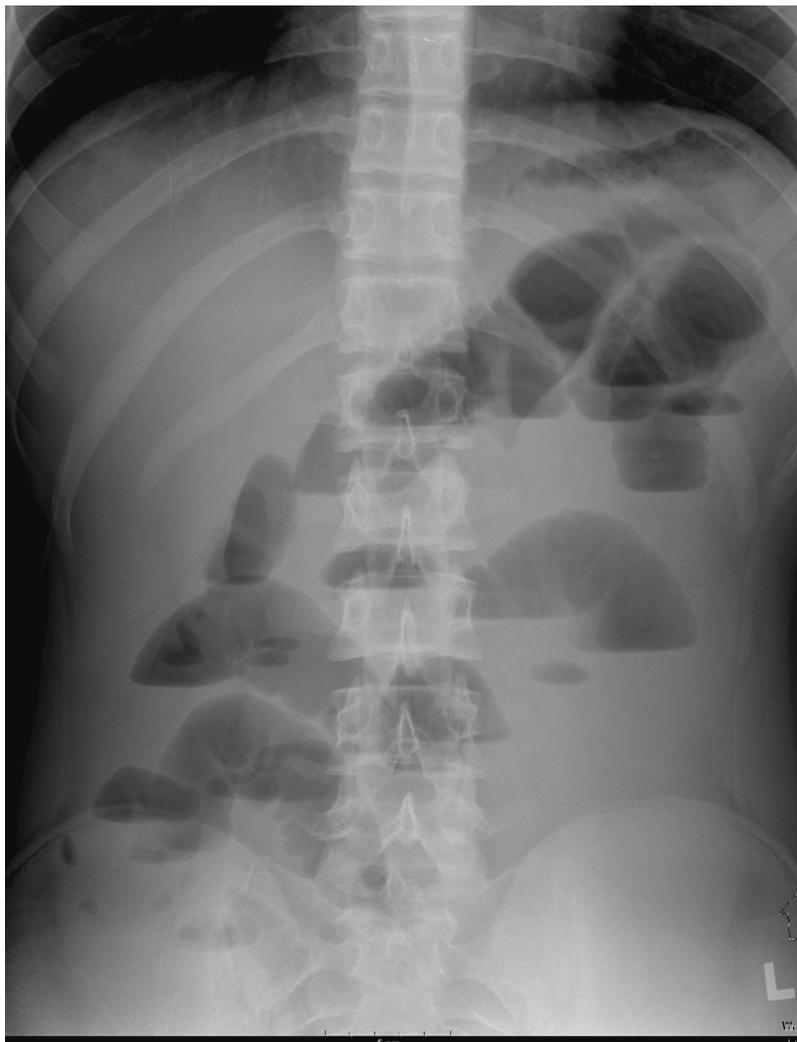


Fig. 10. SBO. Upright view of the abdomen demonstrating unequivocal SBO. Multiple distended loops of small bowel are seen with air–fluid levels. No colonic gas can be identified.

and the diffuse and proportional dilation of the small bowel and colon characteristic of paralytic ileus (Fig. 11).

If there is doubt as to the diagnosis after plain films, contrast studies of the bowel help to separate mechanical obstruction from ileus. If colonic ileus or distal colonic obstruction with an incompetent ileocecal valve is suspected, barium enema is fast and inexpensive. The SBFT examination has been used to triage patients with suspected SBO into surgical and nonsurgical management categories, but has largely been replaced by the widespread use of abdominal CT. The major disadvantages to the SBFT include: inability of patients with suspected SBO to ingest large quantities of contrast; difficulty in assessing distensibility and fixation of the small bowel; flocculation and dilution of barium in high-grade obstruction with incomplete bowel opacification; and the length of exam—hours or longer before contrast reaches the point of obstruction (26).

Enteroclysis challenges the distensibility of the bowel wall and exaggerates the effects of mild or subclinical obstruction (Fig. 12). Intubating the small bowel bypasses the stomach and allows direct delivery of nondiluted barium or iodinated contrast (CT enteroclysis) directly into the jejunum. Advantages include: controlled infusion of contrast promotes antegrade flow toward site of obstruction despite diminished



Fig. 11. Ileus. Supine radiograph of the abdomen demonstrating diffuse gaseous distention of the small and large bowel.



Fig. 12. Spot radiograph from an enteroclysis demonstrating differential dilatation of proximal versus non-dilated (arrow) mid-jejunal loops secondary to low-grade adhesive disease.

bowel peristalsis; facilitates detection of fixed and nondistensible bowel segments; high sensitivity (100%) and specificity (88%) for SBO and high accuracy in determining the cause of obstruction (86%); detects multiple levels of obstruction; and most importantly, is highly reliable in diagnosing partial low-grade obstruction or excluding the diagnosis compared to conventional CT or SBFT (26). SBO is excluded by enteroclysis when contrast passes unimpeded through normal caliber small bowel loops from duodenum to right colon. Mechanical obstruction is confirmed by the demonstration of a transition zone from a proximally distended segment to collapsed distal segment beyond the obstruction (26,30).

Conventional CT is highly sensitive in distinguishing high-grade small bowel obstruction from ileus (Fig. 13), and should be the initial study of choice in patients with suspected SBO in the immediate postoperative period or when the clinical presentation suggests underlying abscess, closed loop obstruction, or strangulation (26,31). In the absence of clinical suspicion of these conditions, or if the CT findings are equivocal for obstruction, CT enteroclysis may establish the diagnosis by providing volume-challenged distention of bowel loops. Water-soluble contrast is initially infused through an enteral catheter placed in the proximal small bowel at fluoroscopy, followed immediately by CT during continued contrast infusion. This technique overcomes the insensitivity of conventional CT for diagnosing lower grades of obstruction and is equivalent to barium enteroclysis in patients with low-grade partial SBO. CT

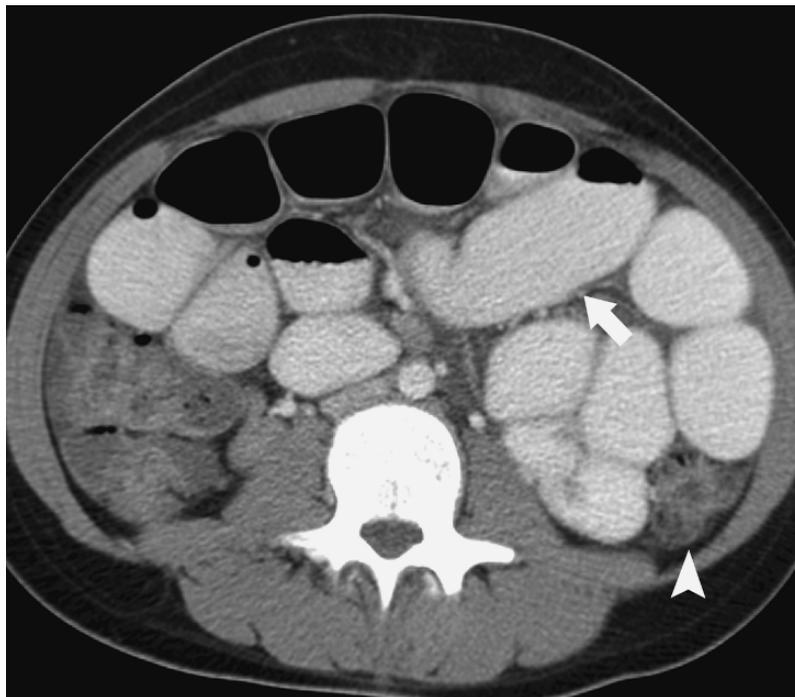


Fig. 13. SBO. Axial image from abdominal CT demonstrating dilated small bowel (arrow) and collapsed colon (arrowhead). Not shown is high-grade distal ileal obstruction caused by severe inflammatory bowel disease.

enteroclysis with multiplanar reformatting can map the location of parietal and visceral adhesions, helping to select appropriate access sites into to the peritoneal cavity (32).

MRI currently has a limited role in mechanical SBO. MRI enteroclysis has the potential to change the assessment of the small bowel through its multiplanar imaging capabilities, its lack of ionizing radiation, and the functional information and soft tissue contrast that it can provide (26,33). Further research and experience may clarify whether MRI imaging and enteroclysis will play a role in evaluating SBO or be used as a problem solving examination.

ESOPHAGEAL DISORDERS

Foreign Body

Ingestion of foreign bodies is common in the young child, coins being the most frequent. Fortunately, the large majority of swallowed objects pass through the gastrointestinal tract without complication. Foreign bodies can become lodged in the esophagus at characteristic locations of physiologic narrowing. The thoracic inlet is most common (75%), followed by the level of the left mainstem bronchus (20%), and least frequently just proximal to the gastroesophageal junction (5%). Patients with underlying esophageal pathology (i.e., caustic ingestion, tracheoesophageal fistula repair, or vascular ring), have foreign body retention in nonphysiologic positions (34,35).

The initial radiographic evaluation should include radiographs of the neck, chest, and abdomen to evaluate the entirety of the intestinal tract from mouth to anus.

Esophageal foreign bodies are characterized as radiopaque or radiolucent, sharp or dull, and single or multiple (34). The imaging appearance will influence treatment options. Complications of foreign body retention should be sought and include: high-grade esophageal obstruction with air–fluid levels; tracheal narrowing owing to local edema or mass effect; perforation with pneumomediastinum; mediastinal migration of the foreign body; and a mediastinal mass secondary to abscess formation.

A contrast esophagram is required for nonradiopaque foreign bodies. The radiolucent foreign body may be outlined by contrast or demonstrated as an irregular contour at the base of the contrast column in complete obstruction (Fig. 14). Contrast extension beyond the confines of the esophageal lumen defines perforation. CT can identify small foreign bodies not seen on standard radiographs, further characterize the features of a foreign body, and evaluate the paraesophageal anatomy for edema, inflammation, or abscess formation. Esophageal foreign bodies associated with prior esophageal surgery may be retrieved via an endoscope or via transoral interventional radiological snare retrieval with fluoroscopic guidance. Fluoroscopically guided snare removal will either be successful or define foreign bodies that are adherent with granulation tissue that require surgical removal.

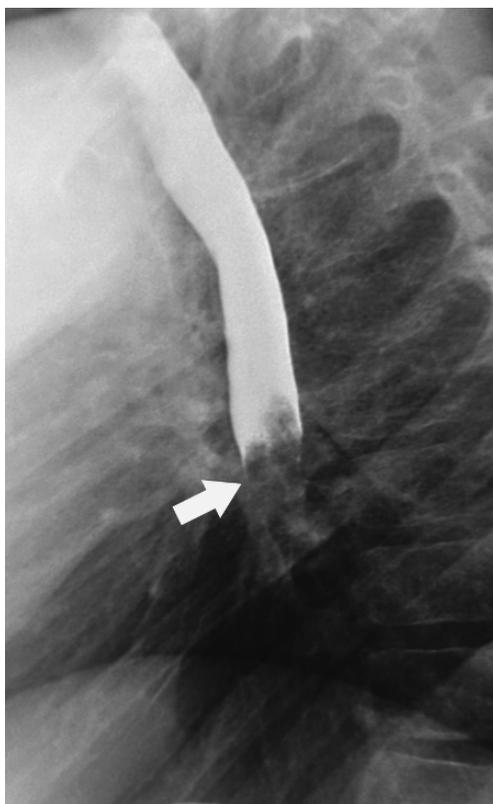


Fig. 14. Radiolucent foreign body. Lateral view from an esophagram demonstrating complete esophageal obstruction due to meat impaction (arrow).

Achalasia

Achalasia is a primary esophageal motility disorder characterized by increased lower esophageal sphincter (LES) tone and decreased lower esophageal peristalsis. Histologic degeneration of the myenteric plexus of Auerbach is the common cause of achalasia and leads to loss of inhibitory postganglionic neurotransmitter (nitrous oxide and vasoactive intestinal peptide) production, which is responsible for the relaxation of the LES and coordinated esophageal peristalsis (36). Impaired emptying and gradual esophageal dilation are responsible for the development of clinical symptoms (37). Patients present with progressive dysphasia to solids and liquids, chest pain, and regurgitation of undigested food.

Achalasia may be identified on the upright chest radiograph as a dilated esophagus with an air–fluid level and a paucity of gastric air. A barium esophagram should follow as the initial diagnostic examination of choice. Classic findings identified in achalasia include smooth tapered narrowing of the distal esophagus (“bird’s beak” appearance) associated with atonic dilation of the lower two-thirds of the esophagus and a column of contrast in the esophageal lumen (37) (Fig. 15). CT scans are not generally recommended in the evaluation of achalasia unless there is suspicion of a mass

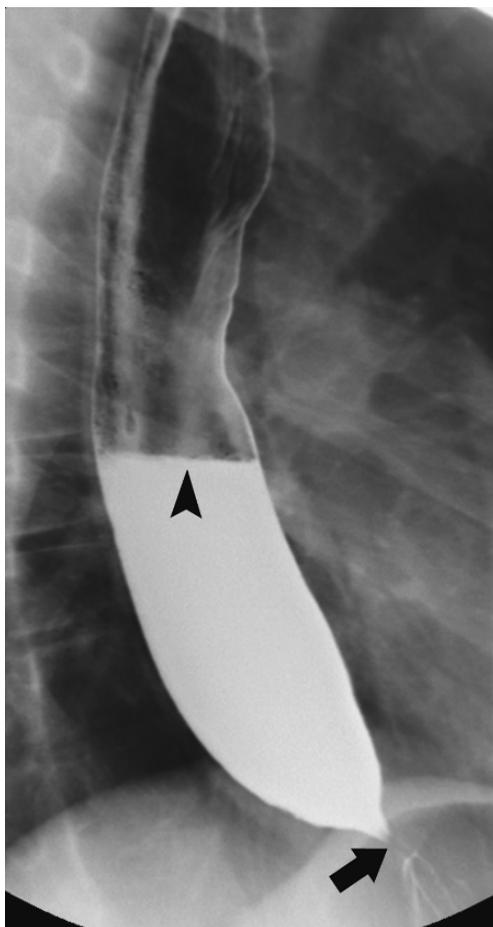


Fig. 15. Achalasia. Lateral view from an esophagram demonstrating a dilated esophagus, air–fluid level (arrowhead) and obstructed distal esophagus (arrow).

(adenocarcinoma, lymphoma, or others) at the gastroesophageal junction mimicking the findings of achalasia. Radiological intervention following surgical treatment of achalasia centers on balloon dilation of postmyotomy strictures. Balloon dilation with fluoroscopic guidance of distal esophageal stricture associated with achalasia is safe, to a diameter of 30 mm.

Esophageal Perforation

Instrumentation of the esophagus accounts for approximately two-thirds of all reported cases of esophageal perforation and can be seen following esophagoscopy, esophageal dilatation (bougienage or pneumatic), sclerotherapy, intraesophageal tube placement, and endotracheal intubation. Other causes include spontaneous perforation (Boerhaave's syndrome), foreign body ingestion, blunt chest trauma, operative injury, tumor, and severe esophagitis (38,39). Rapid development of necrotizing mediastinitis, empyema, sepsis, and multiorgan failure account for the high mortality associated with esophageal perforation. Accurate diagnosis and early treatment are essential to the successful management of patients.

Lateral neck films may detect subcutaneous emphysema before identification on chest radiographs or by physical examination in cervical esophageal perforation (40). When thoracic or abdominal perforation is suspected, posteroanterior and lateral radiographs of the chest as well as an upright or decubitus view of the abdomen should be obtained. Chest radiographs detect 90% of esophageal perforations; however, they can be negative if taken too early. Soft tissue and mediastinal emphysema are seen after 1 hour, whereas pleural effusions and mediastinal widening take several hours to develop (39,40). The presence of pleural effusion, pneumomediastinum, subcutaneous emphysema, hydrothorax, hydropneumothorax, or subdiaphragmatic air on radiographs is highly suggestive of esophageal perforation (41).

The contrast esophagram remains the standard for diagnosis of esophageal perforation (Fig. 16). Water-soluble contrast is recommended for initial screening of suspected perforation. Contrast extravasation is identified in only 50% of cervical and 75–80% of thoracic perforations. If no perforation is initially identified, barium esophagography is recommended. Barium improves detection of small primary or unsuspected secondary perforations. The detection rate increases to 60% for cervical and 90% for thoracic perforations (42). Contrast studies have an overall 10% false negative rate.

CT identifies esophageal perforations that are difficult to diagnose or when contrast esophagrams cannot be performed. Abnormal findings suggestive of perforation include extraluminal air in the soft tissues of the mediastinum, esophageal thickening, visible communication of the air-filled esophagus with a contiguous mediastinal or paramediastinal air–fluid collection, or abscess cavities adjacent to the esophagus in the mediastinum or pleural space. Left-side pleural effusions are highly suggestive. In patients who fail to improve after initial treatment, CT assists in localizing pleural fluid or abscess collections amenable to interventional radiological drainage catheter placement.

Complications of Fundoplication

Gastroesophageal reflux disease (GERD) is a common condition for which medical management is effective in the large majority of patients. Those with intractable vomiting, persistent esophagitis, apnea, and pulmonary infections require further surgical management (43). Antireflux surgery has been shown to be highly effective

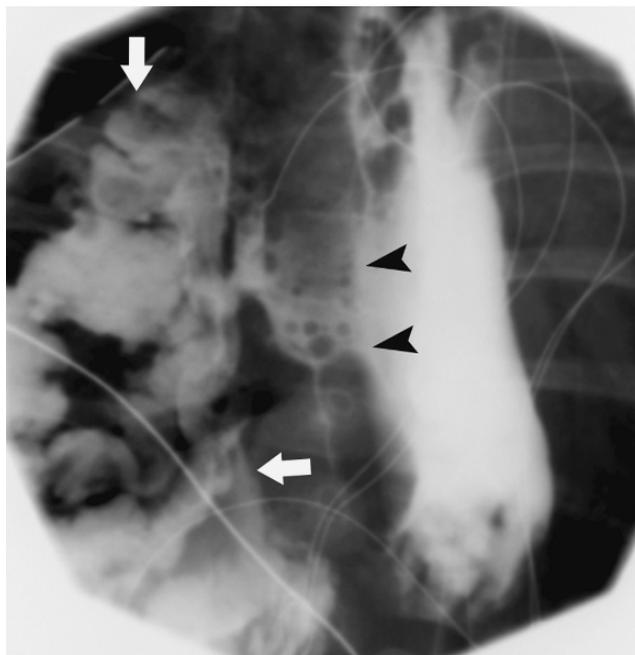


Fig. 16. Esophageal perforation. Anterior view from an esophagram demonstrating a large thoracic esophageal perforation (arrowheads) with extravasation into the right thorax (arrows).

in managing complications related to GERD. With advances in minimally invasive techniques, laparoscopic Nissen fundoplication has become the preferred surgical technique (44). Despite a success rate of more than 90% and low mortality, antireflux surgery is not without complications.

The upper gastrointestinal series (UGI), with barium administered orally or through a gastrostomy tube, is the study of choice to evaluate the integrity and the function of the wrap. Loosening or breakdown of the fundoplication is identified by contrast filling the folds of the wrap. Small amounts are generally inconsequential, whereas larger amounts reflect more significant loosening. In complete breakdown of the wrap, there is loss of the normal “pseudotumor” identified at the fundus. Hiatal hernias are seen when the fundoplication and upper stomach herniate through the hiatus. The wrap may remain intact. Paraesophageal hernias represent extension of a portion of the wrap through the hiatus, typically along posterolateral margin of the left side of the wrap where there is an absence of sutures. The key differentiating point from hiatal hernia is that the gastroesophageal junction remains below the diaphragm. A malpositioned fundoplication results from partial disruption of the sutures, allowing distention of the fundus above the remaining sutures. This has a similar appearance to the paraesophageal hernia, but the deformed fundus remains entirely below the diaphragm. Esophageal obstruction can result from an excessively tight wrap leading to esophageal dilation and delayed transit of contrast through the fundoplication (Fig. 17). Partial esophageal obstruction may be found in the immediate postoperative period secondary to edema; persistent obstruction, however, requires treatment (43).

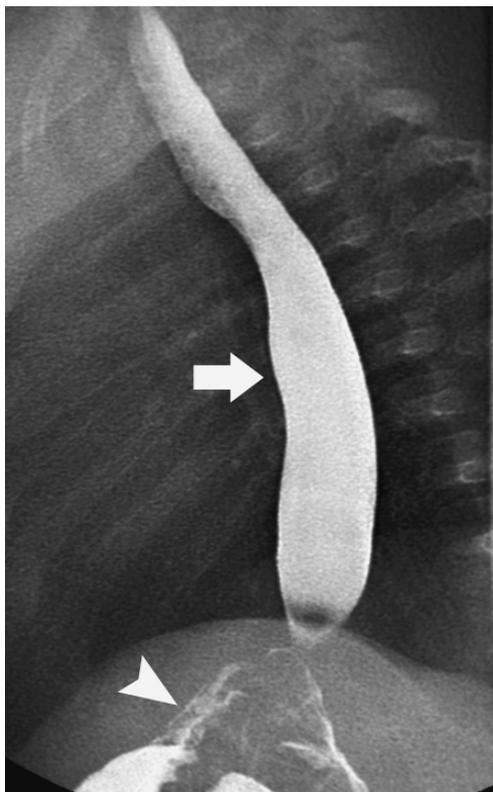


Fig. 17. Delayed esophageal transit. Lateral view from an upright esophagram demonstrating an obstructed esophagus following Nissen fundoplication (arrow). Contrast in gastric fundus shows intact Nissen (arrowhead). Contrast was placed through gastrostomy tube prior to esophagram.

Esophageal Strictures

Esophageal strictures in the pediatric age group are most frequently benign. Strictures may be congenital, but are most frequently acquired following surgery (repair of esophageal atresia, interposition surgery, restrictive Nissen fundoplication), caustic or foreign body ingestion, achalasia, infection (herpes virus, candidiasis), inflammation (epidermolysis bullosa, chronic granulomatous disease), reflux esophagitis, and following sclerotherapy of varices. Anastomotic strictures following esophageal atresia repair occur in 30–70% of patients and are the most commonly acquired stricture of childhood (45).

The radiologic evaluation of esophageal strictures requires an esophagram with water-soluble or barium contrast (46). The diagnosis of stricture is confirmed when a persistent area of circumferential narrowing is identified within the esophageal lumen. The involved segment shows lack of distention during passage of the contrast bolus. The proximal esophagus is often dilated with delayed passage of the contrast bolus through the stricture. The degree of dilation and contrast delay is determined by the severity of the stenosis. Retained secretions, ingested food matter, or foreign bodies may be seen within the lumen of the proximal esophagus (Fig. 18). Esophageal strictures following surgical intervention are most commonly found with repair of esophageal atresia and are effectively and safely treated with low-profile angioplasty balloon dilation. Long-term



Fig. 18. Esophageal stricture. Anterior view from an esophagram demonstrating a focal esophageal stricture (arrow) with dilated proximal esophagus and retained food within the lumen. Patient had esophageal atresia repair as an infant.

balloon dilation of esophageal anastomotic strictures is successful in nearly 100% of patients, and success is greatest when started within 6 months of surgery, in patients without GERD, and when solid food ingestion is permitted to maintain the patent esophageal lumen. The incidence of perforation from balloon dilation of anastomotic strictures ranges from 0–1.8% (47,48).

Anastomotic Leak and Recurrent Fistula

Complications seen following repair of esophageal atresia include anastomotic leak, recurrent tracheoesophageal fistula, and stricture (discussed previously) formation. The incidence of anastomotic leak following repair is approximately 20% (49). Causes include: the use of silk sutures; excessive anastomotic tension; and excessive distal esophageal mobilization. A greater incidence of anastomotic leak is noted in patients who undergo colonic interposition, attributable to impaired blood supply in the cephalad end of the graft (49). Clinical suspicion of leak occurs when saliva is seen in the chest tube. An esophagram confirms the clinical suspicion by demonstrating contrast extravasation from the anastomotic site into the mediastinum or pleural space.

Recurrent tracheoesophageal fistulae are seen in approximately 3–12% of repairs. The etiology is related to an anastomotic suture line leak with erosion through the previous site of repair (49). Diagnosis may be difficult and delayed for years. A contrast esophagram may directly demonstrate the fistula (Fig. 19); however, an esophagram performed after placement of a nasogastric or orogastric tube may be

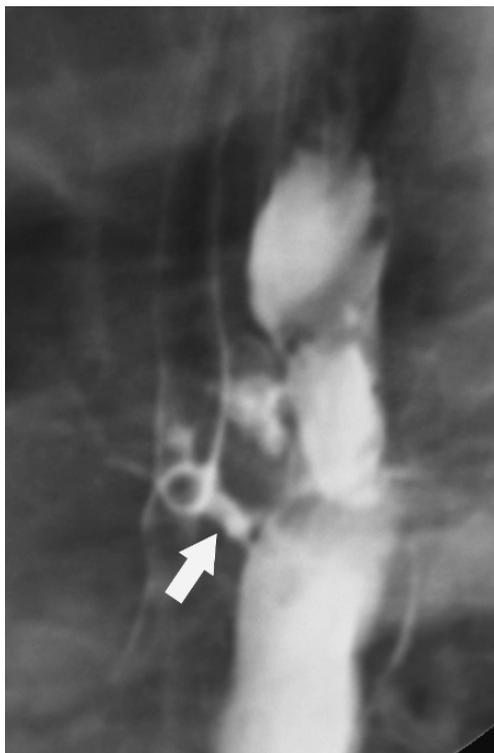


Fig. 19. Recurrent tracheoesophageal fistula. Oblique view from an esophagram demonstrating a recurrent fistula between the esophagus and the trachea (arrow). Patient had prior repair of esophageal atresia with tracheoesophageal fistula. Coughing with feeds was the clinical presentation.

required. Repeated contrast injections during withdrawal of the tube achieve greater local luminal distention and increased likelihood of fistula visualization. Direct canalization of the fistula is less often performed, but will directly opacify the fistula or serve as site for injection of methylene blue during bronchoscopy (35).

STOMACH

Gastroparesis

Gastroparesis is characterized by impaired gastric emptying in the absence of a mechanical gastric outlet obstruction. Clinical symptoms include early satiety, postprandial bloating, nausea, vomiting, and abdominal pain. Nutrient composition (volume, osmolarity, osmolality, caloric density, viscosity, Ph, and type of macronutrients), degree of gastric distention, and intestinal feedback from concentration-dependent chemoreceptors along the proximal intestine all interact to determine gastric transit (50). Water or human milk empties faster than formula, and liquids empty faster than solids. Medium chain triglycerides empty faster than long-chain triglycerides.

Nuclear medicine scintigraphy remains the gold standard method for measuring gastric emptying. Gastric emptying studies are performed by combining a radiopharmaceutical (technetium sulfur colloid) with food and measuring the residual radioactivity within the stomach over time (generally 1, 2, and 4 hours) as a percentage of the initial gastric activity at ingestion (51). Radiolabeling of both liquids and solids is

feasible, although the dynamics of emptying are different. Solid meal scintigraphy is preferred over liquids for detecting delayed gastric emptying because the rate of solid emptying will be affected earlier by abnormalities in receptive relaxation and tone generated by the fundus, as well as by abnormalities in motility and coordination of antropyloroduodenal smooth muscle (52).

Regardless of the technique used, normal values of gastric emptying vary widely among different individuals and day-to-day within the same patient who undergoes repeated tests. Lack of standardization of test meals makes published normal values of gastric emptying difficult to compare between institutions. In routine practice, a T1/2 (time for 50% gastric emptying to occur) or, more commonly, the percentage of residual gastric activity at 1, 2, and potentially 4 hours is calculated (Fig. 20). The published range of normal gastric emptying for solid phase is 10–50% retention at 2 hours. For those patients with normal or borderline emptying (45–55% retention) at 2 hours, extending the study out to 4 hours has been suggested (51). The range of normal gastric retention at 4 hours is narrower (0–10% retention) and a greater number of symptomatic patients may be demonstrated to have delayed gastric emptying at this time. The key is to establish a standard protocol for

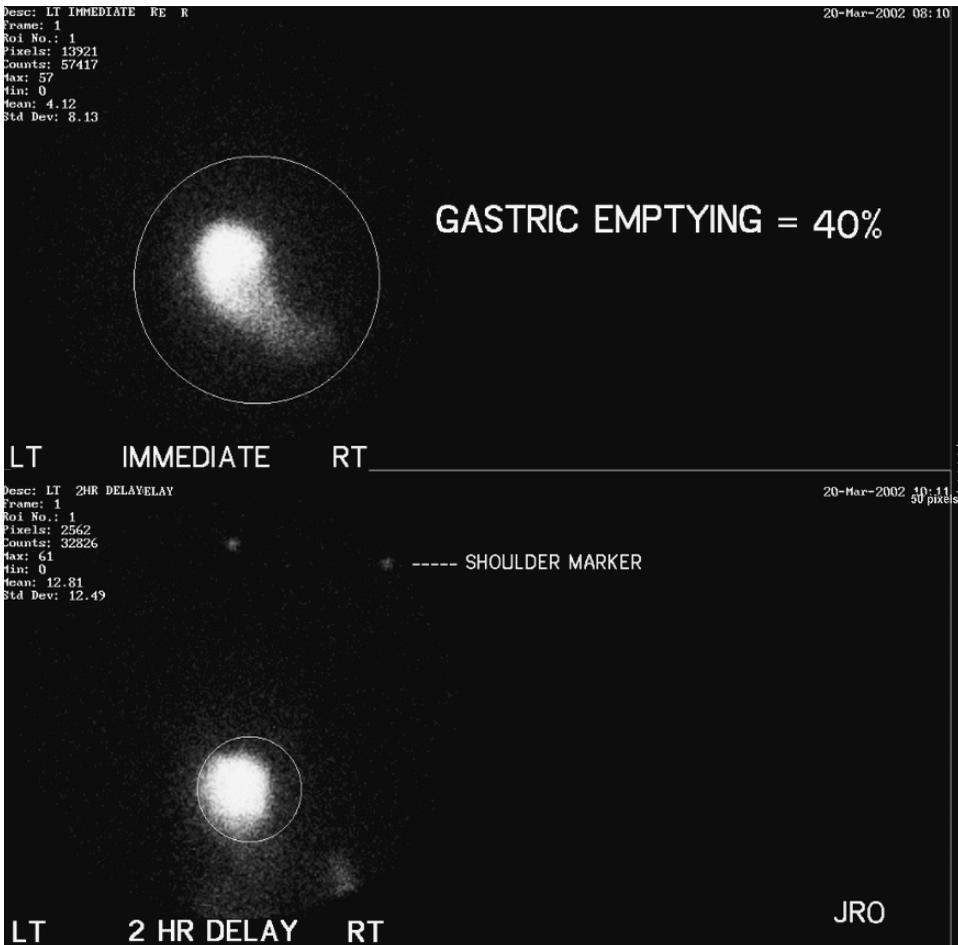


Fig. 20. Gastric emptying scan demonstrating delayed emptying at 2 hours.

scintigraphy at one's own institution so that variations in gastric emptying are physiologic and not technically related.

Gastric Perforation

The etiology of the gastric perforation is age dependent. In the newborn infant, gastric perforation is abrupt in onset and occurs in 96% during the first week of life. Proposed etiologies of perforation include: hypoxia or stress-induced necrosis; gastric hyperacidity in the first few days of life; gastric ulceration; overdistention of the stomach caused by distal obstruction or mechanical ventilation; prematurity; indomethacin or dexamethasone therapy; vigorous respiratory resuscitative measures; and iatrogenic (53). Perforation in the older child and adult most commonly arise from peptic ulceration, although iatrogenic and traumatic injuries, postoperative complications, and necrotic or ulcerated malignancies are also reported (54).

Supine, upright, left lateral decubitus and crossfire supine views of the abdomen should be obtained, depending on the age and the clinical status of the patient. The characteristic finding of gastric perforation is pneumoperitoneum. Associated absence of an air–fluid level in the stomach and decreased bowel gas are very suggestive of the diagnosis. Recognition of free air on a supine abdominal radiograph include: increased lucency over the liver compared to the abdominal wall musculature (Fig. 21), the “football” sign (air outlining the falciform ligament) (Fig. 22); The “inverted-V” sign (air outlining the median umbilical folds); and the “Rigler” sign (air on both sides of the bowel wall) (Fig. 22). On the upright abdominal view, air collects between

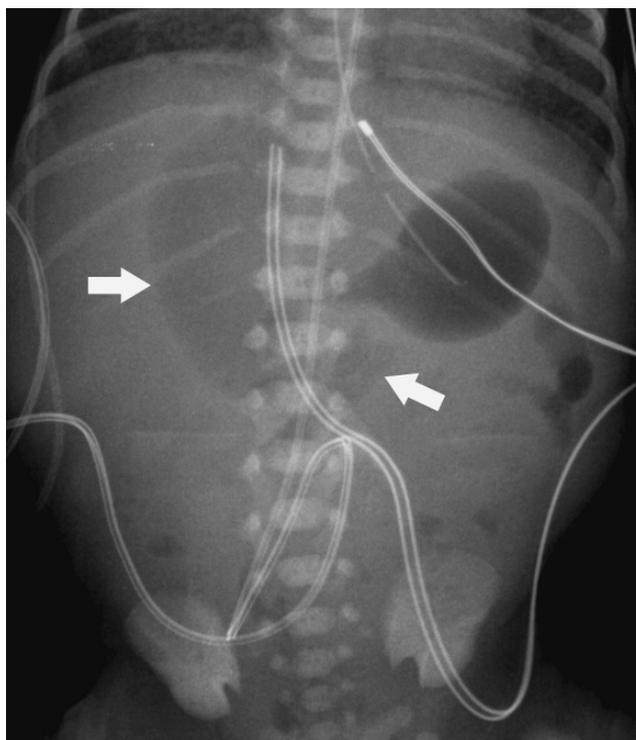


Fig. 21. Pneumoperitoneum. Free air on supine radiograph (arrows) seen as abnormal lucency in the right upper quadrant.

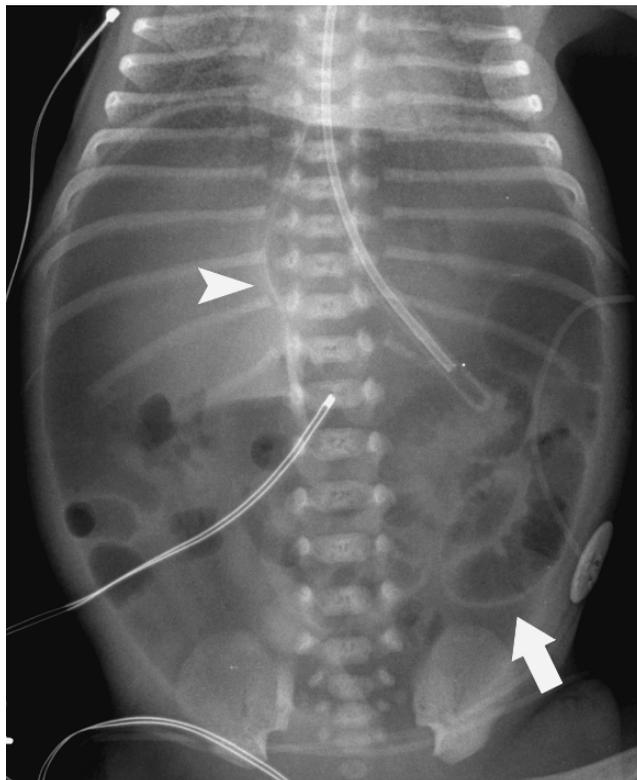


Fig. 22. Pneumoperitoneum. Massive pneumoperitoneum on supine radiograph. Free air outlines the falciform ligament (arrowhead) and is seen on both sides of the bowel wall (arrow).

the diaphragm and the upper abdominal viscera. On the decubitus view, air collects between the liver and anterior abdominal wall. A triangular air collection is seen on supine crossfire views when air collects between the anterior abdominal wall and two adjacent loops of bowel. Free air may not be seen initially if the perforation is small, self-sealed, well contained by adjacent organs, retroperitoneal, or the result of fluid extravasation rather than air (54). The reported sensitivity in the detection of extraluminal air by plain film radiography is 50–70%.

When perforation is suspected, but not demonstrated on abdominal radiographs, a contrast examination may be obtained. A water-soluble contrast agent is used initially in any suspected case of intestinal perforation. If a perforation is not identified on the water-soluble contrast exam, barium administration should immediately follow. Identification of any extraluminal contrast confirms the diagnosis of perforation (Fig. 23). Free extravasation of contrast into the peritoneal space, however, is detected in approximately 50% of patients with anterior perforation of the stomach, the other half forming walled-off collections (55). Penetrations of the posterior wall of the stomach may involve the pancreas, lesser omentum, transverse mesocolon, liver spleen, biliary tree, or colon. Less than 50% of these patients will demonstrate contrast extravasation, likely caused by early sealing of the leak.

Abdominal CT is the recommended imaging modality for detecting gastrointestinal perforations that are not identified by either plain films or contrast examinations, and



Fig. 23. Gastric perforation. Extravasation of contrast (arrow) is demonstrated through posterior gastric wall perforation.

in atypical clinical presentations. CT is superior to plain films in demonstrating small collections of free intraperitoneal air or small collections in the retroperitoneum. CT images need to be viewed in wide window settings (lung window) that assist in discriminating low-density air from fat (Fig. 24). In addition, the site and cause of the perforation as well as associated complications of phlegmon, abscess, and peritonitis can be assessed (54). Administration of oral contrast is recommended, although extraluminal extravasation is not a frequent CT finding in perforation. Diagnosis of perforation is based on direct findings of extraluminal air or luminal contrast material and direct visualization of a focal ulceration or discontinuity of the stomach wall. An indirect sign of perforation is an inflammatory phlegmon in direct continuity with the wall of the stomach.

Gastrostomy Tube Dislodgement

Insertion of gastrostomy tubes has become commonplace, particularly for care of the special-needs child. Three methods of gastrostomy placement are available today: surgical gastrostomy, percutaneous endoscopic gastrostomy (PEG), and interventional radiological percutaneous gastrostomy (56). Whether done surgically or percutaneously, gastrostomy placement requires a tract be created through the anterior abdominal wall into the stomach for an enteral feeding device. Only the surgical technique involves suturing of the stomach to the anterior abdominal wall. The other techniques rely on scarring to take place between the stomach and the anterior abdominal wall.

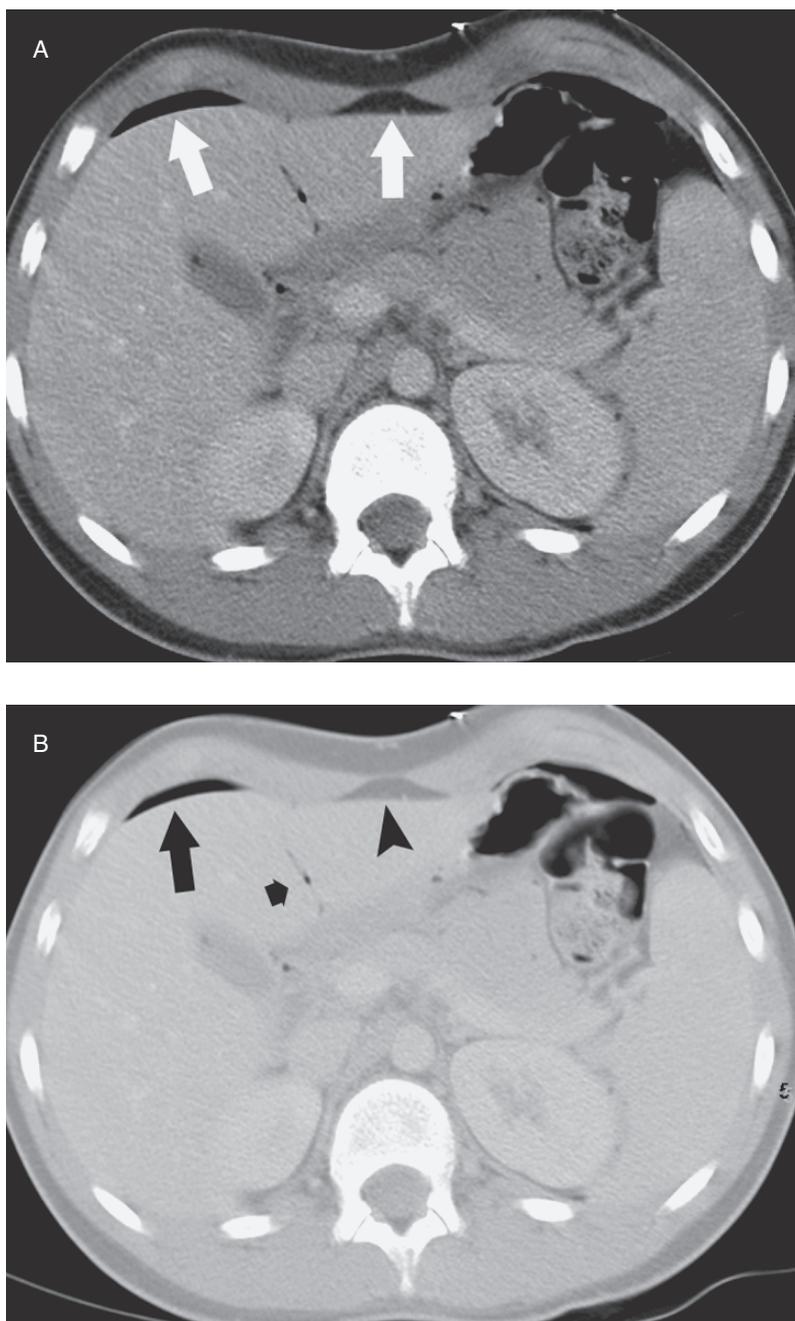


Fig. 24. Pneumoperitoneum. (A) Axial CT image of the upper abdomen viewed in abdominal window demonstrating poor discrimination between free air (left arrow) and fat (right arrow) densities. (B) Same image viewed in lung window shows better discrimination between air (arrow) and fat (arrowhead) density. Small bubble of gas is also noted along falciform ligament (short arrow).

A serious complication of PEG placement is early dislodgement of catheter prior to formation of a mature tract. This is reported to occur in up to 7.8% of patients undergoing the procedure (57). Radiologic management in these patients is limited. If there is a concern for dislodgement of the catheter into the peritoneal cavity, a water-soluble contrast examination performed through the gastrostomy tube may be obtained to confirm the intraperitoneal position. Attempts to blindly replace a completely dislodged catheter, however, are contraindicated owing to the high risk of false passage creation. Under fluoroscopic guidance, the interventional radiologist may on occasion have success in reestablishing continuity between the gastrostomy and the stomach by using catheter-directed guidewire manipulation.

Late dislodgement of gastrostomy tubes poses little risk to the patient with a mature tract. Immediate replacement is not necessary, although urgent replacement is suggested because of spontaneous closure of the gastrostomy sites within approximately 6 hours (58). This can be prevented by instructing the parents or caregivers to immediately replace the gastrostomy tube, or an equally sized Foley catheter, through the gastrostomy site and tape it in place. A new gastrostomy tube can be directly replaced under fluoroscopic guidance when the tract remains open; however, if the gastrostomy tract has stenosed, fluoroscopically directed dilation of the tract may be performed. This allows replacement of a similar size gastrostomy tube.

HEPATOBIILIARY DIAGNOSIS AND INTERVENTION

Hepatobiliary disease in children is typically from a benign cause. Postoperatively, the etiology may be caused by obstruction, bile leak, or portal vein thrombosis. With obstruction, the patients usually present with jaundice, although abdominal pain, fever, vomiting, or abnormal lab values may also be the initial problem (59). Bile duct injury, retained stones, and strictures (particularly with transplantation) (60–62) are the most common causes. Bile leaks after surgery are often accompanied with abdominal pain and fever (Fig. 25). Portal vein thrombosis can occur after abdominal sepsis, such as appendicitis. The presentation is usually occult. Septic postoperative or trauma patients are also at risk for acalculus cholecystitis.

Imaging in these patients should be focused on the specific clinical question. With possible obstruction, imaging should confirm or exclude that assumption, and identify the level of obstruction. If a bile leak is suspected, this needs to be identified, including the site of the extravasation.

Ultrasound is an ideal initial scanning technique for these patients (59). Intrahepatic and extrahepatic biliary duct dilation can be readily identified, and retained stones may be visible. Abnormal fluid collections are often identified. Doppler analysis can evaluate for the presence and direction of portal venous flow. Cholecystitis can be easily identified as an enlarged gallbladder with wall thickening, pericholecystic fluid, and a hyperemic wall. The ability of ultrasound to be performed portably is valuable in patients in the intensive care unit (ICU).

CT can add valuable information in patients with postoperative disease (59). Bile duct dilatation can be identified, and the level of obstruction may be better seen than with ultrasound. Retained bile stones are rarely seen, as they do not usually contain significant calcium. Abdominal fluid collections such as a biloma are more readily identified. Contrast enhancement of the hepatic and portal vessels allows for evaluation of thrombosis, occlusion, or portal hypertension.

Nuclear medicine is not utilized as much as in the past for these patients. Cholecystitis can be diagnosed with ultrasound, biliary obstruction is better evaluated with imaging, and significant bile leaks are identified as fluid collections on imaging.

MRI has an increasing role in the patients (59). Magnetic resonance cholangiopancreatography (MRCP) is a valuable way to visualize the biliary tree in patients with obstruction, and can replace diagnostic endoscopic retrograde cholangio

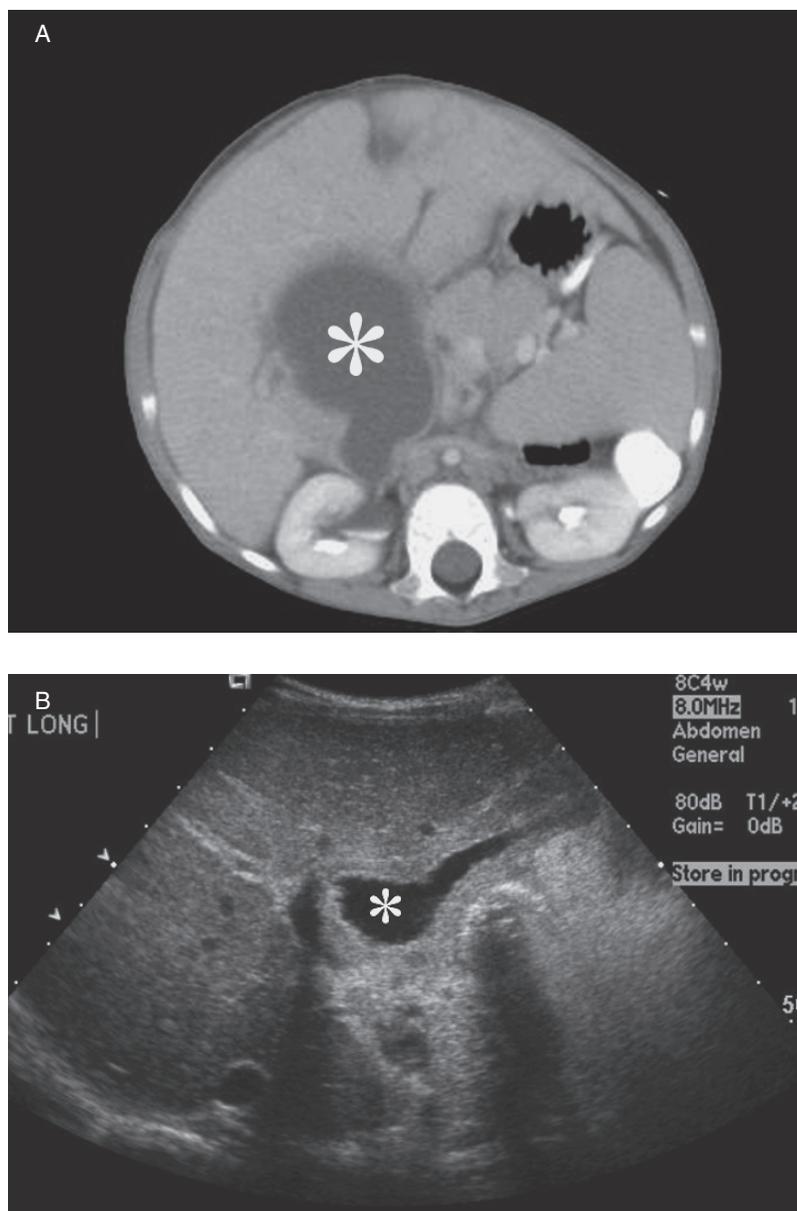


Fig. 25. This patient had fever after cholecystectomy. (A) CT scan showing an abnormal fluid collection in the gallbladder fossa consistent with a biloma (*). (B) The fluid collection (*) on ultrasound which will guide drainage. (C) A drain in the biloma (*). Injection of the biloma shows filling of the intrahepatic bile ducts (arrows).

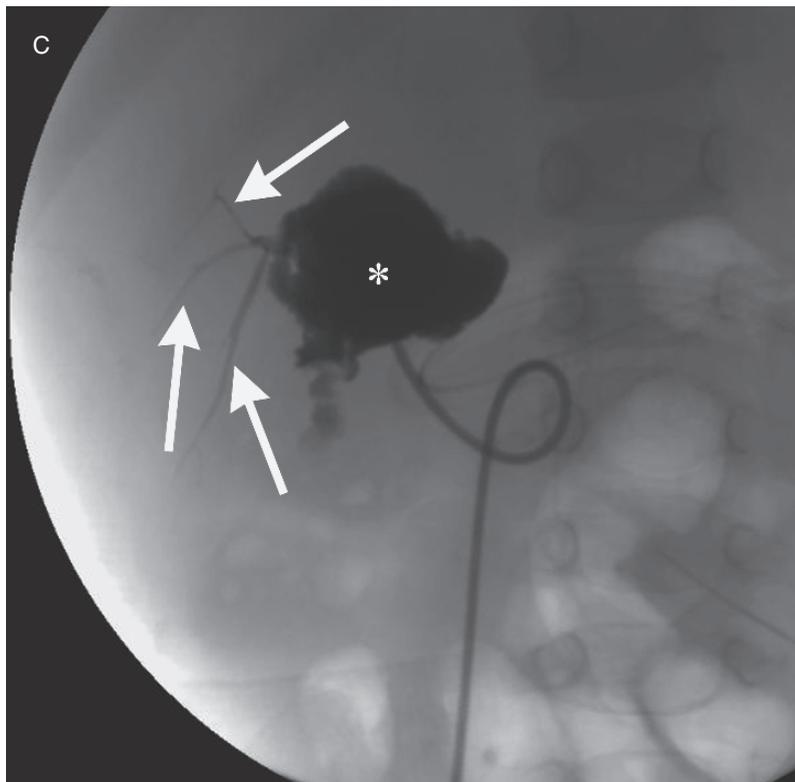


Fig. 25. (Continued).

pancreatography (ERCP) in some instances. Bile duct dilatation and the level of obstruction are readily identified, and retained stones can be seen.

Obstructive Jaundice

In patients with obstructive jaundice, imaging as described previously will identify the level and cause of obstruction. ERCP is usually the initial interventional approach in adults; however, in children this may be more difficult. Relief of the obstruction by external or internal drainage can be accomplished with interventional radiology (IR) techniques (63). The first decision is the need for solitary or multiple drains. If the obstruction is at the level of the common hepatic duct or more distal, a solitary drain will treat both the right and left hepatic lobes. Intrahepatic obstruction may require multiple drains to adequately treat the patient. Both right and left lobe approaches are available, each with different advantages and disadvantages. The right lobe approach drains a larger percentage of the liver, but has an increased risk of transpleural traversal (64). This predisposes the patient to pneumothorax, hemothorax, and biliary-pleural fistula; although this usually can be avoided. A left approach has fewer lung complications, but has a smaller liver volume with which to work (Fig. 26).

These patients all need appropriate antibiotic coverage before the procedure. Ultrasound guidance is the preferred method; although with minimally dilated ducts, fluoroscopically guided placement of a small-gauge needle via the right mid-axillary line is also possible. Typically, a small needle (22 G) is used to enter a peripheral bile

duct. After insertion of an 0.018-inch wire, a stiff introducer system is used to gain access with a 4- to 6-Fr sheath. Through this sheath, diagnostic cholangiography can be performed followed by catheter and guidewire manipulation through the biliary system. In patients who are critically ill, a drain can be left within the liver. Otherwise, attempts are made to pass the area of obstruction, and establish internal/external drainage by placing a drain through the intrahepatic ducts, across the level of obstruction, and into the duodenum.

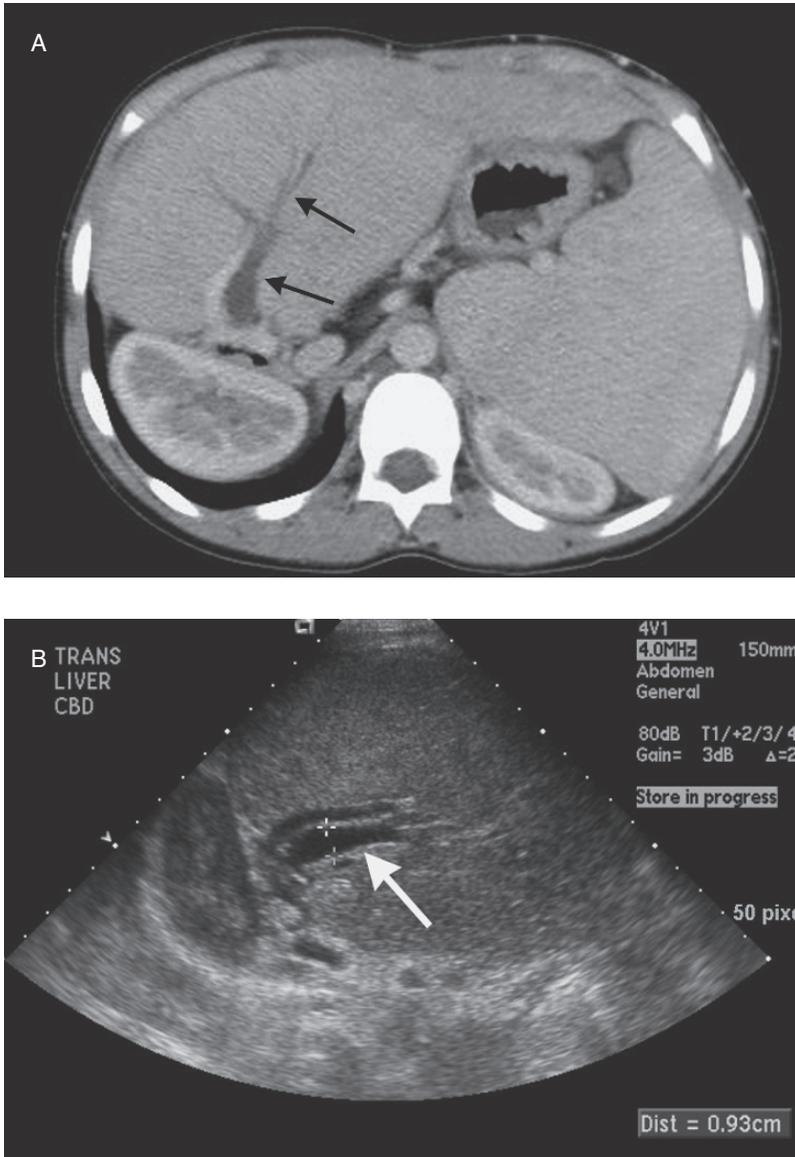


Fig. 26. Biliary stricture. This patient has had a partial hepatic resection for hepatoblastoma. He had a choledochojejunostomy, but presents years later with jaundice. (A) Intrahepatic bile duct enlargement (arrows), confirmed on (B) ultrasound (arrow). (C) Small needle access (arrows) into a peripheral bile duct (arrowhead). Subsequently, a drain was placed. (D) Cholangiogram through the drain, showing obstruction (arrow) at the choledochojejunostomy.

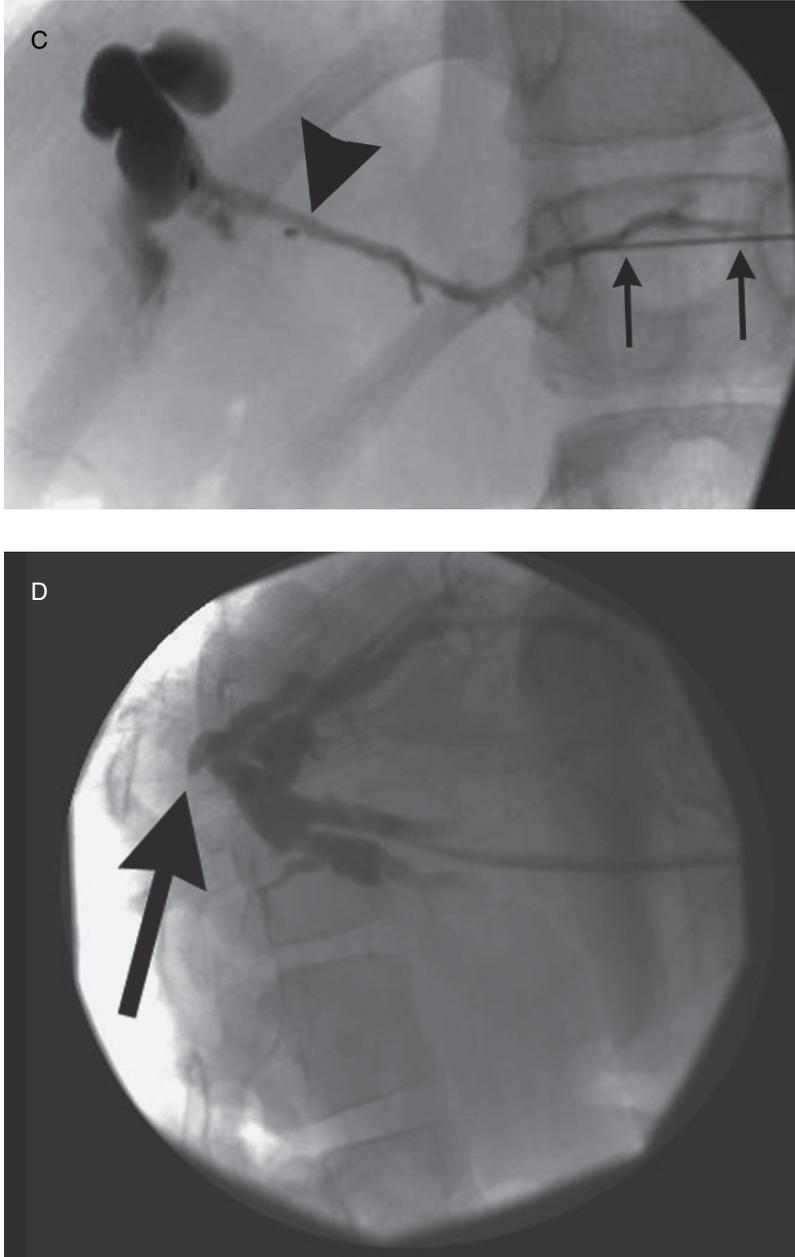


Fig. 26. (Continued).

After crossing the obstruction initially, or at a later date after the patient's sepsis has resolved, the reason for the obstruction can be addressed. Strictures can be dilated with an angioplasty balloon, and retained stones can be removed via IR fluoroscopically guided techniques, or with a combined IR/surgical fluoroscopic/cholangioscopy technique (65,66) (Fig. 27). A drainage tube is left in place for several weeks, and only removed if the patient does well after capping the tube for a week (67). Metallic stents are contraindicated for most benign biliary obstructions.

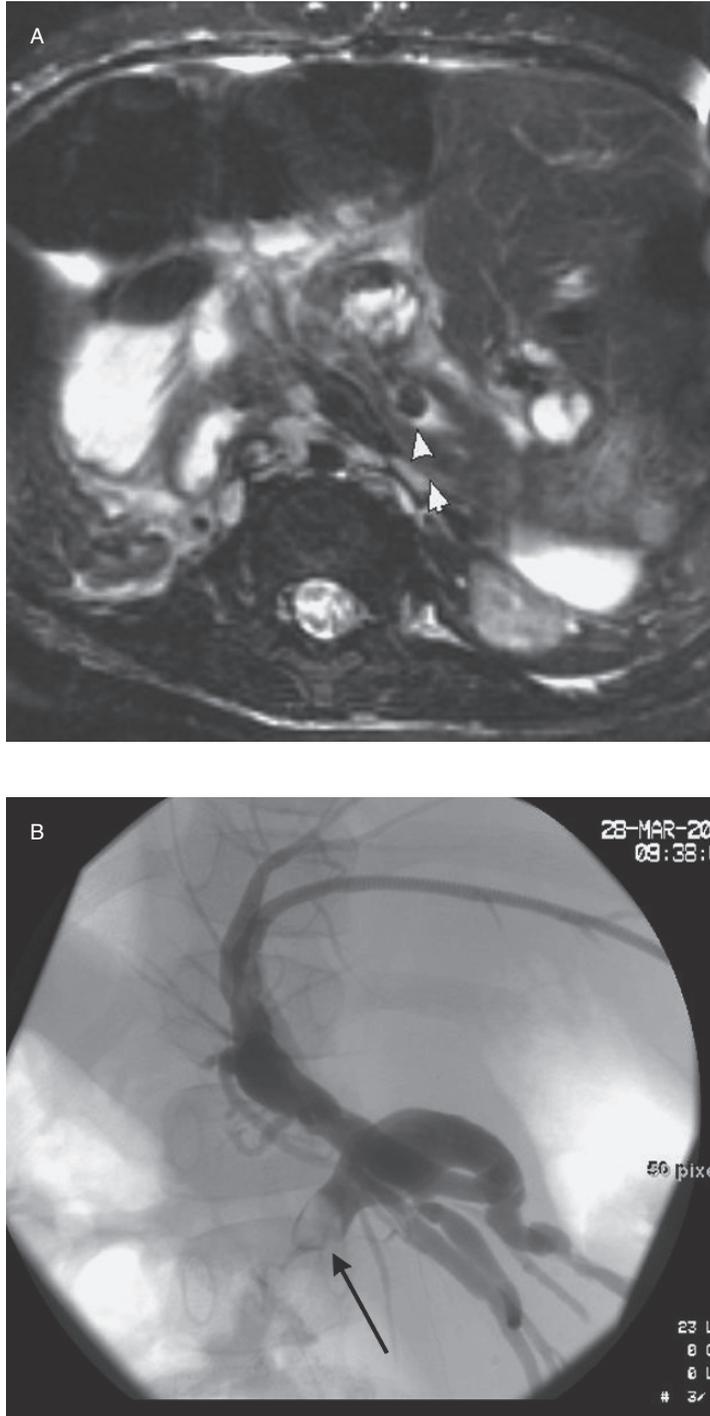


Fig. 27. Retained biliary stone. This patient had jaundice after cholecystectomy. She has situs inversus. **(A)** Retained biliary stone (arrowhead) on the MR. **(B)** Biliary drainage and a cholangiogram with the retained stone (arrow) at the distal common bile duct. **(C)** The obstruction has been crossed with a wire (arrow) for internal drainage. **(D)** At a later date combined fluoroscopy and cholangioscopy were used to remove the stone. The endoscope (arrowheads) was guided with fluoroscopic guidance to the stone.

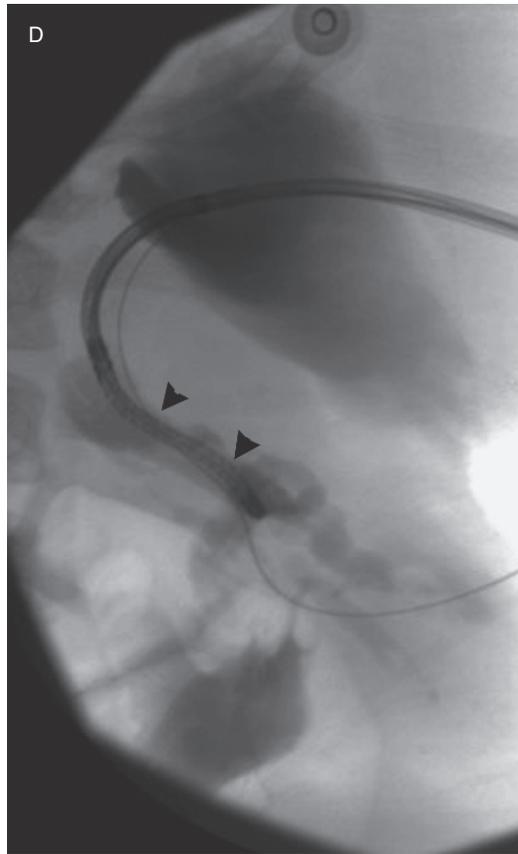
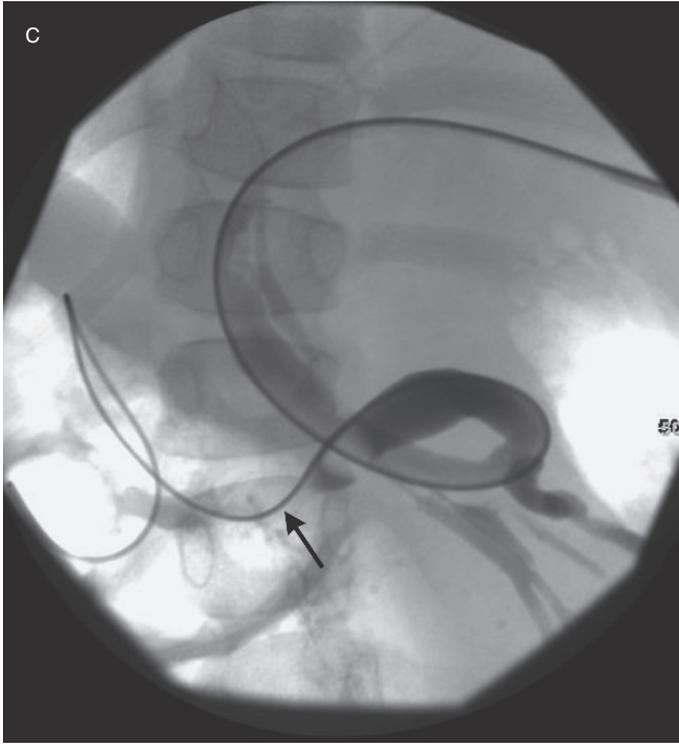


Fig. 27. (Continued).

Biliary Leaks

Fluid collections identified with imaging in symptomatic patients after hepatobiliary surgery are often bilomas (62) (Fig. 25). The initial treatment for these collections is external drainage of the collection. These techniques are described in the section on abdominal abscesses. External drainage is usually the only necessary treatment; however, some collections show continued drainage caused by an active bile leak. When

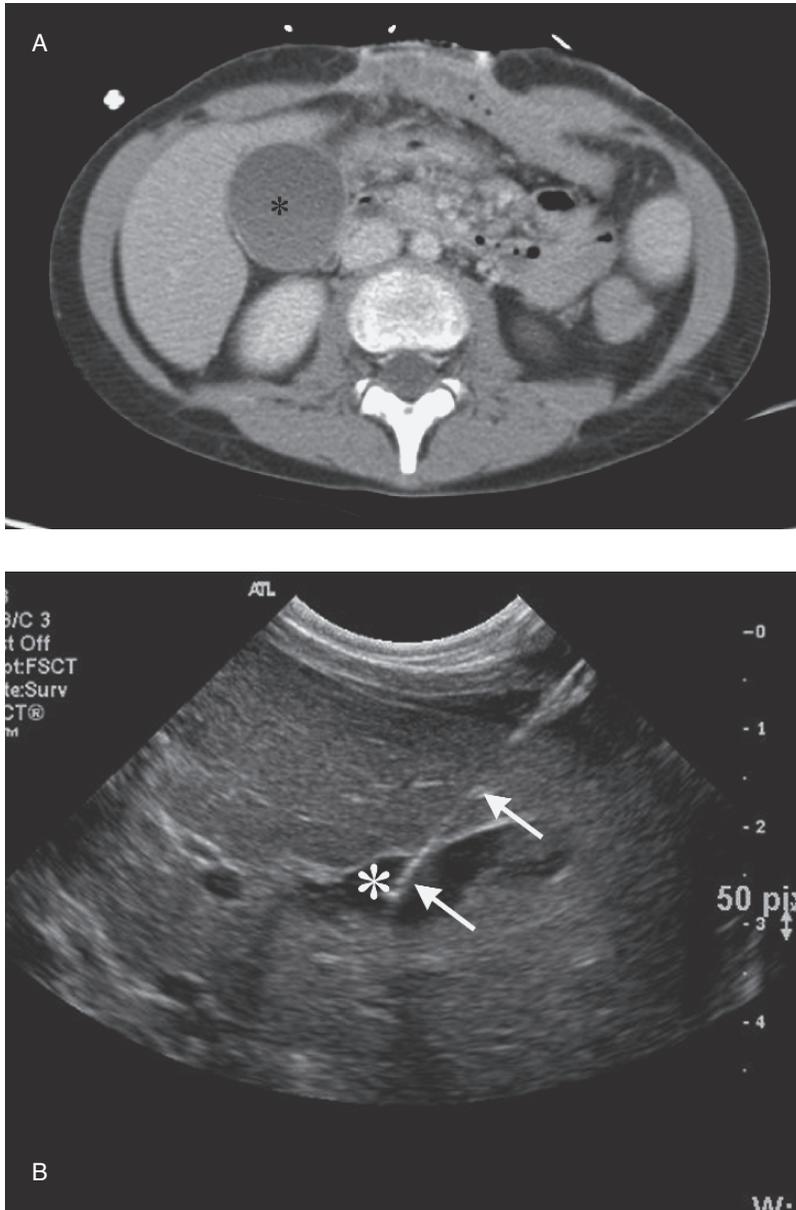


Fig. 28. (A) CT scan from a septic postoperative patient shows a distended gallbladder (*). (B) Ultrasound guided needle (arrows) placement into the gallbladder (*). (C) Injection of the drainage tube confirming placement in the gallbladder (*).

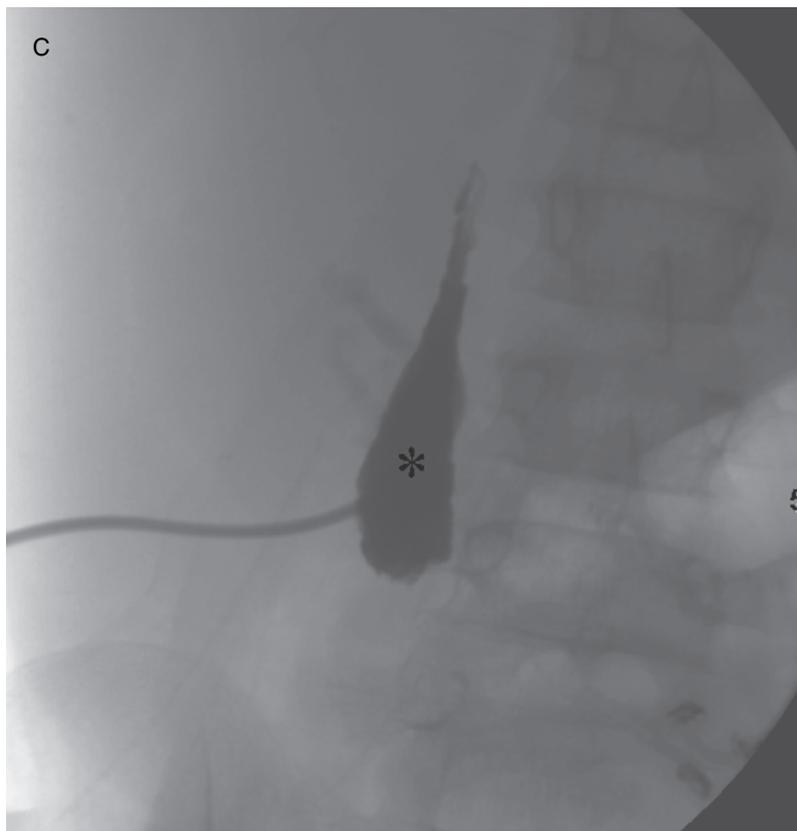


Fig. 28. (Continued).

persistent drainage is present, cholangiography and possible drainage of the intrahepatic bile ducts is indicated. If cholangiography shows the bile leak, then diversion of the bile flow from the leak will allow the leak to close.

Percutaneous Transhepatic Cholecystostomy

Septic patients with acalculus cholecystitis benefit from gallbladder drainage (9). This procedure is often performed portably in the ICU. Ultrasound guidance allows the placement of a drain into the gallbladder, usually with a transhepatic approach to decrease biliary leak after tube removal (Fig. 28). The catheter is typically left in place until the sepsis resolves.

DEEP VEIN THROMBOSIS (DVT) AND PULMONARY EMBOLUS

Venous thrombosis in children is less common than adults, with idiopathic thrombosis seen in 4% of children as opposed to 30% of adults (68). Venous thrombosis in children is most often associated with a central venous catheter (CVC), trauma, malignancy, sepsis, immobility, cardiac disease, and inherited coagulopathy (68–72). DVT is frequently regarded as originating from lower extremities; however, with increasing use of upper extremity CVCs, upper extremity venous thrombosis is an increasing source

of asymptomatic and symptomatic thrombosis (71,72). Pulmonary thromboembolism (PTE) is uncommon in children, yet fatal in 30% of untreated cases (74). In pediatric trauma, older children with high Injury Severity Scores, thoracic injuries, spinal injuries, and CVCs are at greatest risk of venous thromboembolism. Diagnostic imaging modalities for thromboembolism are best used in a targeted fashion: venous sonography for upper and lower extremity thrombosis; multidetector CT angiography for diagnosis of PTE. CT angiography has largely replaced ventilation/perfusion scanning as a screening study for PTE, and with greater than 90% sensitivity (Fig. 29), has largely replaced conventional pulmonary arteriography for definitive diagnosis of PTE

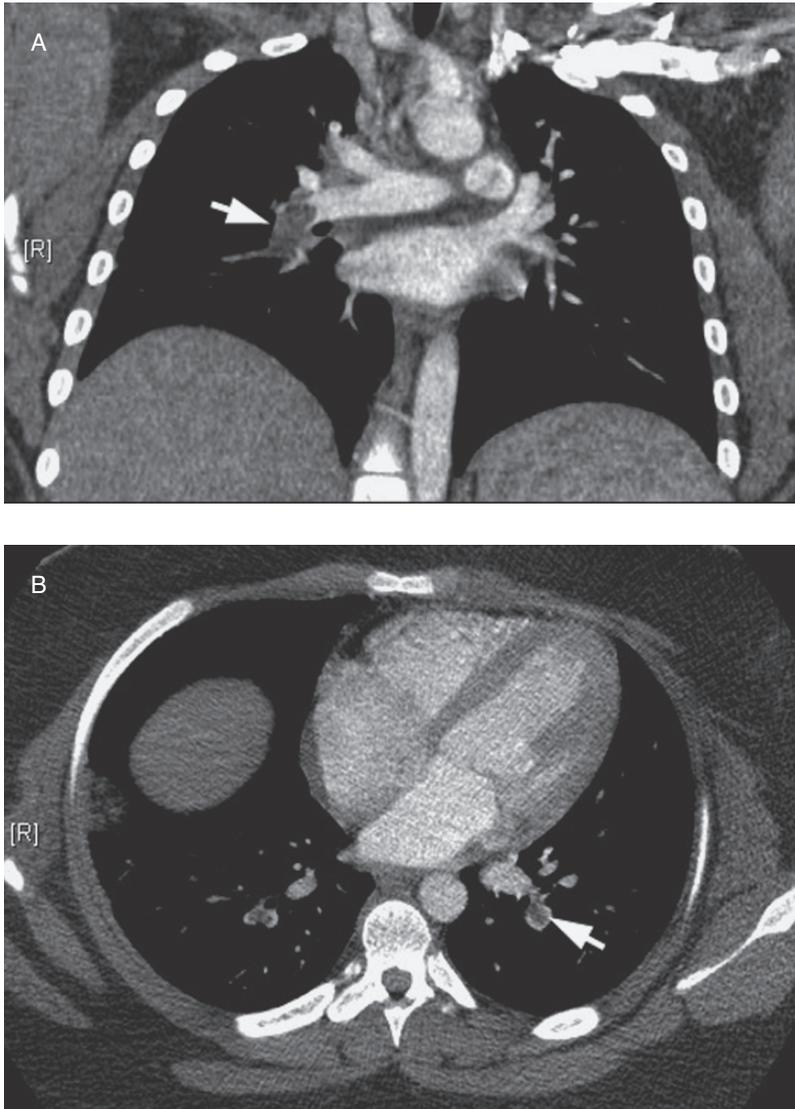


Fig. 29. CT angiogram (CTA) of pulmonary embolus. (A) Coronal reconstruction of CTA demonstrates the large right lower lobe pulmonary artery embolus (arrow) outlined by contrast media. (B) Transaxial CTA image demonstrates bilateral lower lobe pulmonary emboli, with prominent left lower lobe embolus (arrow) clearly outlined with contrast media.

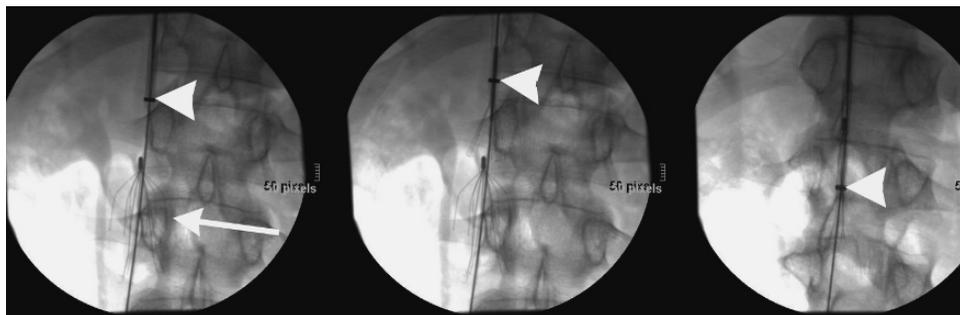


Fig. 30. This patient had extensive DVT, proven PTE by CT, and spinal trauma precluding anticoagulation. A retrievable IFC filter (arrow) had been placed. A sheath (arrowhead) has been advanced through a right internal jugular approach, and is advanced to subsequently remove the filter.

(75–77). Although a superb diagnostic modality when all factors are favorable, the most common causes for an indeterminate scan are motion artifact and poor contrast enhancement (75). Treatment of DVT is primarily done with anticoagulation therapy, to include prophylaxis with low-molecular-weight heparin. IR adjunctive treatment includes the use of intravascular thrombolytic therapy and placement of temporary or permanent vena caval filters (VCFs) (77–83). Thrombolytic therapy with streptokinase, urokinase, and tissue plasminogen activator (tPA, discussed in greater detail in a later section) has been used safely and successfully in children to treat PTE (77–79). With improved device technology and the development of retrievable VCFs, the VCF provides a safe and effective treatment option (Fig. 30) for patients with either DVT or PTE who are not candidates for anticoagulant therapy, patients who have failed anticoagulant therapy, and those with idiopathic infrarenal inferior vena cava (IVC) thrombosis (79–83). The authors are using VCFs more commonly than in prior years, especially the removable VCF, for these indications and in patients who are at significant risk of PTE from DVT.

THORACIC MANAGEMENT ISSUES

Postoperative Pneumothorax

Pneumothorax following primary surgical intervention is most frequently effectively treated with thoracostomy tubes placed at the time of surgery. Occasionally, thoracostomy tubes may fail to drain the pneumothorax because of mechanical occlusion or persistent bronchopleural fistula. In the setting of unresponsive treatment of pneumothorax, the interventional radiologist is able to accurately place a small-bore pigtail (8–14 F) catheter under fluoroscopic guidance. When a persistent air leak is present with a bronchopleural fistula, pleural suction of 30–60 cm H₂O may be required for evacuation of the pleural air to allow the visceral and parietal pleural surfaces to oppose and seal the pleural leak.

Pneumonia

Pneumonia is a potential complication of surgery, particularly with prolonged endotracheal intubation and immobilization. The interpretation of plain radiographs with postoperative atelectasis that persists longer than 2–3 days should include

pneumonia in the differential diagnosis to facilitate timely treatment of an early pneumonia. The lower lobes are the most common location of postoperative pneumonia, although any segment or lobe may become infected. Pneumonia most commonly is treated adequately with antibiotic therapy without complications. The diagnosis of pneumonia is most commonly made with plain radiographs, with CT scanning reserved for pneumonia in which complications are suspected. Bacterial pneumonia is most commonly complicated with parapneumonic effusion, pulmonary necrosis, pulmonary abscess, and empyema. Pulmonary necrosis in children, with or without cavitation, is most often treated without further surgical or radiological intervention. Radiographs 3–12 months after detection of necrotizing pneumonia most often demonstrate complete healing of the necrotic segments without other sequelae. A pulmonary abscess that fails to respond to antibiotic therapy is often amenable to catheter (5–12 F, age and size dependent) drainage, especially when abutting the pleural surface, with little risk of pneumothorax or other complications.

Pleural Effusion and Empyema

Bacterial pneumonia is common in children, with most cases being caused by *Streptococcus pneumoniae* (84–87). Multiple reports have cited the incidence of associated pleural effusion to be 36–57%, with 15–20% progressing to empyema, defined as pus within the pleural space (88–92). Empyema is classically divided into three separate stages (85,87–90,93,94):

1. Early exudative: Lasts from 24–48 hours, and fluid forms because of pleural inflammation.
2. Fibrinopurulent: Lasts from 7–10 days, and is characterized by thickened exudates, fibrin deposition, white cell infiltration, and formation of loculations.
3. Organized: Starts 2–4 weeks from effusion onset. There is fibroblast formation and creation of a thick peel.

Grading of pleural disease based on laboratory criteria have also been described, with pH, glucose levels, lactic dehydrogenase (LDH), Gram stain, and cultures all being performed (90,91). However, this lab evaluation does not define how the patient should be treated, with the exception of antibiotic coverage for the appropriate pathogen (95). Ultrasound has also been used to grade empyemas (Fig. 1A and 1B), as Stage I (anechoic fluid), Stage II (loculations), and Stage III (solid peel) (96). This information is prognostic, concerning the need for additional treatment such as fibrinolytics or surgery (91,92,96).

It is well established that appropriate treatment of empyema will help control sepsis, restore pulmonary function, and prevent lung entrapment from the fibrous peel (88,91). In prior years, empyemas were treated initially with large-bore chest tubes and/or with open thoracotomy with decortication. Two more recent techniques for dealing with empyema were then described: video-assisted thoracoscopic surgery (VATS), and small tube placement with fibrinolytic therapy. These techniques are now considered the most appropriate treatments for patients with empyema, with thoracotomy reserved for patients with a large peel. However, which is best is quite controversial (85,88, 89,97,99). At institutions where IR performs image-guided small tube placement and fibrinolytic therapy, success rates are 77–98% (90,91,93,95,100,101). It is obvious that these small tubes are easier on patients. VATS is also usually successful, and might have some potential advantages (87,99,102–105). Some authors think that a patient's

symptoms might improve more quickly, and the hospital length of stay may be less (89,94). However, with IR drainage, the temporal improvement in symptoms is similar to VATS, the operating room (OR) risks are obviated, and the patients avoid having a large chest tube.

A simple and effective treatment protocol has been developed for treatment of empyemas at the authors' institution (101). All patients with empyemas considered for percutaneous treatment are evaluated by IR following chest CT. Chest CT helps define parenchymal disease, and areas of necrosis are readily identified. This gives prognostic information about the duration of therapy, and possible need for future interventions (Fig. 31). Prior to insertion of the chest tube in IR, the empyema is

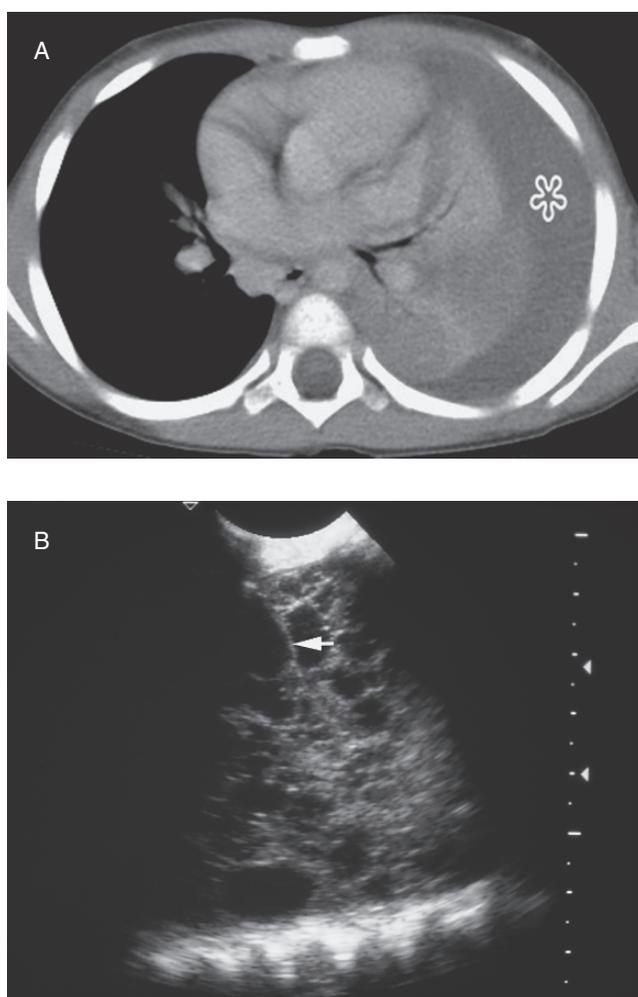


Fig. 31. Percutaneous treatment of organized empyema. **(A)** Chest CT demonstrates left lower lobe pneumonia with large left pleural empyema (asterisk). **(B)** Sonogram of the left pleural empyema demonstrates grade 2 organization with large amount of organization with fibrin strands (arrow) separating purulent locules. **(C)** Chest radiograph following placement of 12-F pigtail catheter for fibrinolytic therapy and drainage. **(D)** Chest CT following fibrinolysis demonstrating resolution of the empyema with a trace amount of pleural thickening following therapy (arrow).

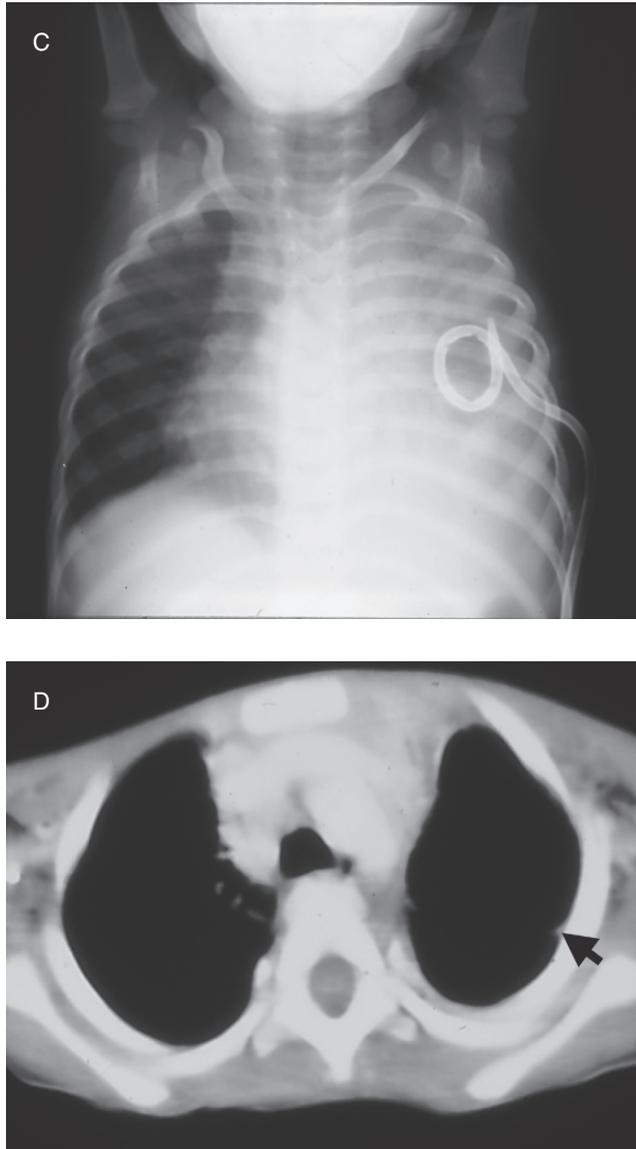


Fig. 31. (Continued).

assessed with sonography. Patients with a solid peel as identified by ultrasound were referred for surgery. All other patients had small-bore chest tubes (10–14 F) placed, with fibrinolytics given if the fluid was echogenic or had loculations, or if the patient had suboptimal response defined as residual fluid, or no symptomatic improvement (Fig. 31). The tube is then attached to a water-seal chest tube system and is placed to 20-cm water suction. The tPA dose is 2 mg mixed in 20 mL of saline. The tPA is injected into the tube, the tube is clamped for 1 hour, and then the tube is returned to suction. This is repeated every 12 hours until fluid output decreases to less than 25 mL in a 12-hour period. Chest tube output, daily chest radiographs, and patient symptoms are monitored throughout the protocol. At 48 hours, if the patient has not

improved, and residual fluid is suspected because the tube output is less than expected or the chest radiograph got worse, repeat chest CT is performed. If significant pleural disease is present, then tube drainage with fibrinolytics is considered a failure, and the patient is referred for surgical intervention. Otherwise, the tube is removed when output falls below 25 mL in a 12-hour period, and one of the two following criteria are met: (1) The patient is afebrile and off supplemental oxygen; or (2) Chest radiograph has demonstrated no significant pleural disease.

This protocol of drainage and fibrinolytic therapy was effective in 92% of patients, with the patients' mean length of hospitalization at 8.4 days and average number of tPA doses at 5.9. Most initial failures in the evolution of this protocol were patients where tPA was either started late, or stopped after a few doses. Since publication, expanded use of this protocol has demonstrated efficacy in 98% of patients.

DISPLACED VENOUS CATHETER

CVCs may be subject to malposition or fracture, resulting in the need for either repositioning, removal, or retrieval of migrated catheter fragments. Subclavian CVCs may infrequently penetrate either the subclavian vein or superior vena cava (SVC) with resultant pleural effusion from the infusate. This malpositioning is treated with pigtail catheter thoracentesis as well as removal and replacement of the CVC. Femoral CVCs will infrequently be placed into lumbar veins rather than the IVC, requiring replacement over a guidewire with fluoroscopic guidance, or complete replacement. The most common site of CVC malposition is either an arm percutaneously inserted central catheter (PICC) or subclavian CVC in which the tip has flipped into the ipsilateral internal jugular vein. Flexible PICC lines may be repositioned using a simple technique of rapid saline injection, coupled with deep inspiration, both working in concert to relocate the catheter tip in the SVC. When simple maneuvers such as these fail, the tip may be repositioned with fluoroscopic guidance and guidewire manipulation or CVC tip redirection using an angiographic catheter placed through the brachial/basilic or femoral venous routes. In the rare instance of CVC fracture and migration, nitinol retrieval snare techniques may be used, most frequently from a transfemoral access route, safely capturing and removing the CVC fragment.

VENOUS AND ARTERIAL THROMBOSIS

Imaging of Suspected Vascular Thrombosis

Vascular thrombosis is much less common in children than in adults. Thrombosis is related to Virchow's Triad, where slow blood flow, an abnormal endothelium, and hypercoagulable states are predisposing factors (108). Although children may develop lower extremity DVTs or effort thrombosis (Thoracic outlet syndrome, Paget-Schroetter) as in adults, a larger percent have thrombosis related to a procedure such as central venous or arterial catheterization (109–114). Vascular catheters injure the endothelium, and alter the blood flow in the vessel. In addition, many patients who need vascular catheters have inflammatory processes that can cause a hypercoagulable state (115,116). Most of these thromboses lead to occult occlusions. No acute symptoms are present, and obstruction is only identified later with inability to recannulate the vessel. However, some of these patients will have extremity swelling or limb ischemia

and will present acutely with their thrombosis. Dialysis access is another condition where vascular thrombosis is important. Thrombosis of an arteriovenous fistula or graft precludes hemodialysis until remedied or a catheter is placed (117,118).

Ultrasound is the best initial imaging technique in patients with suspected thrombosis (119,120). The vessel is often filled with hyperechoic clot, and has no Doppler flow (Fig. 32). Secondary findings may be present such as inability to compress the vessel, and the lack of Doppler findings such as respiratory waveform variation and augmentation. Some vascular territories are outside of the typical ultrasound windows. The subclavian veins are difficult to visualize with ultrasound, and collateral veins in the area may mimic a patent subclavian vein. The IVC can be obscured with bowel gas. Although less common, arterial thromboses in similar positions may be obscured.

When ultrasound evaluation is inconclusive, or when the area of interest cannot be evaluated, angiography is indicated, and can identify thrombosis or occlusion. With hemodialysis access, angiography will identify thrombosis, and the underlying vascular problem (118).

MRI with angiographic techniques can evaluate for vascular patency (120,121). This technique is valuable in patients with suspicion of prior venous occlusion (usually owing to multiple prior lines). MRI is most useful in patients for planning for additional IV access, as ultrasound and angiography are indicated for acute thrombosis (Fig. 33).

CT angiography techniques are a newer way to evaluate for thrombosis. Multidetector imaging allows for images to be obtained with a more uniform contrast bolus. Volumetric acquisition allows for better image reconstruction and processing (120).

When vascular thrombosis is diagnosed, it is important to determine if the occlusion is acute or chronic. Acute thrombosis can be treated with thrombolysis and treatment of the underlying problem. Chronic occlusion will not respond to these treatments. If the thrombosis is acute, the benefit of the procedure must be weighed against the risks. Contraindications to thrombolysis include brain tumors, recent surgery, or known stroke (122). Young children undergoing thrombolysis must be admitted to the pediatric ICU. If they cannot hold still during the treatments, they may need paralysis and intubation. For simple asymptomatic venous thrombosis related to a catheter, this is not typically worth the risks. However, if the limb is swollen, access is critical, or for venous or arterial thrombosis with limb ischemia, the other problems can be managed. The next decision is for systemic or local thrombolytic therapy. If the throm-

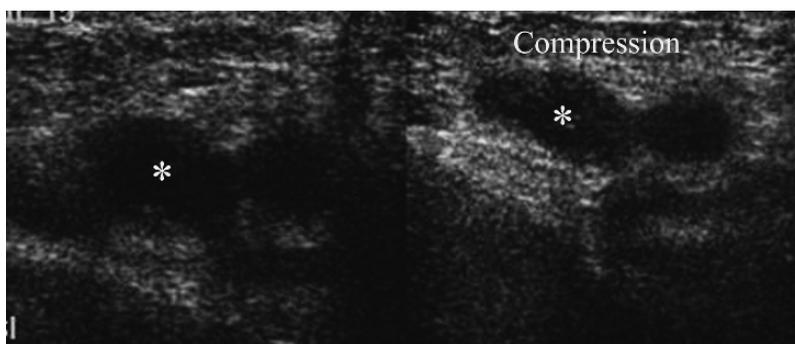


Fig. 32. Ultrasound without (left) and with (right) compression demonstrates a noncompressible right common femoral vein (*) consistent with DVT.

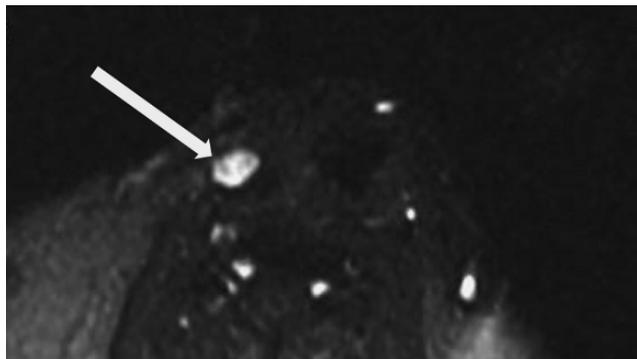


Fig. 33. This source image from an MR angiogram shows a patent right internal jugular vein (arrow). The left is not visualized.

basis is diffuse, systemic therapy is indicated (123,124) (Fig. 34). For a focal clot, catheter-directed thrombolysis is best, and an infusion catheter is placed across the thrombosis (Figs. 35 and 36). Dosing of the current preferred thrombolytic, recombinant tPA is not well established. Pediatric doses have ranged from 0.02 mg/kg/hour to 0.5 mg/kg/hour, but are not standardized for systemic or catheter-directed therapy (114). Adult dosing may be either weight-based or set for arterial (0.5 mg/hour) or venous (1.0 mg/hour) administration (125). The authors use a weight-based dose of 0.05–0.1 mg/kg/hour for catheter-directed therapy with maximum doses of 0.5 mg/hour for arterial and 1.0 mg/hour for venous administration. The infusion is continued for 6–12 hours. Follow-up angiography is then performed, with continuation of the thrombolysis, treatment of underlying vascular pathology, or discontinuation of treatment done depending on the results. All patients are placed on one-half-dose heparin during thrombolysis to prevent thrombosis around the catheters. Residual stenoses are often identified from a prior central venous line or effort vein thrombosis, and these can be treated with angioplasty. Stenting of vascular stenoses is controversial in children. Stents will not grow with a pediatric patient, and intimal hypertrophy will limit the long-term patency in these vessels.

Thrombosed dialysis grafts or fistulas are more of a problem for adult IR physicians, although some pediatric patients have these devices. Thrombolysis, graft studies, and angioplasty can be performed as in adult patients. The dose of tPA to clear a thrombosed graft is 2–4 mg, which is an acceptable dose in most children old enough for a dialysis shunt. National Kidney Foundation guidelines should be followed (118). Portal venous thrombosis may occur in children with abdominal sepsis or dehydration. Usually occult initially, these patients may present with portal hypertension and “cavernous transformation” of the portal vein; although these patients may present with nonspecific abdominal pain (Fig. 37). This diagnosis can be confirmed with ultrasound, CT, or MRI. The diagnosis of portal hypertension may be difficult if varices or splenomegaly are absent. Techniques such as wedged hepatic venous pressures will not be accurate because of the discontinuity of the portal triad with the mesenteric vascularity. Transhepatic portal systemic shunts (TIPSS) are typically difficult because of the occluded portal vein, and are a temporary solution in a child with a long-term problem. Surgical shunts are possible, and ultrasound and CT imaging are used to assess patency. IR techniques can be used to identify and treat thrombosis or stenosis

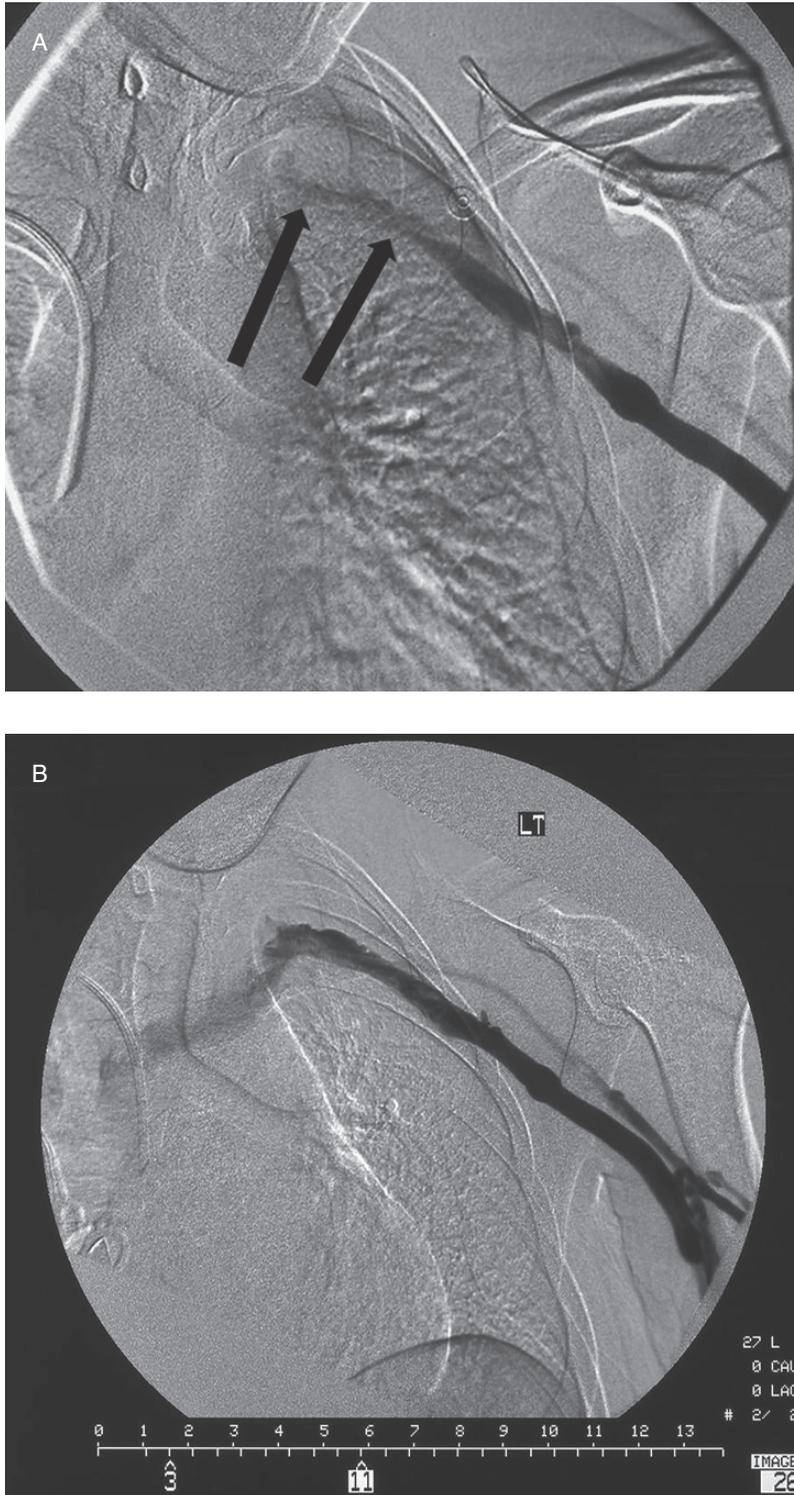


Fig. 34. SVC syndrome. This patient with bilateral central lines had acute head swelling and headache consistent with SVC syndrome. (A) Occlusion at the level of the subclavian vein (arrows). (B) Restoration of flow after system thrombolytic infusion into the central lines.

of these shunts (126–128). Arterial or venous thrombosis in transplant patients can be treated similarly (129,130).

IVC filters decrease the risk of pulmonary embolism from lower extremity and pelvic DVTs (131,132). Indications include a patient at risk for PTE who cannot

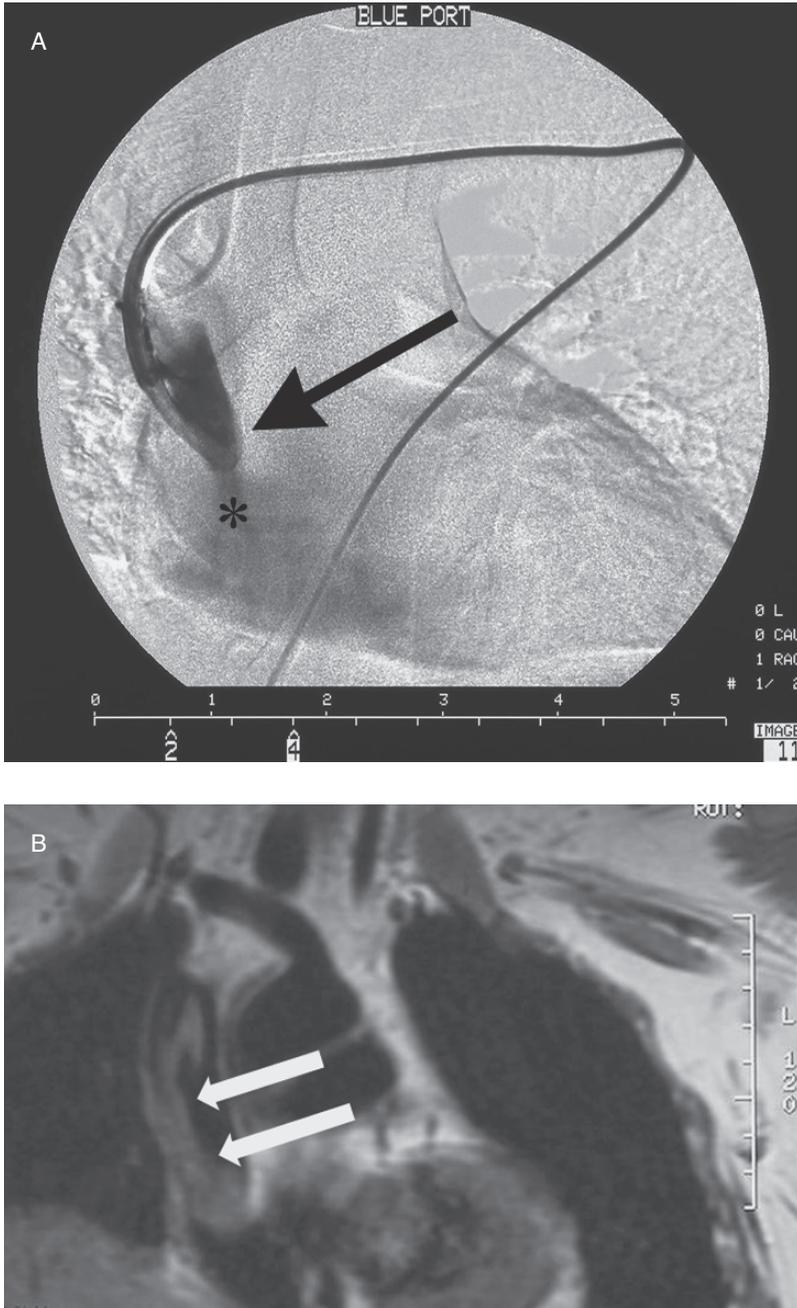


Fig. 35. SVC thrombosis. (A) Central line injection shows a narrowing (arrow) in the SVC with a jet (*) into the right atrium. (B) MR demonstrates a filling defect (arrows) along the wall, representing clot. (C) Image after thrombolytic infusion shows a patent SVC without clot (*).

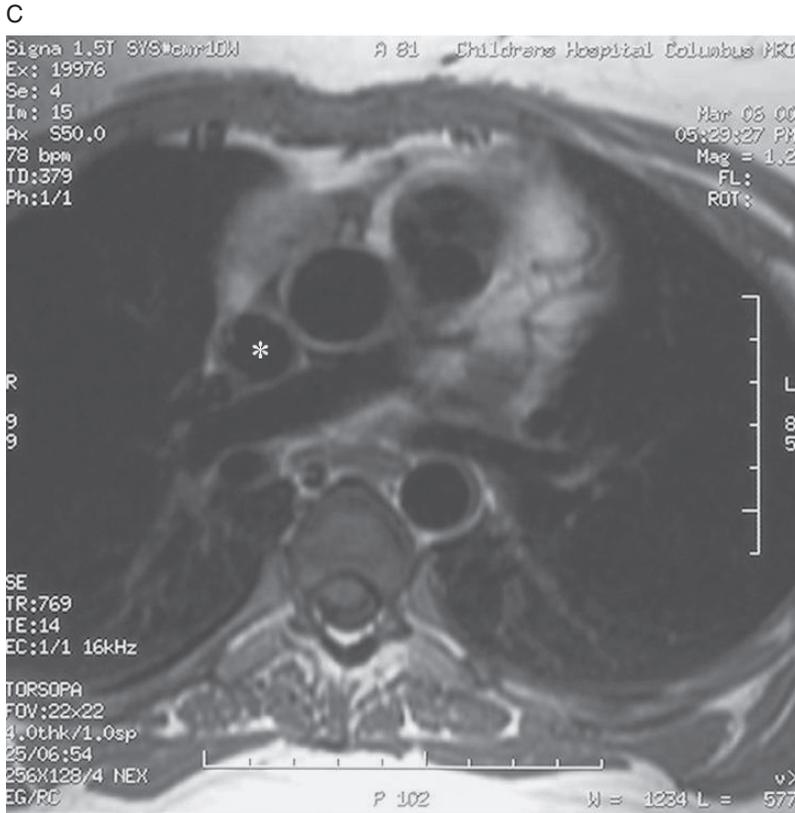


Fig. 35. (Continued).

be anticoagulated, or a patient with recurrent PTE on appropriate anticoagulation. Both permanent and removable filters are available, and the removable filters can be retrieved when the patient can be anticoagulated or when there is no more risk of PTE (Fig. 30).

REOPERATIVE TREATMENT OF VASCULAR ANOMALIES

In the reoperative care of patients with vascular anomalies, a clear understanding of the treated tumor or malformation is essential for both the surgeon and treating interventional radiologist. Vascular anomalies are categorized into two main groups: vascular tumors (infantile hemangioma) and vascular malformations. Vascular malformations are commonly divided into two major groups: high-flow and low-flow lesions (133). Arteriovenous malformation is a high-vascular-flow lesion; with capillary malformations, venous malformations (VM), and lymphatic malformations (LM) constituting slow-flow lesions. The most common reoperative care scenarios in this spectrum are residual VM and LM following primary surgical resection. These two lesions will provide the focus for the following discussion.

VMs are most often a network of cavernous venous channels with poor intercommunication and obstructed venous drainage mechanisms. Reoperative VMs are found in two primary settings: residual foci of disease following attempts at primary surgical resection, and venous components of a mixed venolymphatic malformation that enlarge

years after primary resection of the LM in childhood. Radiological diagnosis is best performed with contrast-enhanced MRI, demonstrating the full extent of the lesion in a global perspective; sonography is best to plane procedural mapping and clarify discrepant cases (i.e., differential diagnosis between LM and VM).

Given the poorly demarcated and often infiltrative nature of the VM, and lack of clear feeding arterial and venous structures in VMs, IR techniques are playing an increasing role in the treatment of VMs. IR techniques provide the surgeon and patient minimally invasive technique options that are highly effective, safe, and avoid major blood loss and cosmetic deformity. VMs are effectively treated with a combination of embolization and sclerotherapy for endolesional therapy. Coil embolization is used in VMs with significant outflow channels in order to provide lesional venous stasis, in which sclerosant agents will work to produce endothelial cell death and permanent fibrosis of the vascular spaces. In lesions with a favorable slow-flow state, a variety of sclerosant agents have proven effective in the majority of VMs. The two most commonly used agents are sodium tetradecyl sulfate (STS) and ethanol (133,134).

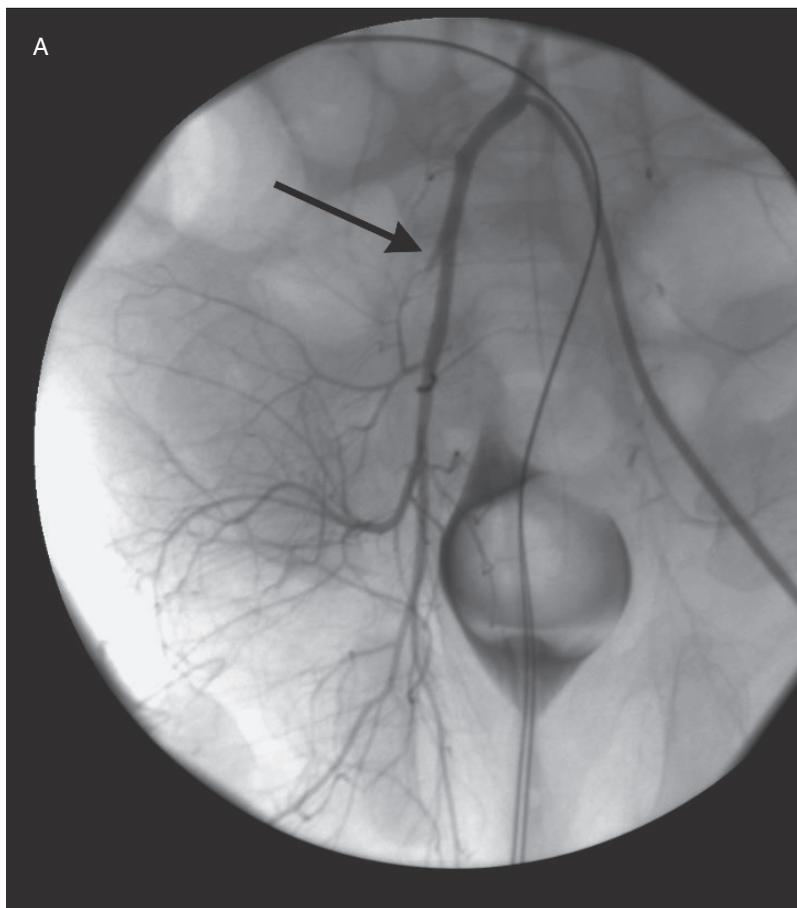


Fig. 36. Neonatal arterial thrombosis. This patient was a premature infant who developed right lower extremity ischemia after right femoral central line insertion. **(A)** Abrupt occlusion (arrow) of the right common iliac artery. After receiving catheter directed thrombolytics, the follow-up **(B)** demonstrates resolved thrombus in the superficial femoral artery (arrows) and the trifurcation (arrowhead).

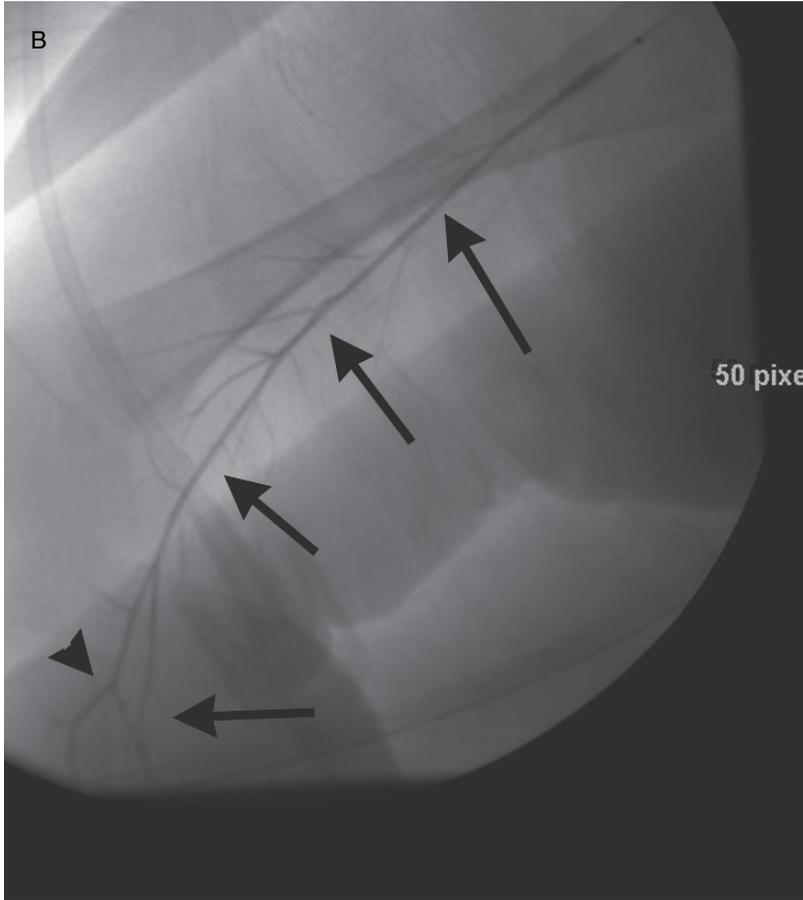


Fig. 36. (Continued).

Other sclerosants include sodium morrhuate, ethanolamine, and polydocanol. Ethanol is a superb sclerosant agent; however, ethanol carries a lower maximum dose allowance for each procedure and the risk of death from acute cardiopulmonary collapse if ethanol is injected into the systemic venous supply at a significant rate and/or dose. STS is most effective in the 3% solution (134) and as an agitated foam solution rather than as the simple liquid form. Sclerosant agents are able to be injected via small-gauge needle systems (22–23 G) (Fig. 38) or via small catheters following coil embolization of outflow channels. Preliminary results suggest efficacy (pain reduction 100%; reduction of swelling 63%) of intralesional laser therapy of symptomatic VMs when sclerosant therapy is not effective or desired (135).

LMs present at all ages from fetus through adulthood. The typical LM is often a combination of large macrocysts with small foci of accompanying smaller cysts (microcystic disease). Less commonly, LM presents solely as a small number of large macrocysts. LM is most frequently diagnosed in the head and neck, but can present in numerous locations to include the orbit, mediastinum, retroperitoneum, abdomen, extremities, scrotum, and penis. A LM that is diagnosed at birth usually presents as a soft, spongy, nontender mass. In older children and adults, a LM may present with rapid development of a soft or firm nontender mass, this occurring as a result of

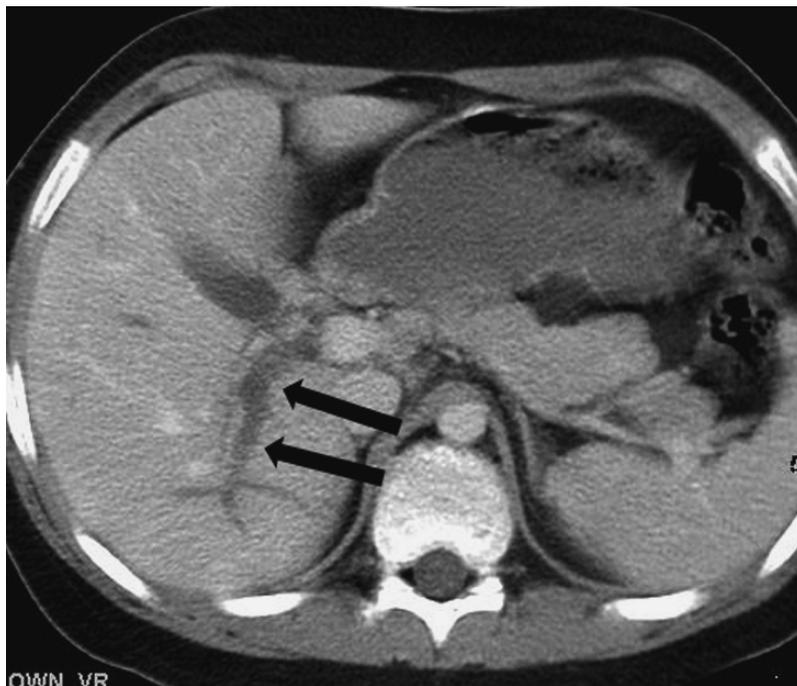


Fig. 37. PTE. This CT scan in a patient with diffuse abdominal pain after appendectomy shows a nonenhancing right portal vein (arrows) consistent with thrombosis.

hemorrhage into previously undiagnosed LM locules. Less frequently, LM presents with infection of the mass or pain.

Diagnostic imaging is best performed with a combination of MRI and sonography. MRI is the imaging modality of choice for global assessment of the extent of a LM, including involvement of airway and osseous structures. Sonography may be the sole diagnostic imaging modality if the lesion is well localized in a superficial location. Sonography during a preoperative consultation facilitates preprocedural mapping and assessment of adjacent structures that may impact procedural efficacy.

Traditionally, LM is treated with surgical resection, followed by interventional radiological sclerotherapy for recurrent or unresectable lesions. Surgical resection may be difficult, and when foci of microcystic disease are left in the operative bed, recurrence can be expected. Fluid collections that occur following resection of LMs are most often postoperative lymphoceles or expansion of residual microcystic disease in the operative field. In the authors' experience, simple drainage of either lymphoceles or residual LM is not effective for long-term resolution. In the authors' experience, lymphoceles are effectively treated with catheter-based (dual-drug regimen as described in the following paragraph) sclerotherapy, with a high technical success rate, less than 1% recurrence, and a less invasive alternative to further surgical therapy.

LMs that recur following attempts at surgical resection occur both as macrocystic (cyst locules greater than 1 cm) and microcystic disease. Macrocystic disease may be treated over a 1-year time period with a series of direct-injection sclerosant techniques with agents such as liquid ethanol, ethanol gel, OK-432, doxycycline, and bleomycin, all long-term efficacy (80–90%) in cyst size reduction (133). These direct-injection

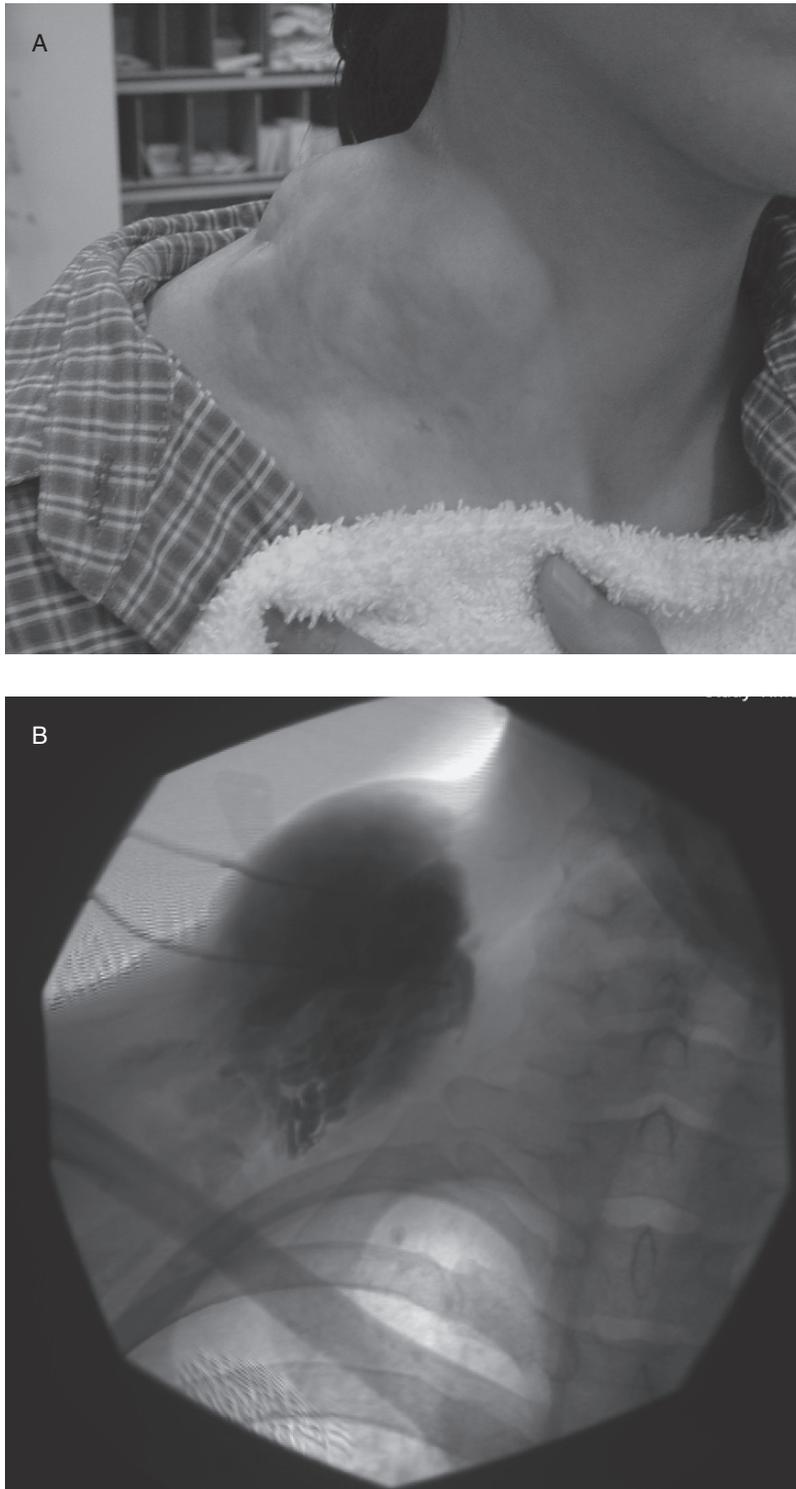


Fig. 38. Cervicothoracic venous malformation with percutaneous therapy. (A) Clinical photograph demonstrating an engorged and painful VM in the right neck and thoracic inlet following prior surgery. (B) Contrast venogram prior to treatment demonstrates the large volume of cavernous VM at the base of the neck. (C) Clinical photograph following one session of treatment demonstrates near complete resolution of the mass effect (and complete resolution of pain).



Fig. 38. (Continued).

methods result in slow reduction of cyst size, frequently require multiple treatments, and have variable risks of extrusion with skin necrosis and peripheral nerve injury. A new technique for treatment of macrocystic LM has been developed at Columbus Children's Hospital, which provides complete ablation of treated macrocysts within 3 days of treatment (136). Prior to treatment, cytology specimens are collected to confirm the diagnosis of LM. Sclerotherapy of macrocystic LM is performed with indwelling pigtail catheter placement (5–8 F) and time-limited dual-drug (SDS followed by ethanol) sclerosant contact followed by suction drainage. Following aspiration of the ethanol, the catheter is connected to a Jackson-Pratt suction bulb system and maintained for 3 days (Fig. 39).

Ultrasound examination performed 1 month following a single session of sclerotherapy demonstrates complete ablation of the treated cysts in 98% of cases (136). Macrocystic LMs have been successfully treated with the previously described protocol, in the neck, face, orbit, chest, abdomen (Fig. 40), mesentery, and extremities without complications. In the treatment of large cysts, there is a 2% incidence of repeat hemorrhage in cysts following sclerotherapy. Following repeat hemorrhage, a second treatment has been effective for ablation in 100% of cases (136). In our patients, no cases of neuropathy, unexpected skin necrosis, or myoglobinuria have been encountered with sclerotherapy. In LM that involves the cutaneous epidermis, sclerotherapy targets the LM and involved skin to provide a clean tissue bed for successful skin grafting.

In the natural history of LM, microscopic cysts that are not detectable by sonography or the naked eye at surgery mature to a point where they are larger than 1 mm and detectable with surveillance sonography. In prior years, minimally invasive therapy for LM was limited to macrocystic disease. Foci of residual or maturing microcystic disease can now be successfully treated with targeted IR therapy, avoiding complications of infection, sepsis, and hemorrhage. A new microcystic sclerotherapy technique developed at Columbus Children's Hospital provides precisely targeted high-concentration doxycycline that allows 50 or more cysts to be treated in a single setting (137). In this microcystic treatment technique, cysts as small as 2 mm are drained

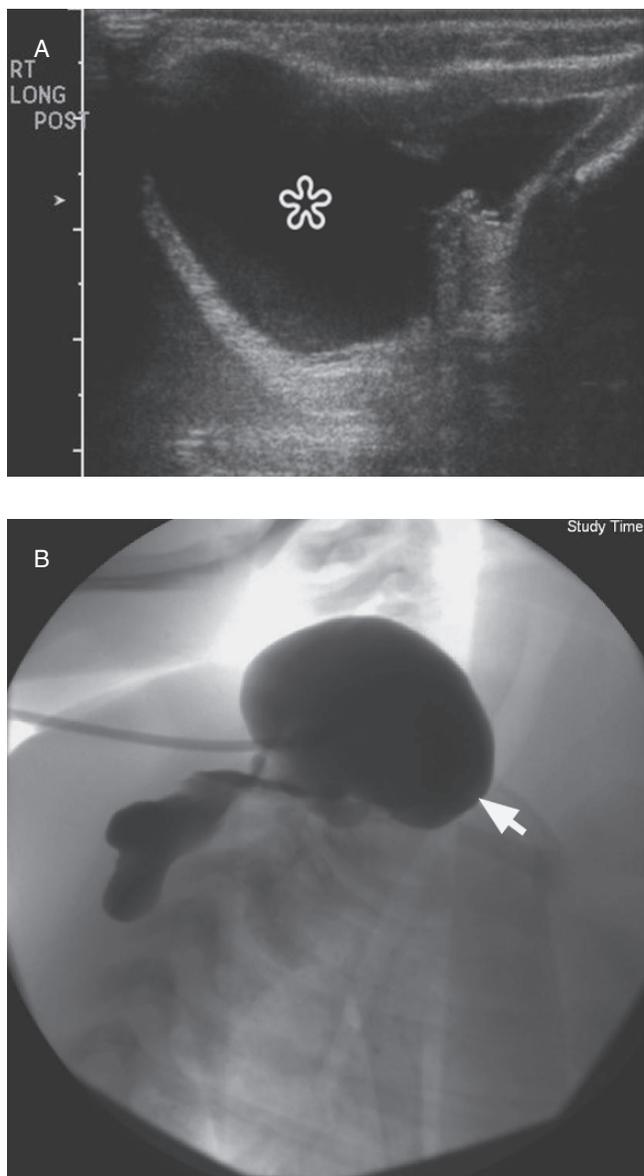


Fig. 39. Macrocytic LM treated with dual drug time-limited ablation therapy. **(A)** Neck and chest sonogram demonstrate the macrocytic LM (asterisk). **(B)** Contrast angiogram during treatment demonstrates the vast extent of the macrocytic LM (arrow) in the neck and shoulder. **(C)** Fluoroscopic image during treatment demonstrates complete evacuation of intracystic contrast prior to dual-drug ablation therapy. Clinically, there was resolution of the mass 3 days following treatment. **(D)** Neck and shoulder sonogram 6 months following treatment demonstrates complete resolution of the LM with no evidence of residual or recurrent disease. Normal neck muscle planes are present (arrow) at the site of prior LM, with no evidence of scarring.

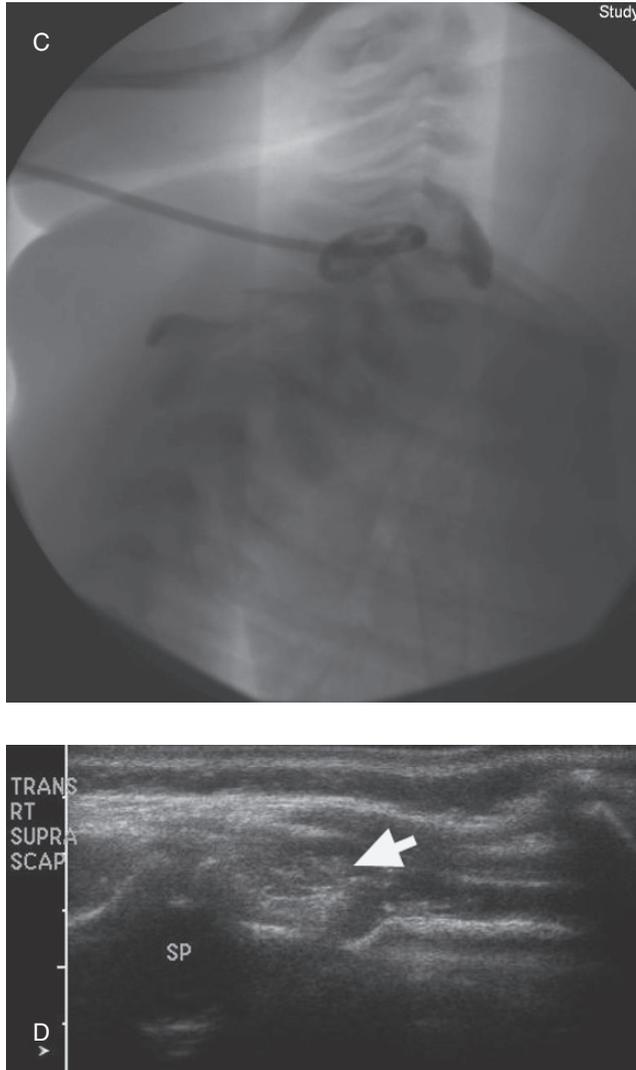


Fig. 39. (Continued).

and ablated with intracystic doxycycline sclerotherapy. In the authors' experience, controlled intracystic treatment of microcysts has resulted in 100% cyst ablation and allows for controlled and staged treatment of large areas of microcystic disease.

TUMOR ABLATION AND EMBOLIZATION

Percutaneous image-guided radiofrequency ablation (RFA) of tumors continues to gain momentum in adult patients as a viable and effective therapeutic option in the treatment of solid tumors in a variety of locations, such as the skeleton, liver, spleen, kidney, adrenal gland, lung, and pancreas (138). In children, RFA has been most widely used in the treatment of osteoid osteoma. The authors use RFA for a growing number of pediatric indications, to include treatment of osteoid osteoma, recurrent/residual hepatoblastoma, hepatic adenoma (139), palliative treatment of pulmonary metastatic

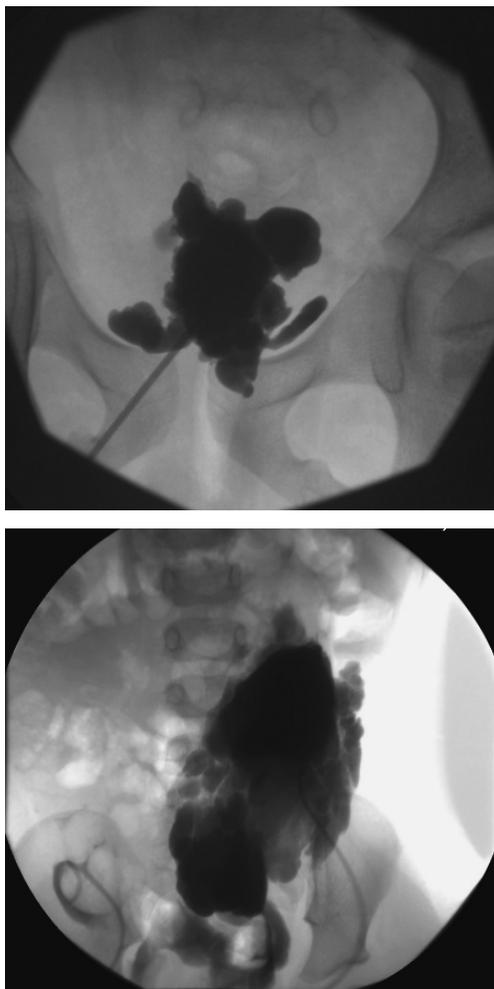


Fig. 40. Recurrent intraabdominal LM treated with dual-drug time-limited ablation therapy. **(A)** A 6-year-old male with recurrent LM following two surgical resections. Contrast study during percutaneous ablation procedure demonstrates large macrocystic LM in the abdomen and pelvis. Imaging follow-up 2 years later demonstrates no recurrence of the macrocystic LM. **(B)** A 1-year-old male with large recurrent intraabdominal and retroperitoneal LM following three surgical attempts at resection. **(C)** Contrast study from the percutaneous ablation procedure demonstrates the largest of three macrocystic components of the LM. Follow-up sonography demonstrates no recurrence of this macrocystic mass following one treatment. Microcystic disease in the pelvis was treated in a subsequent procedure with no recurrence.

osteogenic sarcoma, metastatic leiomyosarcoma, unresectable leiomyoma, aneurysmal bone cyst, and chondroblastoma. Promising uses of renal RFA include treatment of multifocal Wilms tumor, especially in a solitary kidney, and patients with Hippel-Lindau disease; percutaneous RFA is well tolerated, providing an effective nephron-sparing alternative to surgery.

Hepatic and Lung/Tumor Ablation

The majority of experience documented in adult hepatic tumor RFA is in treatment of hepatocellular carcinoma or metastatic disease from organs such as the colon,

pancreas, lung, and breast (138). Treatment of hepatic tumors (hepatoblastoma) in children has been sporadically reported (138). In our clinical practice, focal liver tumors that have been treated with RFA include hepatoblastoma, hepatic adenoma (139), and metastatic leiomyosarcoma. Patients with hepatoblastoma who present the greatest clinical challenge are those with Beckwith-Wiedemann, with multiple hepatoblastomas developing over time. Indeed, these are ideal candidates for RFA, given the risks of repeated surgeries for resection. A percutaneous approach is used in the majority of hepatic tumor RFAs (Fig. 41). In RFA of pediatric liver tumors, we use a 200-W RF generator with internally cooled electrodes (Radionics; Burlington, MA), either

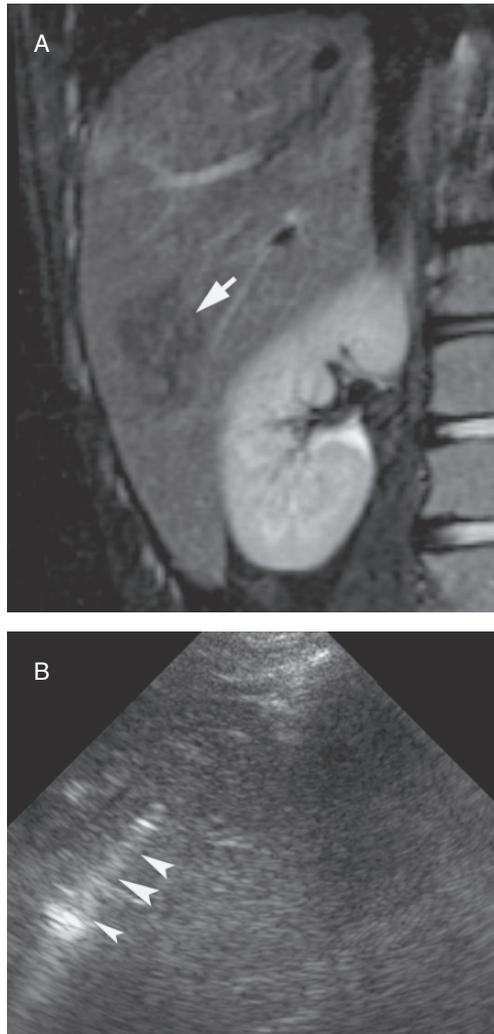


Fig. 41. Hepatic adenoma primarily treated with radiofrequency ablation (RFA). (A) Coronal MRI of the right hepatic lobe demonstrate the focal mass, proven to be hepatic adenoma by percutaneous core needle biopsy (arrow). (B) Sonographic image during RFA demonstrates the RF needle probe (arrowheads) in the adenoma at the beginning of the ablation treatment. (C) CT scan 1 year following treatment demonstrates focal scar (arrow) at the site of the RFA with no residual tumor (CT and MRI 2 years later demonstrated further retraction of the hepatic scar and no recurrent tumor).

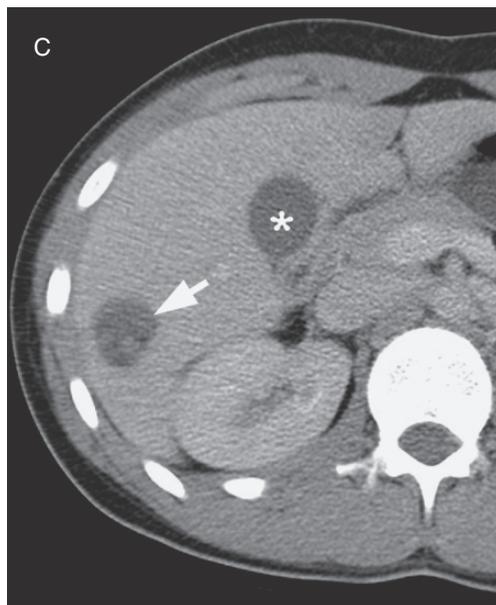


Fig. 41. (Continued).

as a single electrode, or a cluster of electrodes for larger lesions. Overlapping RFAs are performed, with each site reaching a target temperature of at least 60 °C over 12 minutes.

In the treatment of focal liver tumors, the experience in children is currently small, as pediatric-unique guidelines are developed. At the current time, the authors apply guidelines similar to adult literature (138), such that we can expect complete tumor ablation in 90% of patients with tumors smaller than 2.5 cm. Tumors 2.5–3.5 cm in diameter can be expected to be ablated in 70–90% of cases; ablation of tumors 3.5–5.0 cm in diameter will be seen in only 50–70% of cases. In large tumors (greater than 3.5 cm), and with conditions such as Beckwith-Wiedemann with recurrent hepatoblastoma, palliative RFA is a reasonable goal, if treatment can prolong life expectancy with limited systemic symptoms and few minor complications. The volume of effective tumor ablation can be enlarged with hypertonic saline tumor pretreatment and with combination chemoembolization (138).

In the lung, our experience is limited to the treatment of metastatic osteogenic sarcoma patients who are not operative candidates. Internally cooled RF electrodes are used with a 200-W RF generator (Radionics). Large lesions are treated with a three-electrode cluster, each burn reaching a minimum target temperature of 60 °C over 12 minutes. We have used both ultrasound and CT guidance for lung RFA cases, with CT proving to be most useful when lung precludes an effective sonographic window. We have provided palliative RF treatment for metastatic lesions as large as 8 cm. The insulating effect of lung limits the extension of necrosis into adjacent tissue.

Reoperative Tumor Embolization

Tumor embolization in children is most frequently performed for emergency treatment of hemorrhage of unresectable tumors and treatment of multifocal recurrent

tumors, such as renal angiomyolipoma in tuberous sclerosis patients. Uncontrolled hemorrhage in malignancy is rare, yet occurs most commonly with retroperitoneal tumors such as extensive renal tumors, large chest wall tumors, and exophytic hepatoblastoma with peritoneal hemorrhage. More frequently used as the first-line therapy, embolization of hepatic hemangiomas is useful as a means of decreasing the vascular shunting that causes heart failure, diffuse intravascular coagulation (DIC), and death. Diagnosis of tumor-related hemorrhage is made primarily with clinical factors, to include need for repeated transfusions. CT scan is the most common noninvasive modality for diagnosis, rarely demonstrating active extravasation of contrast from a site of hemorrhage. Embolization is performed via transarterial catheterization of the native organ, first diagnosing the focus of hemorrhage with arteriography. Once the site of origin is identified, microcatheter techniques can isolate the selective arteries with subsequent embolization with gelfoam slurry or polyvinyl alcohol particles. If necessary, the main artery supplying the organ of origin (e.g., renal artery with primary renal malignancy) may be coil embolized as the means of controlling hemorrhage.

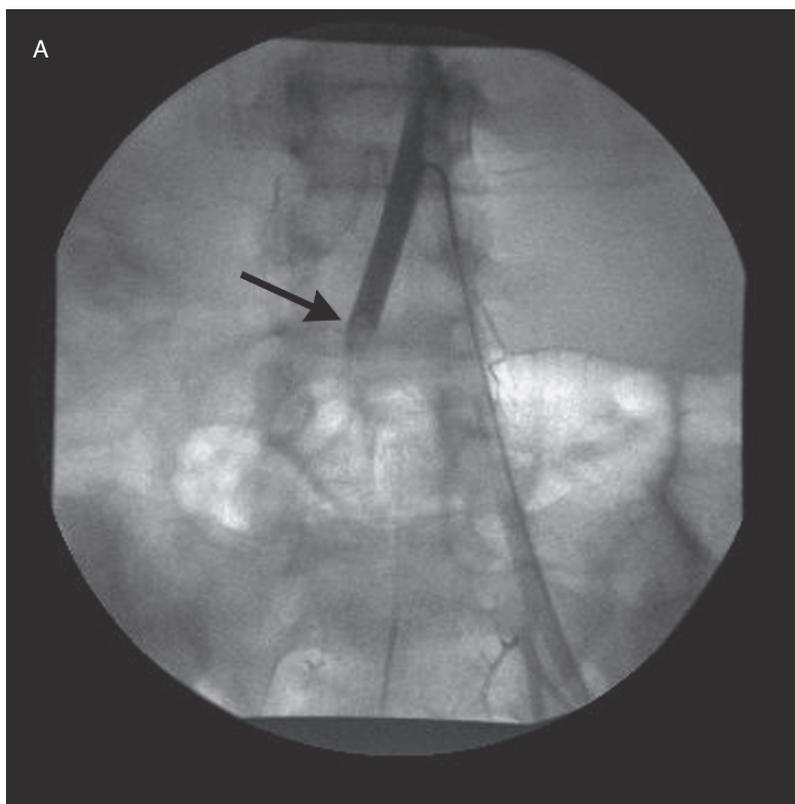


Fig. 42. This is an angiogram obtained on a trauma patient with right lower extremity ischemia. **(A)** Contrast injection from above, and occlusion (arrow) of the right common iliac artery. **(B)** Retrograde injection from the right common femoral artery, and demonstrates thrombus within the artery (arrows). An attempt was made to cross the occlusion, but was not successful. At surgery, the artery was completely transected.

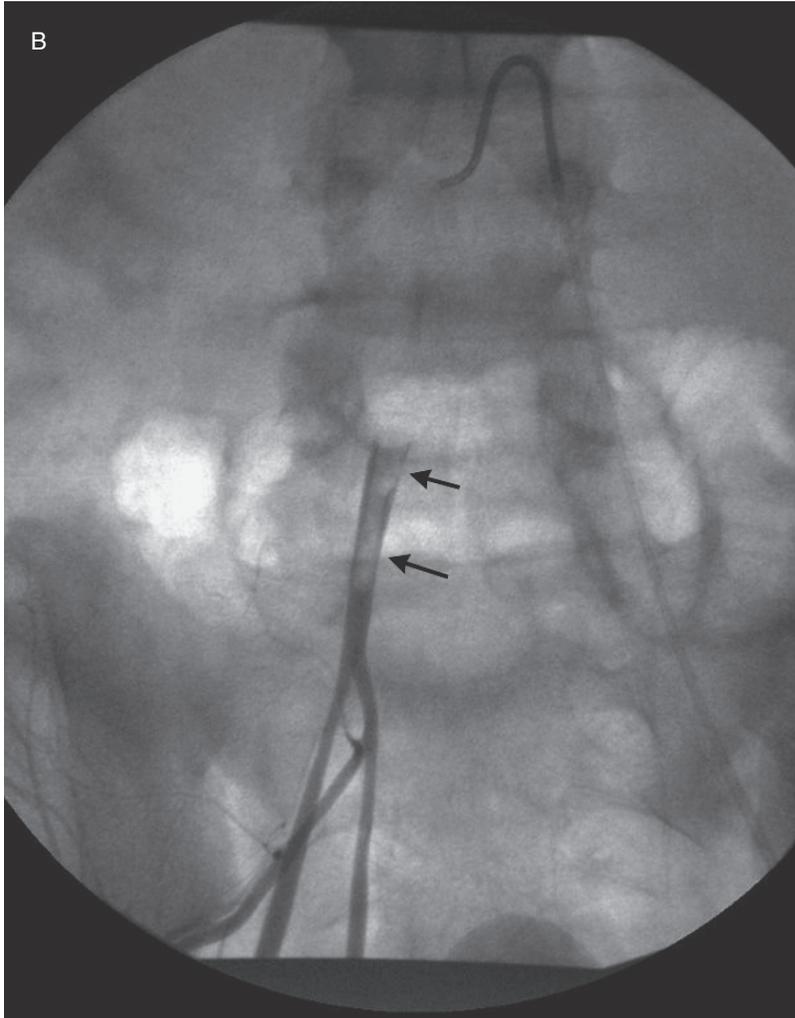


Fig. 42. (Continued).

VASCULAR INJURY

Vascular injury can be either after external trauma or iatrogenic. Trauma victims can have direct vascular injury such as amputation, avulsion, crush injury, or occlusion or hemorrhage related to fractures. Iatrogenic injury can present after surgery as bleeding (such as after resection or biopsy, or at an anastomotic site), or ischemia. CT is excellent for excluding vascular trauma (140–142). CT scanning can identify hematomas, and newer CT angiography techniques can accurately depict vascular injury and occlusion (3). However, CT does not have the dynamic information present with angiography that can better identify the occlusion, length of involved segment, amount of reconstitution, and associated findings such as an intimal flap or pseudoaneurysm formation (Fig. 42). This information is important for surgical planning, and angiography is still indicated in certain circumstances when vascular injury is identified by imaging.

Angiography can provide diagnostic information and guide therapeutic interventions in patients with vascular injury (143–155). If there is hemorrhage in an unstable patient,

the bleeding site can be identified and embolized using particulate embolic material, coils, or other agents (147–151) (Fig. 43–45). Splenic embolization can be performed for hemorrhage in unstable patients (148–150). Bleeding from pelvic fractures can be controlled with internal iliac arterial embolization (151).

Vascular occlusion is readily identified. If there is an obvious intimal flap from a dissection, a stent may be indicated, although this is controversial in children (143–146). Endovascular techniques with aortic stent grafting are becoming more common in adolescents and adults with trauma and aortic injury (143,147). There are reported cases in adults of recanalization of posttraumatic occluded arteries (144–146). In our



Fig. 43. This patient presented with hemobilia after surgery for a large posttraumatic hepatic hematoma. (A) Angiogram that shows a pseudoaneurysm (arrow), which is better seen after superselective angiography (B) (arrow). (C) Embolization with coils (arrow), and occlusion of the pseudoaneurysm. The hemobilia resolved.

experience, occlusion of a peripheral artery in a child is often caused by complete disruption, although vasospasm stops continued hemorrhage (Fig. 42). Attempts at recannulation in these patients might lead to greater problems. Arterial occlusive disease is uncommon in children. However, fibromuscular dysplasia or other arteritides, trauma (external or iatrogenic), venous stenoses from prior central venous access, and anastomotic strictures (usually after transplantation or with dialysis shunts) may present with vascular stenosis (152–155) (Figs. 46 and 47]. Angioplasty is indicated in these patients, with stenting reserved for elastic recoil or dissection after the dilatation (152–155).

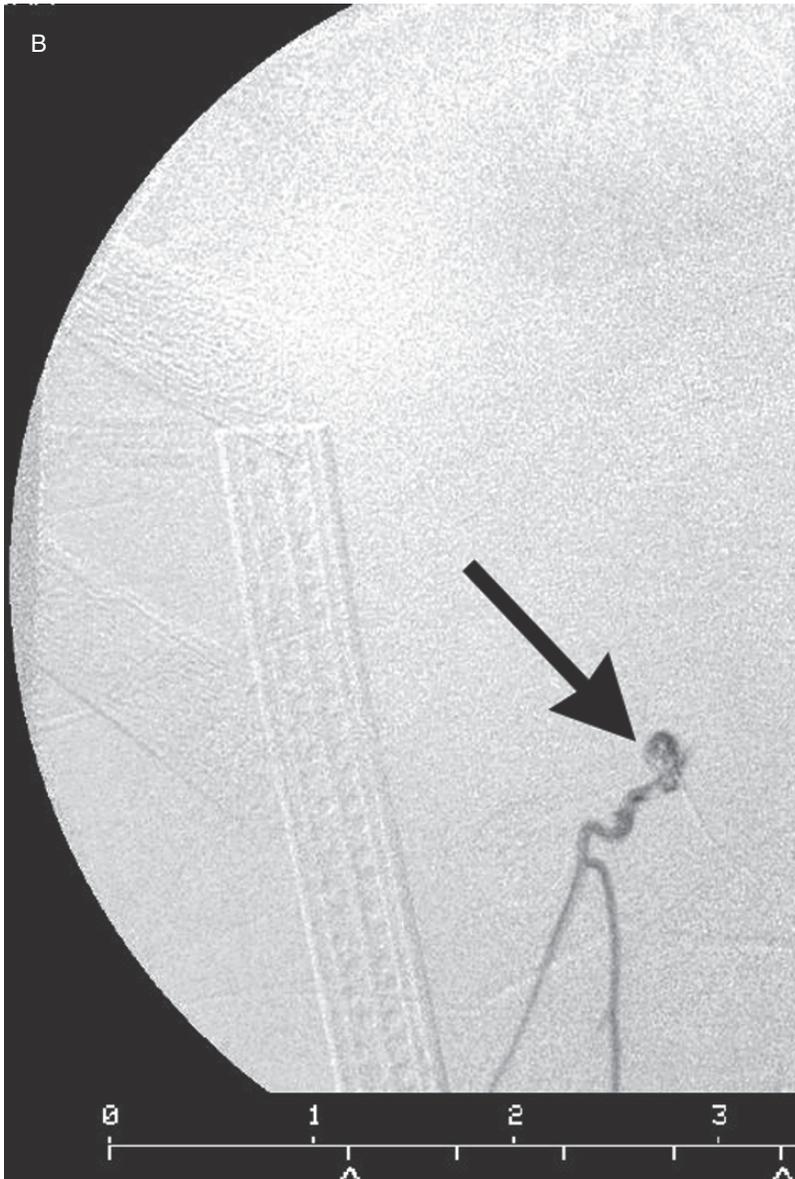


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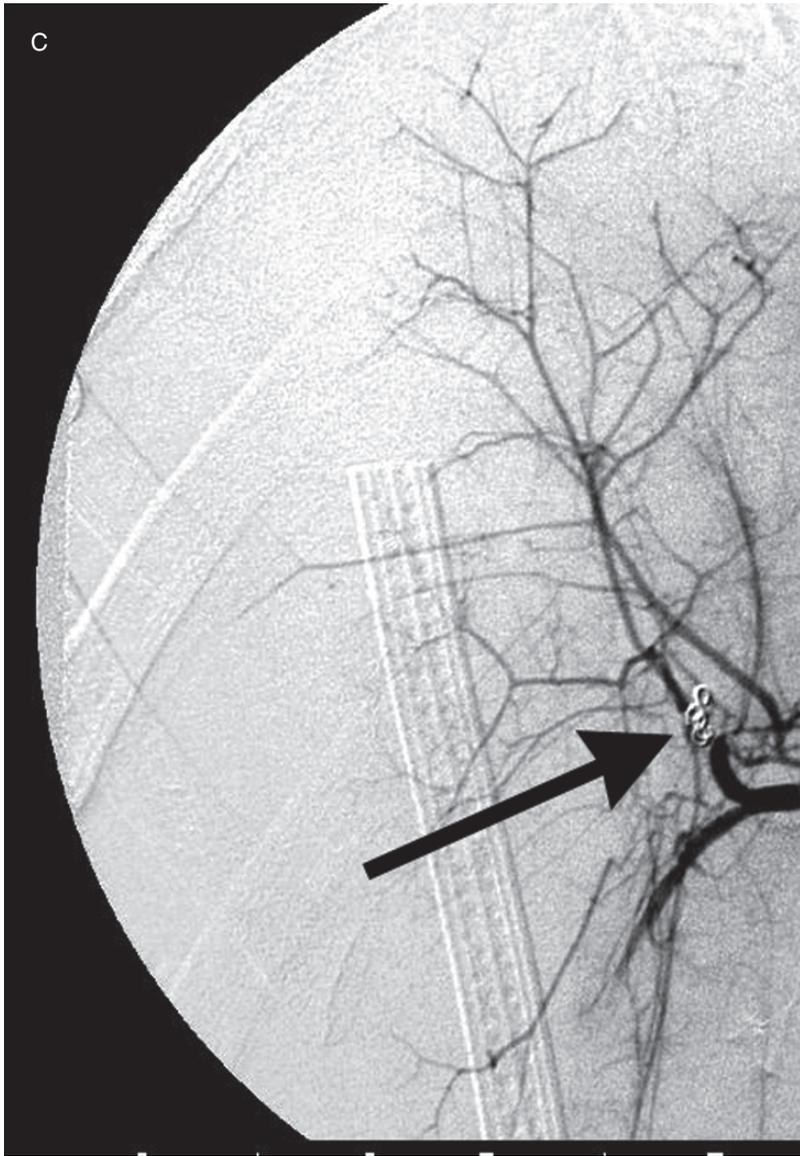


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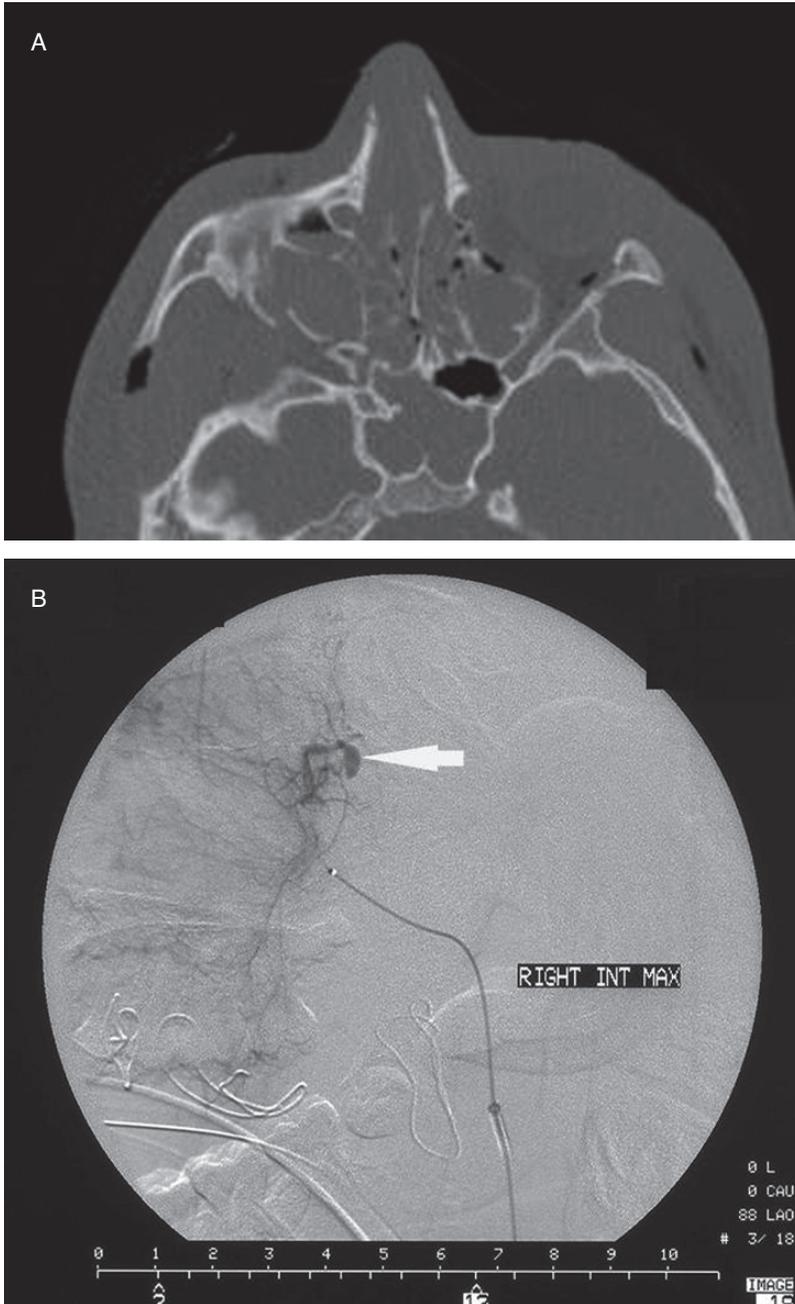


Fig. 44. Traumatic pseudoaneurysm. (A) Facial CT showing extensive facial fractures. The patient had uncontrollable transfusion dependent bleeding despite surgical packing. (B) Pseudoaneurysm (white arrow) on an angiogram. (C) Angiogram after coil (black arrow) embolization, with occlusion of the pseudoaneurysm.



Fig. 44. (Continued).

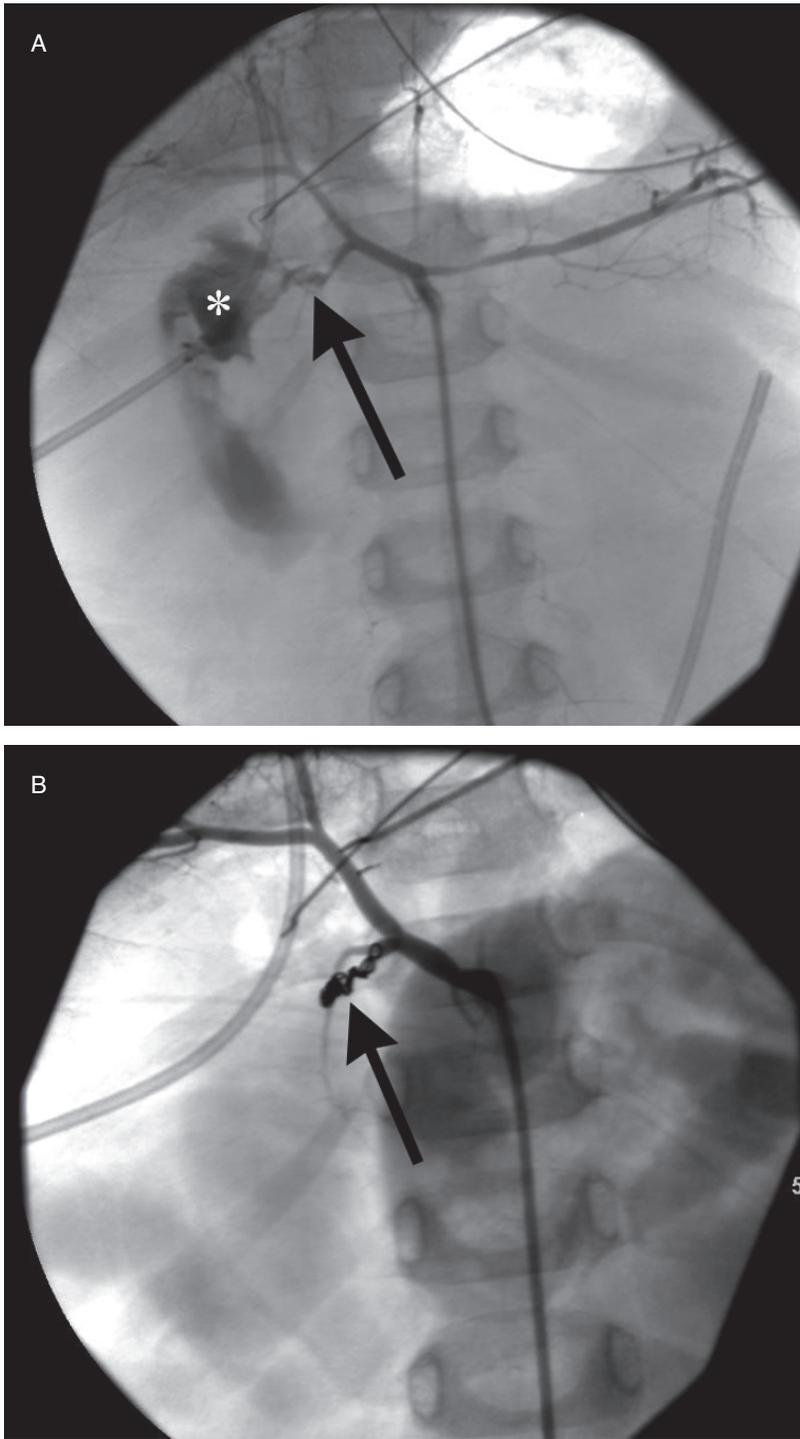


Fig. 45. This postsurgical child had significant hematemesis. **(A)** Angiogram shows a bleeding site (arrow) with extensive extravasation into a hematoma (*). **(B)** Resolution of the hemorrhage following embolization with coils (arrow).

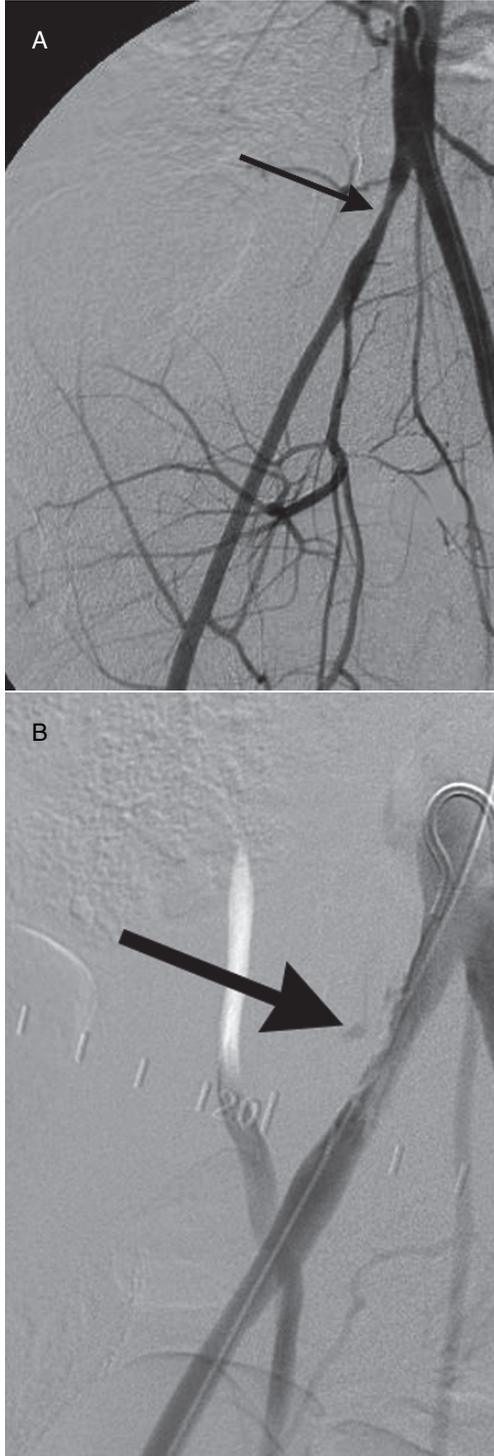


Fig. 46. Arterial trauma with stent placement. This patient had direct right common iliac artery injury from placement of a trocar during a laparoscopic appendectomy. He had surgical repair, but now has claudication. **(A)** Arterial narrowing at the surgical site (arrow). **(B)** After angioplasty, irregularity of the artery and some extravasation (arrow). **(C)** Angiogram after stent deployment; the irregularity and the extravasation have resolved.

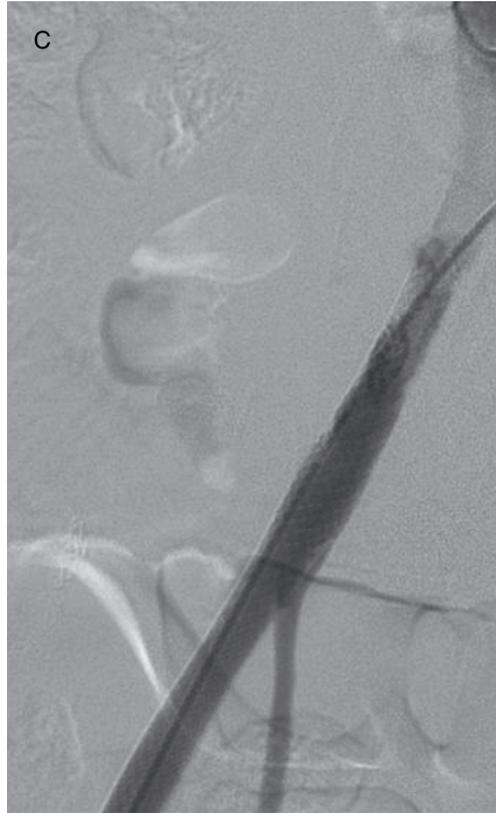


Fig. 46. *(Continued).*

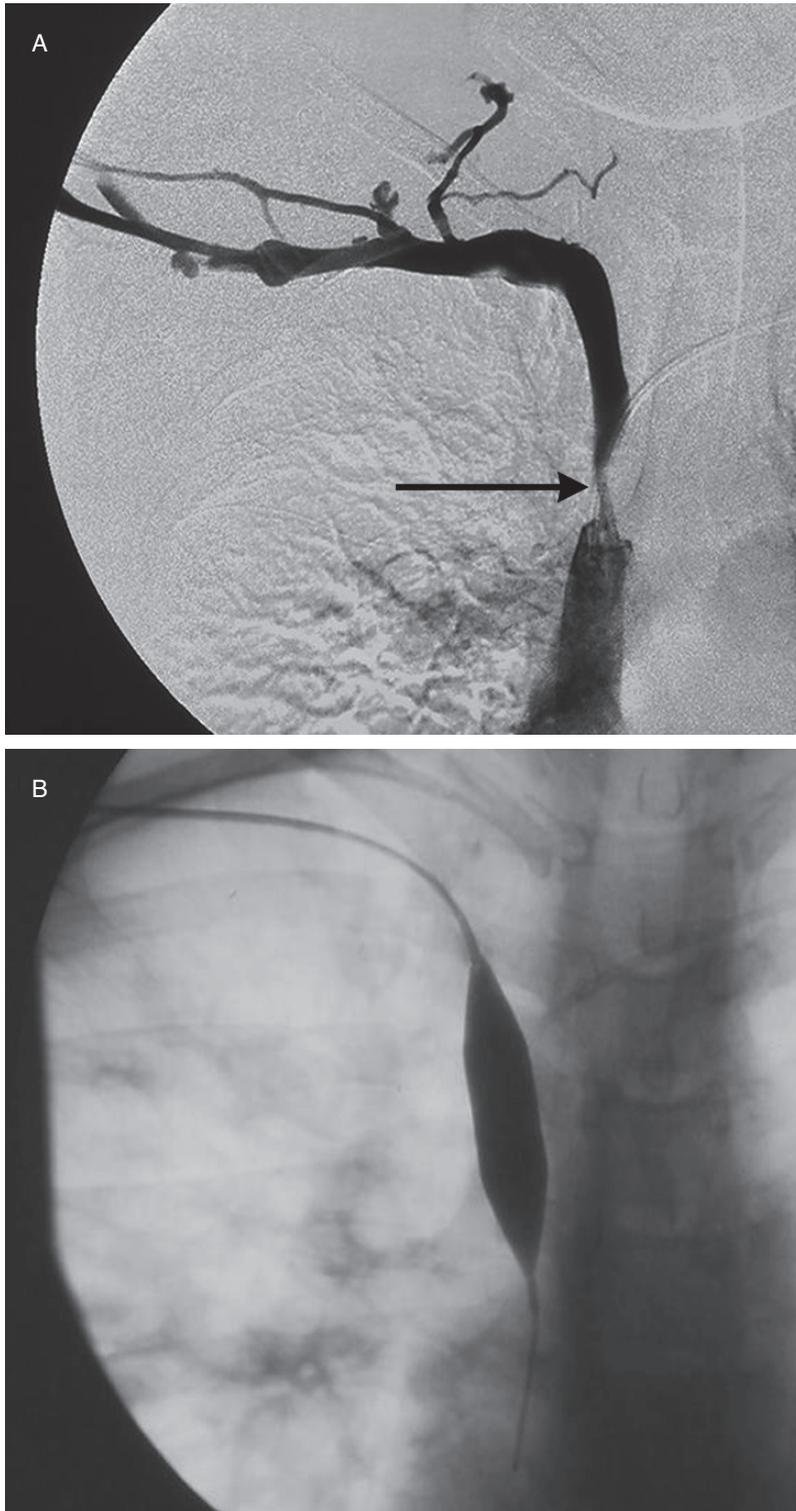


Fig. 47. Venous stenosis. (A) Trauma from the catheter tip has caused a venous stenosis of the SVC (arrow). (B) Balloon angioplasty was performed, with resolution of the stenosis (C) and good flow through the SVC (arrow).

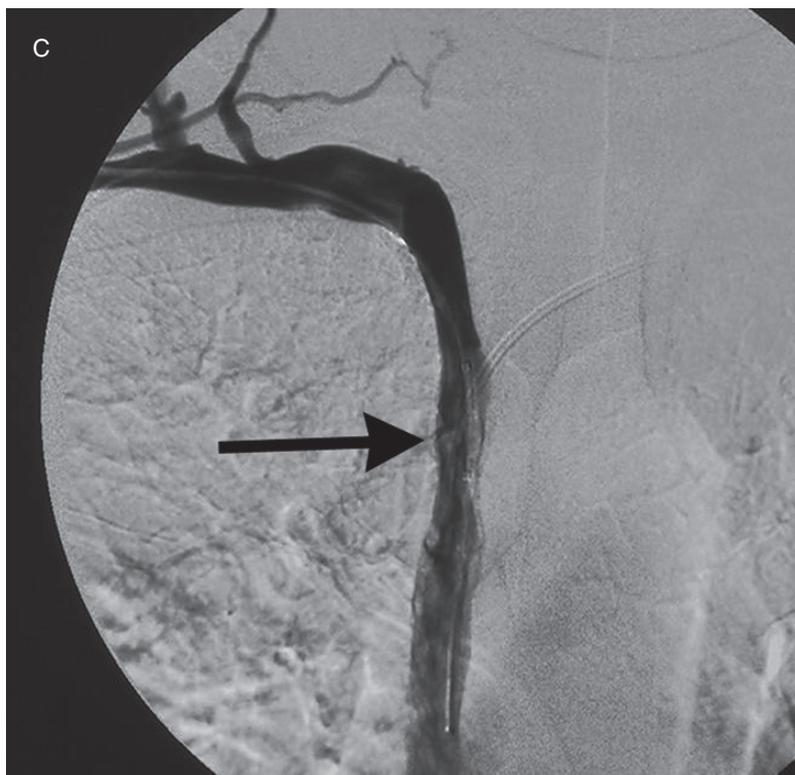


Fig. 47. (Continued).

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2

Techniques for Difficult Venous and Arterial Access

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CONTENTS

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INTRODUCTION

The maintenance of central venous access is critical to the management of pediatric patients in various clinical scenarios, and the establishment of vascular access is one of the most common procedures performed by pediatric surgeons. This is of utmost importance particularly in those who are dependent on parenteral nutrition. Although the internal jugular or subclavian vein is an adequate route of access in most patients, a common pitfall associated with long-term venous access is the eventual exhaustion of traditional sites of peripheral or central catheterization over time. This is caused by multiple complications, including catheter sepsis, venous stenosis and/or thrombosis, or catheter malfunction. It is therefore critical to understand the various options for long-term central venous access in these patients. In this chapter, we provide a comprehensive and multidisciplinary approach to the management of reoperative venous access with particular attention to preoperative planning and imaging, as well as specific techniques in interventional radiology and surgery.

PREOPERATIVE PLANNING

Most patients who require long-term vascular access have an associated complex medical and surgical history. In addition to their underlying disease, such patients have also often had complications related to venous access itself. The development of a rational strategy for central venous access is dependent on the recognition of the disease processes most likely to require long-term vascular access and the

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identification of suitable vein(s) that will provide extended patency. Patients at high risk of requiring prolonged venous access generally include those with intestinal failure, owing to either quantitative (short bowel syndrome) or qualitative (intestinal dysmotility or long-segment Hirschsprung's disease) dysfunction. In other patients, the presence of malignancy may require prolonged central venous access for the delivery of parenteral nutrition, chemotherapeutic agents, and/or frequent blood sampling. Therefore, the prevention of central vein occlusion is a critical goal. In these patients, it is important to avoid ligation of major veins during placement of central venous catheters (CVCs). Minimization of the number of CVCs inserted in these patients is also very important. Referring physicians should understand the risks associated with CVC insertion. Although CVCs greatly facilitate patient care in terms of avoiding needle sticks for blood draws and limiting peripheral intravenous lines, future standard insertion sites may be compromised by the overliberal utilization of CVCs.

Central venous catheter replacement in patients requiring prolonged central venous access should be performed only when absolutely necessary. For example, cuts or breaks in the catheter should seldom dictate the need for a catheter replacement, but rather catheter repair kits should be used to preserve CVCs. In addition, catheter patency can usually be reestablished from occlusion owing to drug, clot, or lipid solutions. In order to salvage a CVC, bacteremia can often be managed with a course of antibiotics through the catheter. Finally, it is important to assess the purpose of central venous access. Insertion of a second CVC may be avoided if a schedule is devised to enable medications and/or blood draws to all be done through an existing catheter.

The patient population requiring prolonged central venous access is prone to catheter-related complications. Technical misadventures during CVC insertion may be avoided by eliciting a careful history, which reveals the need for preoperative imaging. Many factors contribute to catheter-related complications, including catheter tip location, duration, and prior catheterization (1,2). The number and sites of previous CVCs, complications from previous CVC insertion, and the presence of coexisting pathology are all key factors that must be considered. In addition, a history of prior head and neck surgery, previous radiation exposure to the mediastinum, or known mediastinal, lower extremity, or abdominal pathology should alert the clinician that standard sites for central venous access may be unsuitable.

The use of preoperative imaging prior to CVC insertion may be guided by physical examination, as such findings are frequently related to central vein occlusion or stenosis. For the upper extremities and torso, signs associated with occlusion of the veins that drain into the superior vena cava (SVC) may be identified. For example, upper extremity edema, prominent collateral veins, varicosities, and plethora are all characteristic of SVC obstruction. Alternatively, if venous return via the inferior vena cava (IVC) is impeded, similar findings of edema and venous hypertension will be evident in the lower extremities.

PREOPERATIVE IMAGING

Venous anatomy may be imaged by a variety of modalities, but ultrasound and magnetic resonance venography are the most commonly utilized techniques. In the following section, we discuss the use of these and other radiologic modalities. Figure 1

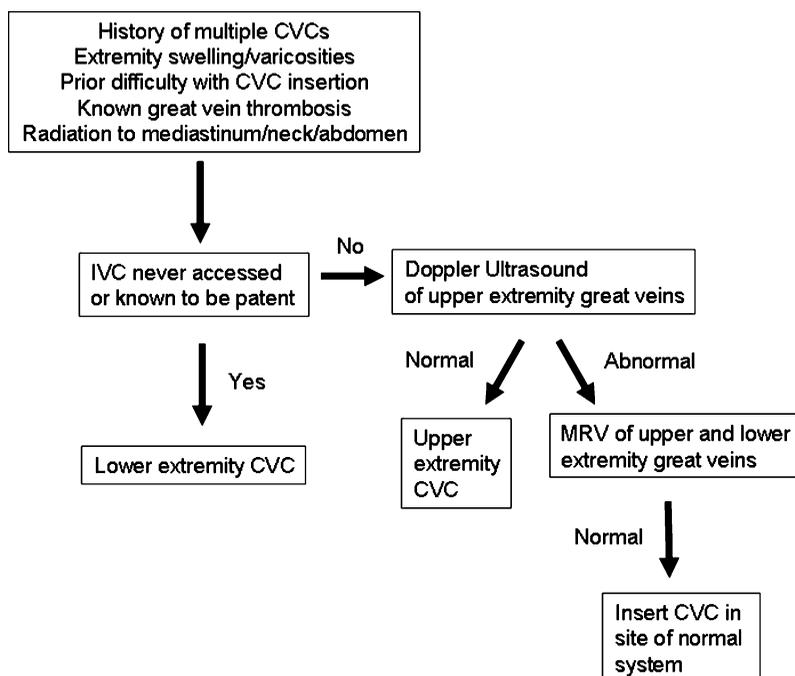


Fig. 1. Proposed algorithm for radiological evaluation of high-risk patients for CVC insertion. MRV: magnetic resonance venography.

depicts a proposed algorithm for the radiologic evaluation of high-risk patients for CVC insertion.

Ultrasound

Ultrasound is a useful noninvasive method to evaluate suspected upper extremity deep vein thrombosis (DVT) and central venous obstruction.(3–5). It is typically performed with high-resolution ultrasound equipment using 7.5- to 10-MHz transducers. A typical ultrasound examination to evaluate the status of upper extremity great veins involves examination of the bilateral internal jugular (IJ), brachiocephalic, subclavian (SC), and axillary veins. Direct visualization of the SVC, brachiocephalic, and medial aspect of the subclavian veins is often limited by the sternum and clavicles.

Accurate diagnosis of upper extremity and central venous obstruction is dependent on three components of the ultrasound examination: (1) real-time gray scale evaluation; (2) color Doppler imaging; and (3) Doppler waveform analysis. The lumina of normal patent IJ and SC veins are hypoechoic. Grey scale signs of thrombosis include variable echogenic intraluminal thrombus (Fig. 2), noncompressibility, loss of respiratory variability and vessel pulsation, and absence of response to Valsalva or sniff maneuvers (6). Color Doppler imaging allows rapid localization of vessels, differentiation of veins from arteries, and delineation of thrombus. Doppler waveform analysis allows evaluation of reflected right atrial pressures and changes associated with respiration. Patency of interrogated veins to the right atrium is implied by normally reflected triphasic atrial waveforms with subtle respiratory variation.

With the patient in the supine position, the IJ veins, which are superficial and easily visible and compressible by the transducer, are followed to their junction with

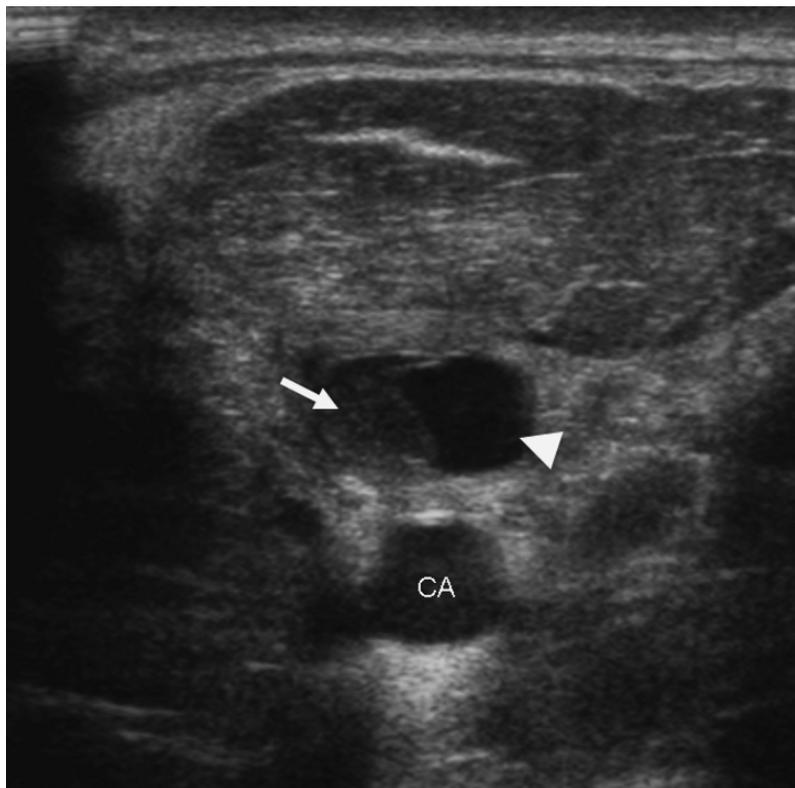


Fig. 2. Transverse ultrasound of the IJ vein with delineation of thrombus (arrow). Arrowhead demonstrates the normal hypoechoic appearance of the patent portion of the lumen. CA: carotid artery.

the brachiocephalic veins. The latter may be visualized from either a suprasternal or medial infraclavicular acoustic window. The SC veins, which are usually more caudal and superficial to the SC arteries, are imaged to their junction with the IJ veins. It is important to differentiate large venous collaterals that may develop in the setting of prior SC vein thrombosis. Compared to the SC veins, such collaterals, however, are typically smaller and in atypical locations relative to the SC artery. Increased velocity and turbulence identified within the vessel signify stenosis. Although it may not be possible to directly image the SVC and brachiocephalic veins, careful examination of the SC and IJ veins for dampened atrial pulsations and abnormal response to respiratory maneuvers may be suggestive of central obstruction. With regard to the detection of SC vein stenosis by ultrasound, false-positive results are rare. False-negative studies may occur in cases of small nonobstructive thrombus or in patients with extensive collateral networks.

Magnetic Resonance Imaging (MRI) Venography

MRI venography is highly accurate in the delineation of central venous anatomy, and the correlation of venous obstruction with contrast venography has been excellent (7–9). The multiplanar capabilities of MR venography are particularly useful in assessing thoracic inlet and mediastinal veins, which may be difficult to image entirely in a

single plane. MR venography is not contraindicated in patients with allergy to iodinated contrast or in those with renal insufficiency. In children, however, imaging often requires sedation because patient motion significantly degrades scan quality.

Two-dimensional time of flight (2-D TOF) sequence, a flow-sensitive method that does not require IV contrast, is the most frequently used method of MR venography. The lack of IV contrast requirement is particularly important in children being imaged for CVC placement, because peripheral venous access is especially difficult in this cohort of patients. Axial, sagittal, and coronal planes are imaged with presaturation bands placed adjacent to the imaged slices to suppress the adjacent arterial signal (Fig. 3). In adults, the sensitivity and specificity for the detection of central venous occlusion by the 2-D TOF imaging are approximately 97 and 94%, respectively, and this technique has excellent correlation with conventional venography (10,11). MR venography in children with suspected CVC-related thrombosis has been shown to be more accurate than ultrasound or contrast studies for defining the extent of thrombosis (7). In that study, venous anatomy and patency for reinsertion of CVCs was correctly defined by MR venography. However, 2-D TOF imaging requires relatively long scanning times, as well as imaging in multiple planes. Thus, three-dimensional gadolinium-enhanced gradient echo MR venography has been developed, which can produce high-quality



Fig. 3. Magnetic resonance venogram of the neck and chest demonstrating an occluded left IJ vein. Arrow indicates absence of left IJ vein signal.

images that can be acquired within a single breath hold (9,12–14). This modality may only be useful in a cooperative pediatric patient or under general anesthesia.

Contrast Computed Tomography (CT)

Contrast enhanced CT is a reliable technique for the evaluation of the etiology of SVC obstruction, and its primary advantage over ultrasound is its ability to image beyond bone and air (15). The SVC is opacified with iodinated contrast material, and the luminal patency and relationship of the SVC to adjacent structures may be studied. Helical CT phlebography can provide multiplanar and three-dimensional reconstruction of the central venous anatomy and mapping of collateral tributaries in cases of SVC obstruction (16). Technical limitations of this technique include streak artifact and intraluminal filling defects caused by contrast dilution by unopacified blood from the IJ and azygous veins and peripheral displacement of intraluminal contrast by laminar flow (17). As a result of these potential flow-related artifacts, the diagnosis of venous obstruction by CT scanning requires two criteria: (1) decreased or absent venous opacification distal to the site of an obstructive intraluminal filling defect or compressive mass; and (2) the presence of collateral pathways (16). As with conventional contrast venography, potential contraindications to CT scanning include the need for iodinated contrast in a patient with a history of renal insufficiency or a history of allergic reaction.

Conventional Contrast Venography

The gold standard for the evaluation of central venous obstruction is conventional contrast venography, and it is used generally when other studies are impractical or equivocal, and in order to thoroughly define venous anatomy in complicated clinical scenarios. The advantage of contrast venography is not only identification of venous stenosis or obstruction, but also real-time evaluation of flow through the central veins and collateral channels. This information is critical in planning the most appropriate approach for CVC placement. Suspected SVC syndrome may be evaluated by conventional contrast venography (18–20). Venography has been reported to be superior to CT scanning in the opacification of collateral venous tributaries and in the determination of the degree of venous obstruction (17).

Evaluation of axillary veins and central veins in the chest requires placement of a large-bore intravenous cannula in the antecubital fossa. Nonionic contrast is hand-injected, and digital acquisition is performed with the arm in a neutral palmar position. The filling of the deep veins of the arm (brachial and axillary veins) may be aided by placement of a tourniquet. To better visualize the central veins, the contrast-filled arm may be elevated or manually compressed to increase contrast drainage, and the SVC can be evaluated by simultaneous antecubital injections. In patients who cannot receive iodinated contrast because of renal failure or allergic reaction, gadolinium or carbon dioxide can be used (21).

INTERVENTIONAL TECHNIQUES

Translumbar Catheterization of the IVC

The use of translumbar percutaneous IVC cannulation for long-term parenteral nutrition in adults was first described in 1985 (22). This technique is a safe and effective option in both adults and children for the administration of parenteral nutrition, long-term antibiotic therapy, and chemotherapy, as well as for stem cell harvesting prior to

bone marrow transplantation (23–26). In the adult population, infectious and thrombotic complications have been reported to be less than 5% (23,26,27). Although it has been described, the risk for IVC thrombosis is less in patients with translumbar catheters than in the setting of femoral venous catheterization, because of the relatively high IVC flow (25,28–30). In the majority of cases of thrombotic complications, management consists of the administration of fibrinolytic agents.

Complications specific to the technique of translumbar catheterization are migration of the catheter into the subcutaneous tissues, retroperitoneum, or iliac veins (29). Contraindications to translumbar CVCs include coagulopathy (owing to the risk of inadvertent aortic puncture) and presence of infection on the lower back or right anterolateral abdomen in the region of the anticipated puncture or tunnel sites. Marked truncal obesity has also been proposed to be a contraindication, because of an increased risk of catheter migration (28,29).

Preprocedure imaging should confirm IVC patency and exclude retroperitoneal pathology. With the patient in the prone position, a puncture site is chosen just cephalad to the right iliac crest and approximately 8–10 cm to the right of midline. Under fluoroscopic guidance, a 21-G microaccess needle is advanced into the infrarenal IVC at the L2 to L3 level by directing it cephalad toward the anterolateral margin of the vertebral bodies. Alternatively, a femoral venous catheter placed with the tip in the IVC can confirm position of the IVC and be used as a target for puncture. One group has recommended a more cephalad puncture of the IVC, with a skin puncture site just below the 12th rib (31). In infants and small children, translumbar ultrasound may be utilized to visualize the IVC, and access can be achieved under real-time ultrasound guidance. After confirmation of IVC access by aspiration of blood, contrast may be injected to confirm IVC placement and exclude entry into the renal vein. By Seldinger technique, a guidewire is then advanced into the IVC, and following placement of a coaxial dilator, a stiffer guidewire is advanced into the SVC or right atrium. The tract is then serially dilated, and an appropriately sized peel-away sheath is placed. A subcutaneous tunnel is extended laterally to the anterior lower abdominal wall, and the catheter is advanced through the peel-away sheath into the central circulation. Final catheter tip position should be as high as possible at the IVC/right atrial junction. Manual compression and supine patient position are sufficient to achieve hemostasis at the time of catheter removal.

Transhepatic Catheterization of the IVC

Although uncommon, occlusion of both the SVC and the infrarenal IVC presents a challenge to central venous catheterization. In such cases, the retrohepatic portion of the IVC is typically patent. Percutaneous access of the retrohepatic IVC can be achieved directly via the liver parenchyma or the hepatic veins. The technique of transhepatic central venous catheterization was first described in adults in 1991 for the administration of long-term parenteral nutrition (32). Transhepatic catheterization has been successful in children for short-term interventional radiology procedures and cardiac catheterizations (33), emergency short-term dialysis (34), and prolonged delivery of parenteral nutrition (35,36). Rates of infectious and thrombotic complications are comparable to standard central venous access approaches (35).

Catheter dislodgement with subsequent bleeding from the hepatic parenchymal tract and hepatic vein thrombosis are potential complications unique to transhepatic CVC placement. Catheter migration and dislodgement may occur because of diaphragmatic

movement during deep respiration. In infants and small children, severe abdominal distension and growth spurts may also result in catheter malposition (35). Hepatic vein thrombosis is usually asymptomatic if the other hepatic veins are patent, although acute Budd-Chiari syndrome and subsequent death has been described in 1 child (37). IVC thrombosis as a complication of transhepatic catheterization has not been reported in the pediatric population. Cholangitis, hemobilia, and liver abscess are other potential complications of this approach. Uncorrectable coagulopathy, massive ascites, and biliary infection represent contraindications to the placement of a transhepatic CVC.

Patient evaluation prior to transhepatic central venous catheterization usually consists of contrast-enhanced CT scanning of the abdomen to evaluate the liver, hepatic veins, retrohepatic IVC, subphrenic space, and perihepatic peritoneal cavity. Ultrasound can further evaluate the hepatic veins and IVC, and is useful in planning the optimal approach (38).

With the patient in the supine position, the right hepatic vein is most commonly accessed via a lateral approach in the mid axillary line through the 10th or 11th interspace. Transpleural puncture must be avoided, as it can result in pneumo- or hemothorax. Alternatively, the middle hepatic vein may be accessed via an anterior subcostal approach. Direct ultrasound and fluoroscopic guidance is utilized to direct a 22-G Chiba needle into the appropriate hepatic vein. Following contrast injection to confirm needle tip position, a platinum-tipped guidewire is advanced into the right atrium. A coaxial dilator is advanced into the IVC, and the guidewire is exchanged for a stiffer wire with the tip placed ideally in the SVC to avoid dysrhythmias. Alternatively, puncture can be made with a sheathed 19-G needle, through which a guidewire can be directly passed. Serial dilation over the wire by Seldinger technique is followed by insertion of a peel-away sheath, and finally the venous access catheter after tunneling to the anterolateral abdomen. Final catheter tip position should be at the SVC/right atrial junction to allow for catheter migration, particularly in infants who are growing rapidly (35). In children who are growing rapidly, chest radiographs can be obtained at 2- to 3-month intervals to assess catheter tip position. If necessary, a new catheter can be exchanged over a wire and placed via a new tunnel and catheter exit site.

Removal of transhepatic catheters carries a risk of intraperitoneal or subcapsular hemorrhage along the catheter tract. Track embolization should be performed by deploying coils or Gelfoam pledgets as the catheter is slowly withdrawn from the liver (34).

Catheterization of Collateral Veins

Percutaneous placement of CVCs can occasionally be placed via enlarged upper or lower extremity collateral veins. Only a few case reports have documented this approach, and they include CVC placement via intercostal veins (39–42) and via thyrocervical collaterals (43). Contrast venography is very useful in evaluating the size and course of the collateral veins, as well as the reconstituted central vein. It is important to recognize the presence and status of other collateral veins, because severe symptoms may be precipitated by catheter-induced thrombosis of a solitary major draining vein. Successful cannulation and advancement of CVCs through tortuous collateral veins usually requires ingenuity and the application of angiographic tools, including hydrophilic selective catheters and torque-control guidewires. Combined interventional radiology and surgical approaches have been described, and in many cases, may offer the best chance for successful CVC placement.

Angioplasty or Stent Placement

In the setting of an indwelling CVC, obstruction of the vein harboring the catheter can result in acute signs and symptoms, including edema, pain, and venous hypertension. In this scenario, thrombolytic therapy has a fairly high rate of success. Subsequent venography often demonstrates a residual venous stenosis. Balloon angioplasty of the residual stenosis should be considered in order to preserve the central vein for continued vascular access (Fig. 4) (44,45). The placement of an endovascular stent is an additional strategy, but this intervention requires further study to determine its utility in the salvage of central venous access sites. One indication for stenting may be “early recoil” of the venous stenosis to its original size after successful dilation. More recently, sharp recanalization of chronically occluded central veins has been described and may offer another option for management of these challenging patients (46–48). These techniques require advanced endovascular skills and are probably limited to centers committed to such interventional radiology procedures.

Peripherally Inserted Central Catheters (PICCs)

Recently, PICCs have become widely utilized for prolonged administration of parenteral nutrition, antibiotics, and chemotherapy, as well as for intermittent blood sampling (2,49–51). It has been reported that with fluoroscopic guidance, PICC placement is successful in children in more than 92% of cases (52). PICCs have been demonstrated to be a reliable and safe route of central venous access in neonates and children, with an infectious complication rate of approximately 2% (53,54). The overall complication rate is low and most commonly involves catheter occlusion, displacement, and fracture, as well as phlebitis and venous thrombosis. In a study in children, a higher risk of complications has been demonstrated more than 30 days after PICC placement, and this has called into question whether PICCs should be used for long-term central venous access (2). A recent review of the use of PICCs for long-term antibiotic and parenteral nutrition administration in adults reported no clear advantage to the use of PICCs compared to traditional CVCs (51). Prospective, randomized studies are required to determine whether an advantage of one technique over the other exists. Contraindications to PICC placement include coagulopathy, lack of peripheral

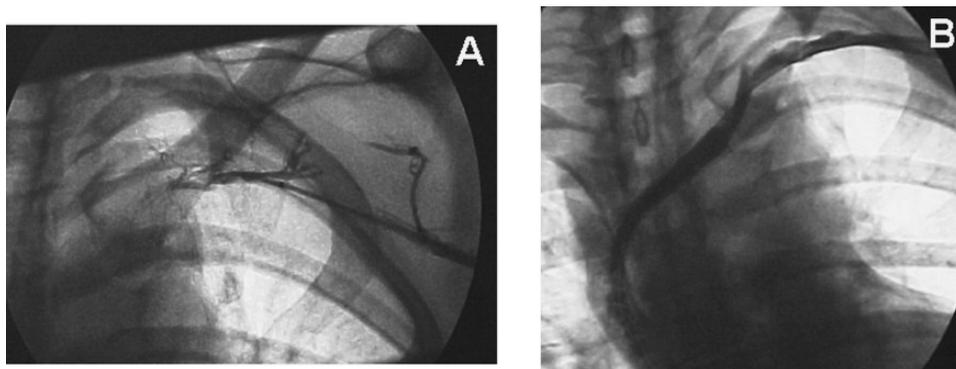


Fig. 4. Balloon dilation of left SC vein stenosis. (A) Venogram demonstrating near-complete occlusion of left SC vein. (B) Venogram after successful balloon dilation demonstrating patency of SC and innominate veins without residual stenosis.

venous access, the presence of venous thrombosis, end-stage renal disease (owing to the potential future need for the creation of an arteriovenous fistula for hemodialysis access), a history of radiation therapy, and prior axillary lymph node dissection.

The most commonly used PICC sizes in children are 2-, 3-, and 4-F catheters. An appropriate vein is selected for cannulation, and usually superficial veins are used, including the cephalic or basilic vein in the upper extremity, the temporal or posterior auricular vein in the scalp, and the saphenous vein in the lower extremity. If necessary, deeper veins, including the axillary, femoral, popliteal, and external jugular veins, may be catheterized for PICC placement. The patient is placed in the supine position with the arm abducted at a 90° angle, and the head is turned toward the arm of insertion. The catheter length is estimated as the distance from a marked insertion site to the midpoint of the sternum. If the insertion site is in the lower extremity, the catheter should be measured to at least 2 cm above the umbilicus. Under sterile conditions, the vein is localized by either palpation or direct visualization or is identified by ultrasound guidance. Venipuncture is performed with the appropriately sized needle, a guidewire is advanced through the needle into the vein, a dilator/peel-away sheath is inserted over the wire by Seldinger technique, and after removal of the dilator and wire, the PICC is advanced through the peel-away sheath to the previously measured catheter length. Alternatively, some centers prefer to access the vein initially with a peel-away sheathed needle, remove the needle, and immediately pass the PICC through the sheath. If the PICC is not placed under fluoroscopy, a chest radiograph is performed to confirm the position of the catheter tip in the SVC, and incorrectly positioned catheters are adjusted as necessary. Because of a high rate of initial incorrectly positioned PICCs in children, it has been advocated that pediatric PICC placement be performed in an interventional radiology suite with fluoroscopic assistance to allow real-time adjustment of catheter tip position and to reduce the number of catheter manipulations required (55). Others have proposed that a dedicated PICC team with ultrasound capability for bedside PICC placement can reduce cost by avoiding the use of expensive interventional radiology facilities (56). The relative cost-effectiveness of these two approaches to PICC placement requires further study.

SURGICAL TECHNIQUES

The SC vein and external or IJ veins are the traditional sites for access to the SVC, and either a percutaneous or surgical cut-down approach may be performed. When the SVC is inaccessible owing to thrombosis of the IJ, SC, or brachiocephalic veins, the IVC should be utilized. Percutaneous and surgical approaches to central venous access via the IVC have included cannulation of the saphenous (57,58), femoral (59–62), inferior epigastric (63–66), gonadal (67–69), iliac (70–72), lumbar (73), and hepatic veins (35,36,38).

Originally considered a less favorable approach owing to activity restriction and increased rates of catheter infection, femoral venous catheters are safe and have infection rates equal to that of SVC catheters, particularly when tunneled to the anterior thigh or abdominal wall. (61,62,74). The approach to placement of a femoral venous catheter is usually via a percutaneous technique, and this is especially useful in the neonatal population (59,75).

Access to the inferior epigastric vein is via the groin (63,65,66). This approach is indicated in cases where the femoral vein cannot be accessed because of the presence of soilage or open wounds. In addition, if the femoral vein cannot be percutaneously

cannulated and the saphenous vein has already been ligated, the inferior epigastric approach provides a good option to cannulation of the IVC. Access to the inferior epigastric vein is via a transverse inguinal incision and exposure of the internal inguinal ring, and the vein courses along the medial margin of the internal ring. The proximal catheter or reservoir can be tunneled up to the abdominal wall, which may lessen the risk of contamination and infection. In cases in which the femoral veins are inaccessible because of interruption or thrombus, catheterization of the iliac vein may be performed via a retroperitoneal approach (71). Retroperitoneal exposure of the iliac vein allows catheter placement by direct venipuncture or via venotomy without the need for vein ligation. The CVC is tunneled onto the chest wall for access. Similarly, the gonadal vein can be accessed via an oblique incision in the iliac fossa (67–69). Gonadal venous catheters are also usually tunneled onto the lower anterior chest wall. If the iliofemoral or lower IVC is inaccessible, percutaneous transhepatic or translumbar techniques should be considered as outlined previously.

If access to both the SVC and IVC is impossible because of vessel thrombosis, central venous catheterization may be accomplished via alternate SVC collaterals, such as the azygous (76), intercostal (39–42,77), and internal mammary veins (78). An additional, more invasive, option is direct right atrial cannulation (79,80). The traditional approach to azygous, hemiazygous, and intercostal vein catheterization is via thoracotomy, and the CVC is tunneled to the anterior chest. The advent of video-assisted thoracic surgery (VATS) has significantly improved these approaches, and has provided a minimally invasive technique for direct right atrial catheterization (36,80). The thoracoscopic approach to right atrial catheterization involves opening the pericardium, exposing the atrium, and placing the catheter over a guidewire. The catheter is secured by using a right atrial pursestring suture. Figure 5 depicts a proposed algorithm for CVC insertion

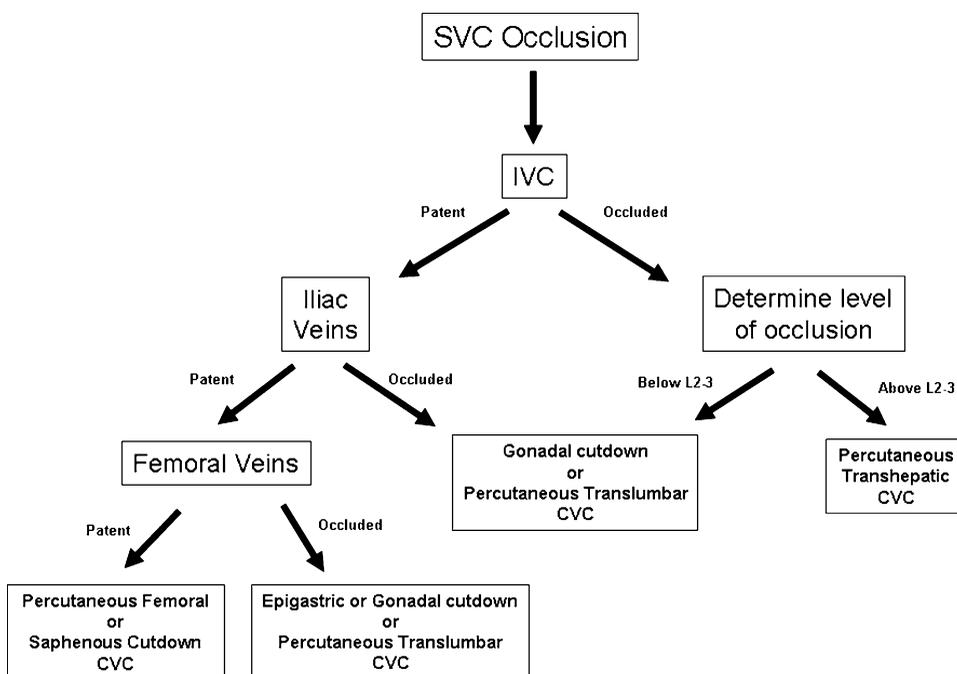


Fig. 5. Proposed algorithm for placement of CVC in patients with chronic occlusion of the SVC.

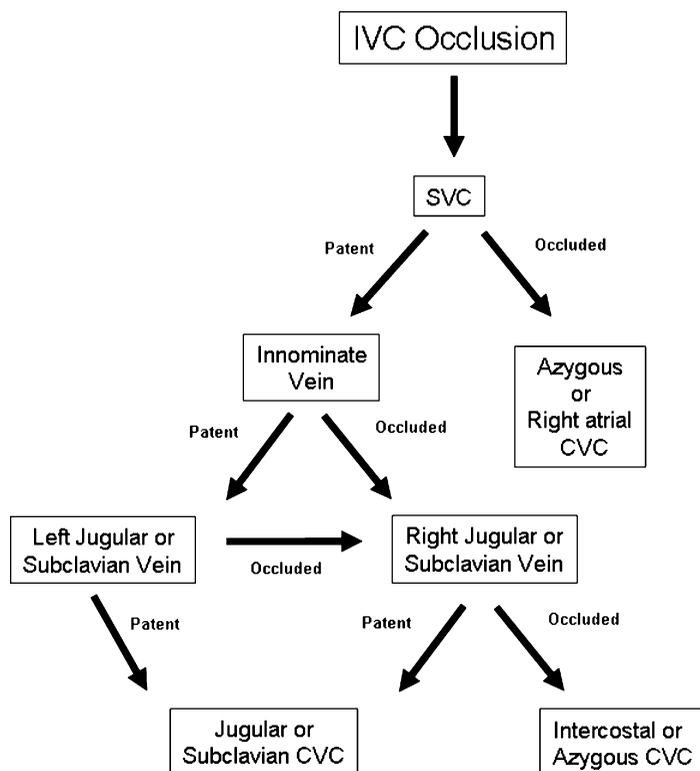


Fig. 6. Proposed algorithm for placement of CVC in patients with chronic occlusion of the IVC.

in the setting of chronic SVC occlusion, and Fig. 6 demonstrates an algorithm for CVC placement in patients with an IVC occlusion.

In conclusion, the establishment of central venous access in patients with a history of multiple previous CVCs and known venous thrombosis is a challenge. There are multiple options for CVC insertion in this difficult patient population, and approaches include both interventional and surgical techniques. In order to avoid exhausting central venous cannulation approaches, it is critical to limit the placement of CVCs as much as possible and to replace them only when absolutely necessary. In high-risk patients, thorough preoperative imaging will identify patent vessels, delineate a rational operative approach, and avoid technical misadventures during CVC insertion.

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3

Reoperative Head and Neck Surgery

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CONTENTS

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Operative procedures performed in the head and neck in children compose a significant element in pediatric surgical practice. There are a broad variety of these procedures, from correction of complex congenital anomalies to treatment of a variety of acquired lesions. The successful conduct of these operations requires a thorough knowledge of normal head and neck anatomy and precise attention to the technical details of the procedure. Inadequate or incomplete resection of congenital or acquired lesions can lead to recurrences, requiring secondary operative procedures. Inadvertent injury to surrounding structures can lead to life-threatening complications or permanent disability. This chapter will review the more common operative procedures performed in the head and neck in children and discuss their potential complications.

THYROGLOSSAL DUCT CYSTS

Thyroglossal duct remnants comprise one of the most common cystic neck masses requiring surgery in children. These lesions usually present as midline neck masses in the first decade of life. Their initial presentation may be as an infected midline cyst. Complications of surgery for thyroglossal duct cysts are uncommon and generally can be avoided by meticulous attention to operative technique during the initial procedure. Preoperative infection should be treated with intravenous antibiotics and surgery delayed until signs of infection have completely cleared. Attempts should be made to avoid incision and drainage of infected thyroglossal duct cysts, as this may result in spread of ductal epithelial cells to the surrounding tissue and increase the chance of recurrence after excision.

Thyroglossal duct cysts arise from tissue associated with the embryologic descent of the thyroid gland from the foramen cecum to its normal anatomic location at the

base of the neck. The thyroid gland has reached its final position in the neck by 7 weeks of gestation. The thyroglossal duct tissue should disappear by 8–10 weeks (1). The path of the thyroid's descent passes through the central portion of the hyoid bone, or immediately adjacent to this bone, anteriorly or posteriorly. Therefore, complete excision of thyroglossal duct remnants requires excision of the cyst itself as well as the midline tissue between the cyst and the foramen cecum, including the central 1 cm of the hyoid bone. Usually there is an evident tract, with or without a visible lumen, but occasionally this "tract" consists of multiple strands of ductal epithelial cells. Sistrunk, in addition to emphasizing the importance of resection of the central hyoid bone, also advocated resecting a wide core of tissue between the hyoid and the foramen cecum (2). Sade illustrated the frequent occurrence of multiple ducts in this portion of the tract, noting multiple tracts in 11 of 14 specimens (3).

Complications of thyroglossal duct cyst excision are listed in Table 1. The diagnosis of a thyroglossal duct cyst is usually a clinical one, with the finding of a midline mass located between the hyoid bone and the isthmus of the thyroid gland. Nonetheless, the preoperative diagnosis will be an error in 25–30% of the cases (4). Most of the erroneous diagnoses will prove to be epidermoid cysts, but some will be lymph nodes. Rarely, the midline lesion will be found to be median ectopic thyroid tissue. In the absence of either preoperative symptoms of hypothyroidism, or the inability to palpate the thyroid gland in its normal location, preoperative imaging with ultrasound or thyroid scans is unnecessary. Incisional infection should occur in less than 2–3% of patients. Generally, surgery should not be performed in the presence of active infection of the cyst. Patients with a history of prior infection should be treated with perioperative antibiotics, selected to cover oral flora.

Recurrent thyroglossal duct cysts should occur quite infrequently if a Sistrunk procedure was done as the original operation. Howard described 1.5% recurrences after a Sistrunk, and approximately 88% recurrences without removal of the hyoid (5). It is said that factors predisposing to recurrence include young patient age, direct skin involvement by the cyst, history of previous infections, lobulation of the cyst, rupture of the cyst at operation, and failure to adequately remove the central hyoid bone and the tissue between it and the foramen cecum. Most recurrences occur within 1 year of surgery, and they usually present as midline neck masses in the region of the hyoid bone. Recurrent cysts require reexcision. This operation should be planned to remove all of the midline neck tissue down to the pretracheal fascia from the isthmus of the thyroid gland to the foramen cecum. The original neck incision should be widely excised with the subcutaneous tissue beneath it. Inferiorly, the pyramidal lobe of the thyroid gland should be resected down to its union with the isthmus of the thyroid. The dissection should then proceed cephalad immediately on the pretracheal fascia, including approximately 1–2 cm of the midline strap muscles on either side of the midline. Complete excision of the recurrent cyst with the hyoid bone, including a wide

Table 1
Complications of Thyroglossal Duct Cyst Excision

-
1. Error in diagnosis
 2. Incisional infection
 3. Recurrence (requires reoperation)
-

excision of the fibrous tissue in the central portion, should be performed. A broad resection of the midline tissue between the hyoid and the foramen cecum should be taken *en bloc*. Occasionally, recurrent thyroglossal duct cysts will present lateral to the midline and may represent missed lobules of the original cyst. In these situations, careful reading of the previous operative note is important to help identify areas of difficult dissection. The lateral cyst should be widely excised *en bloc* with the midline neck tissue described previously. With such aggressive resection of recurrent cysts, second recurrences should be very uncommon.

CERVICAL DERMOID CYSTS

Dermoid cysts are congenital cysts that include both epidermis and epidermal elements, such as sebaceous glands and hair follicles. They generally occur in the lines of embryonic fusion in the anterior neck and are often closely associated with the hyoid bone (6). These lesions are often misdiagnosed preoperatively as thyroglossal duct cysts, because they occur in the midline and may move with swallowing. They often are adherent to the underside of the normal skin overlying them. They are not associated with tracks through the hyoid bone, however. Operative excision of these lesions should include any portion of attached normal epithelium in continuity with the cyst itself. Resection of those lesions attached to the hyoid bone should include removal of the central portion of the hyoid bone with the cyst.

Complications of excision of dermoid cysts requiring reoperation are primarily related to recurrence of the cysts (Table 2).

Factors associated with recurrence include intraoperative rupture of the cyst or incomplete excision of the cyst, either by leaving a portion attached to the overlying skin or to the hyoid bone. Recurrences should be excised with a generous ellipse of the skin around the primary incision and the resulting scar tissue from the primary excision, as well as the recurrent cyst. If the original cyst was attached to the hyoid bone, the entire hyoid should be resected with the recurrence. Second recurrences of these lesions have not been described.

BRANCHIAL CLEFT CYSTS AND SINUSES

Branchial cleft anomalies are some of the most common congenital conditions of the head and neck in children. These anomalies can form simple cysts or sinuses with fistulas from the posterior pharynx to the skin of the neck. Their specific diagnosis is usually not complex, but successful surgical therapy rests upon a thorough knowledge of the embryology and anatomy of the neck and precise surgical technique. First branchial cleft anomalies are associated with the external auditory canal, and may present with fistulous openings either anterior or posterior to the ear. The anterior fistulas lie in close proximity to the facial nerve. Second branchial cleft anomalies,

Table 2
Complications of Cervical Dermoid Cyst Excision

1. Error in diagnoses
 2. Incisional infection
 3. Recurrence (requires reoperation)
-

Table 3
Complications of Brachial Cleft Anomaly Excision

-
1. Infection
 2. Nerve or vascular damage
 3. Recurrence (requires reoperation)
-

which comprise more than 90% of these conditions, may form fistulas that open along the anterior border of the sternocleidomastoid muscle in the lower one-third of the neck or present as a lateral neck mass, usually a little higher than the fistula opening (7). In the Mayo Clinic series, 74% of the second branchial cleft anomalies presented as cysts (1). The internal opening is in the supratonsillar fossa, and the tract usually passes in close proximity to the bifurcation of the carotid artery. Third and fourth cleft anomalies are far less common, and they involve tracks in the superior mediastinum. Branchial anomalies with fistulas are usually diagnosed early in life, because of the small amount of mucoid drainage from the external opening. Branchial cysts, on the other hand, may not present until later childhood or adult life, being discovered as masses in the lateral neck.

The cardinal principal of surgery on branchial cleft anomalies is complete excision of the anomaly with excision of the tract onto the pharyngeal musculature, without injury to surrounding structures. Failure to excise the entire tract may result in recurrences (Table 3). With second cleft fistulas, a cephalad counter-incision may be necessary to achieve complete resection of the tract. Telander described a 6% recurrence rate in 208 patients operated on for branchial anomalies at the Mayo clinic (1). Factors associated with recurrence included a prior history of infection and a history of previous surgical intervention on the lesion.

Recurrence of branchial cleft cysts or fistulas requires reoperation. A preoperative computed tomography (CT) scan of the head and neck may help in defining the precise anatomy of the anomaly, often with contrast injection in an external opening, if present. Whether the recurrence presents as a cyst or a true fistula, the presumption must be that there is a persistent internal opening to the posterior pharynx. This connection must be identified, using operating loops, and suture obliterated at the pharyngeal wall. Similarly, recurrences of a first branchial cleft anomaly must be assumed to still have a connection with the external auditory canal, and this must be identified and closed. Recurrences of first cleft anomalies as cysts in the region of the facial nerve imply incomplete resection in this area at the initial operation. At the time of reexploration, the facial nerve should be identified as it exits the skull and all of its branches carefully identified before excising the recurrent cyst. A superficial parotidectomy may be necessary in some of these cases to allow complete exposure of the facial nerve during this dissection.

ATYPICAL MYCOBACTERIAL CERVICAL LYMPHADENITIS

Atypical or nontuberculous mycobacteria are currently the most common causative agents of mycobacterial lymphadenitis in children in developed countries (8). In immuno-competent hosts, this is considered to be a local infectious process, usually caused by *Mycobacterium avium* or *Mycobacterium intracellulare*. Although true tuberculous lymphadenitis (*Mycobacterium tuberculosis*) had, at one time, nearly

disappeared in the United States, it is currently being seen with increasing frequency in patients suffering with HIV/AIDS. Nonetheless, the vast majority of mycobacterial lymphadenitis in children in the United States remains caused by atypical organisms. These nodes usually occur in otherwise healthy young children between the ages of 1 and 5 years and are most commonly seen in the submandibular triangle (9,10). They are minimally tender and usually present with some discoloration of the overlying skin. They may present with draining sinuses, particularly if percutaneous aspiration has been previously employed. Chest radiographs are normal in these patients and standard purified protein derivative (PPD) skin testing is usually negative or shows only intermediate reactivity (9).

Unlike tuberculous lymphadenitis, atypical mycobacterial infections do not respond to standard antibiotics, and complete surgical excision is the treatment of choice. Because most of these lymph nodes are in the region of the facial nerve, this nerve and its branches are at risk for injury during the initial surgical dissection (Table 4). Full knowledge of the anatomy of the facial nerve and particularly the marginal mandibular branch is required prior to undertaking such a dissection. Care must be taken to specifically identify this nerve and prevent injury to it, even if a portion of the surrounding inflammation must be left adjacent to the nerve. In this case, careful curettage of this region may allow removal of the majority of the infected lymph node tissue with preservation of the nerve. Less than complete excision of the infected node leaves these children vulnerable to development of persistent infection and a chronic fistula through the incision. The surgical specimen should be submitted for pathology examination, with acid fast and fungal stains. Portions of the specimen should be cultured.

Persistent drainage through the operative incision following partial excision of an atypical mycobacterial lymphadenitis must be treated surgically. Sensitivity studies should be performed on the original organism isolated using isonicotinic hydrazide (INH), Rifampin, and ethambutol, as well as the quinolone antibiotics. Bailey has divided atypical tuberculosis organisms into those that are usually responsive to antibiotics and those that are not (11). If the organisms are found to be sensitive to any of these agents, they should be used for perioperative coverage and continued for several weeks postoperatively.

Reexcisional surgery significantly increases the risk of injury to the marginal mandibular branch of the facial nerve. The original skin incision, with the draining sinus tract, should be widely excised and a nerve stimulator used to aid in identifying the marginal mandibular nerve. This nerve should be first identified well proximal to the region of inflammation and dissection should proceed carefully along the nerve to elevate the infected tissue from it. As with the primary operative procedure, it is preferable to leave small fragments of infected lymph tissue on the nerve than to divide

Table 4
Complications of Atypical Mycobacterial Cervical
Lymph Node Excision

-
1. Facial nerve damage
 2. Recurrence (requires reoperation)
 3. Chronic sinus (requires reoperation)
-

the nerve during this dissection. It is generally not necessary to do a wide local nodal excision, although any surrounding lymph nodes adherent to the primary inflammatory mass should be resected with the specimen.

DROOLING

Persistent or uncontrollable drooling is an important clinical problem in between 10–40% of children with cerebral palsy. The presence of such drooling does not indicate an increase in basal salivary secretion, but rather an inadequate clearance of the saliva from the anterior oral cavity. Persistent drooling is a problem for personal hygiene in these patients and often results in maceration and infection of the skin of the face and neck. In addition, it may lead to social isolation of these children. Approximately 60–70% of resting salivary secretion comes from the submandibular glands. Only 20–30% is produced by the parotid glands, and the remainder comes from the sublingual salivary glands (12). Salivary secretion is stimulated by the cholinergic nervous system.

Treatment strategies for children with drooling involve both medical and surgical approaches. Medical approaches use anticholinergic medications such as the scopolamine patch or glycopyrrolate. The use of these medications for treatment of drooling in patients with neurologic deficits is an off-label use. Because these drugs are administered systemically, they have a very high incidence of complications, many of which result in cessation of usage. In most series, between 70–90% of these patients achieve early relief of their drooling, but 70% of the patients have significant side effects, such as xerostomia, restlessness, and blurred vision, and choose to terminate therapy (13). Because of the difficulties with systemic drug administration, more recently botulinum toxin-Type A has been employed for treatment of drooling. Botulinum toxin is injected into the submandibular glands bilaterally, under general anesthesia, with ultrasound control. Approximately 60% of patients respond to Botox injection with reduction in drooling, but this effect only lasts about 3 months. Inadvertent injection of Botox into the tissues surrounding the submandibular glands and diffusion of the material into the surrounding tissues has been implicated in the production of dysphagia in approximately 6% of these patients (14). Attempts at retrograde injection of the Botox through the parotid and submandibular ducts have resulted in a greater percentage of painful swelling and dysphagia in these patients and appears to be poorly tolerated (15).

The surgical principles for treatment of drooling involve attempts to diminish the volume of secretion of saliva or redirect the normal flow of saliva to the posterior pharynx. Submandibular gland excision has been employed with satisfactory relief of drooling in the majority of the patients, but this procedure puts both the marginal mandibular branch of the facial nerve and the lingual nerve at risk (Table 5). Resection

Table 5
Complications of Operations for Drooling

-
1. Failure (requires further therapy)
 2. Dysphagia
 3. Loss of taste
 4. Ranula formation (requires reoperation)
 5. Facial/lingual nerve damage
-

of the parotid gland is a more difficult technical procedure, placing the proximal facial nerve at risk. Attempts to ligate the parotid and submandibular gland ducts intraorally have been successful in a small number of children (16).

The parotid, submandibular, and sublingual glands receive their parasympathetic intervention by nerves that can be exposed in the middle ear. The parotid gland is served by the tympanic nerve, on the medial wall of the middle ear, and the submandibular and sublingual glands are innervated by the chorda tympani nerve, which crosses the midportion of the middle ear. Both of these nerves can be sectioned by reflecting the tympanic membrane under local anesthesia. Relief from drooling is accomplished in approximately 60% of these patients. Complications of this procedure include inability to access the nerves. Section of the chorda tympani nerve results in loss of taste in the anterior two-thirds of the tongue, a complication that is apparently not troublesome in this patient population. There is at least a theoretic possibility of loss of hearing from any procedure performed within the middle ear, but this has not been seen clinically (17).

Recently, a new operation has been evaluated for children with troublesome drooling. In this procedure, the submandibular ducts are transposed to the posterior pharynx, into the tonsillar fossa (18). This operation is occasionally accomplished with simultaneous excision of the sublingual glands, which can be performed from within the mouth. Crysdale reported results of submandibular duct transposition in a group of 194 patients, 80% of whom were less than 18 years of age. Of these patients, 67% had a good or excellent outcome, with a poor result in only 11% (19). Ranula formation was a common complication, and 13% of the patients underwent subsequent sublingual gland excision. The authors have subsequently added sublingual gland resection as a concomitant procedure with submandibular duct transposition.

By far the most common complication of all of these techniques for treatment of drooling in neurologically impaired children has been failure of the techniques to eliminate or improve drooling. This has led to a sequence of procedures in many patients, such as proceeding to submandibular gland excision after failure of duct transposition, addition of section of the chorda tympani nerve after duct transposition and injection of botulinum toxin or institution of drug therapy after failure of any of these procedures. Reoperative procedures all carry a high risk of morbidity and probably should be used as a last resort.

TRACHEOSTOMY

The frequency of use of tracheostomy for airway management in infants and children has declined significantly in the past two decades. Modern neonatal respiratory care has greatly reduced the incidence of acquired subglottic stenosis, as the importance of using relatively small endotracheal tubes has been widely accepted. Currently the three most common indications for tracheostomy in children include: (1) airway obstruction from congenital or acquired conditions; (2) need for ventilatory support (often in children with generalized neuromuscular disorders); and (3) need for pulmonary toilet in patients with a poor or inefficient cough.

The most common complications of tracheostomies are listed in Table 6. Overall, complications arising from tracheostomies in children are estimated to occur in between 20–80% of patients (20). The perioperative complications should be avoided by meticulous attention to the details of the tracheostomy operative procedure. Late complications are minimized by careful surveillance (21).

Table 6
Complications of Tracheostomy in Children

A. Perioperative complications

1. Pneumothorax
2. Bleeding
3. Accidental decannulation
4. Mucus plugging

B. Late complications

1. Accidental decannulation
 2. Granulation tissue formation (requires reoperation)
 3. Tracheal (stomal) stenosis (requires reoperation)
 4. Innominate artery erosion (requires reoperation)
-

Tracheostomies in children should virtually all be performed over a stable airway, usually maintained with an endotracheal tube. The patient's neck should be placed in gentle extension and a transverse skin line incision made in the lower neck. Dissection on either side of the trachea in this region may violate the parietal pleura, which ascends well into the lower neck in a child, resulting in a pneumothorax. The strap muscles are divided in the midline, taking care to assure that the trachea is immediately beneath this. Meticulous hemostasis is required, as there are frequently multiple anterior thyroid veins in this region, which are the source of most immediate postoperative bleeding. When the anterior wall of the trachea is identified, dissection should proceed cephalad to identify the thyroid isthmus. This is bluntly dissected from the anterior tracheal wall and reflected superiorly. The thyroid isthmus crosses at the second tracheal cartilage and provides an important landmark for the tracheotomy. The second cartilage is elevated with a small skin hook and a vertical tracheotomy performed through the third, fourth, and occasionally fifth cartilages. Fine Prolene sutures are placed through the fourth cartilage bilaterally for traction to open the tracheotomy. At this point, an appropriately sized tracheostomy tube should be selected, considering both the diameter and the length of the tube. The endotracheal tube is slowly withdrawn until the tip of the tube is just cephalad to the tracheotomy, and the tracheostomy tube is inserted. When it is assured that the tracheostomy tube is in the trachea, the retractors are removed and the tube is secured to the neck with cotton tapes, tied tightly with a knot. The wings of the tracheostomy tube are sutured to the neck to avoid accidental decannulation. The incision is not closed around the tracheostomy tube, to avoid local infection and pneumomediastinum. The patient is cared for in a monitored setting, with minimal manipulation of the tracheostomy tube for the first 5–7 days postoperatively. At this time, the tube is changed by the operating surgeon, and if a secure tract has been formed, the tracheostomy ties can be loosened and the patient moved to a conventional bed.

Accidental decannulation is always a potential complication of a tracheostomy in a child. Until a stable tract has formed, the tube can retract out of the tracheal lumen and into the mediastinum without being noticed externally. With any episode of significant desaturation or loss of breath sounds, decannulation must be suspected. Within the first few days of the operation, blind reinsertion of the tracheostomy tube may be difficult, even with the traction sutures placed with the original procedure. Adequate retractors and light will be necessary, and one should merely reintubate most patients

by mouth before proceeding to try to reinsert the tracheostomy tube. After 5–7 days postoperatively, reinsertion of the tube should be straightforward.

Obstruction of the tracheostomy cannula with mucus plugs occurs more commonly in children than in adults because of the small size of the tracheostomy tubes used and the lack of an inner cannula. The parents are taught to change the tracheostomy tube on a weekly basis at home, but more frequent changes may be necessary when the patients have increased secretions. The use of humidified air may reduce the incidence of plugging.

Formation of granulation tissues around the tracheostomy tube is a uniform finding in children with tracheostomies. This tissue forms in response to irritation from the foreign body of the tracheostomy tube itself and occasionally can be exuberant enough to cause a foul-smelling exudate. Exuberant granulations can also interfere with the changing of the tracheostomy tube and, in this case, they must be excised or cauterized. Granulations also form on the inside of the airway, usually on the proximal border of the tracheostomy stoma and at the tip of the tube. Children with tracheostomies should undergo bronchoscopy every 6 months and the internal granulation tissue should be removed, before it forms a fibrous stricture of the airway.

Significant erosions of the trachea are less commonly seen with the flexible silastic tracheostomy tubes currently employed, compared to the metallic tubes that were employed years ago. Nonetheless, anterior erosion into the innominate artery is still occasionally encountered. Tracheoinnominate artery fistulas occur from erosion of the anterior tracheal wall caused by pressure at the tip of the tracheostomy tube or at the level of the cuff. Usually there is some indication of a problem, with sentinel bleeding as an anterior ulcer is created (22). Patients with tracheostomies in whom more than 10 mL of blood is seen in the tracheal aspirate, in the absence of any obvious cause, such as tracheitis, should undergo fiberoptic bronchoscopy to look for an anterior ulcer (23). If a superficial ulcer is encountered, the length of the tracheostomy tube may be changed to change the point of impact of the tip of the tube with the tracheal mucosa. Actual innominate artery erosion presents with massive bleeding through the tracheostomy tube and usually is fatal, unless the patient is in a hospital environment. If in such an environment, the tracheostomy tube should quickly be removed and replaced with a cuffed endotracheal tube. That tube is positioned so that the cuff tamponades the bleeding. If this is unsuccessful in stopping the hemorrhage, an index finger may be placed through the tracheostomy stoma and the artery compressed against the sternum (Utley maneuver) (24). The patient should then be rapidly transported to the operating room for a thoracotomy, either by median sternotomy or right anterior third interspace thoracotomy. Proximal control of the innominate artery should be achieved initially, and then distal control of the right subclavian and right common carotid arteries individually. The innominate vessel should then be dissected from the anterior wall of the trachea. The balloon on the endotracheal tube can then be deflated. The margin of the tracheal fistula should be débrided and the fistula closed with interrupted absorbable sutures, placed around adjacent cartilages. The innominate vessel is ligated along with the right carotid and right subclavian vessels. At the completion of the thoracotomy, the endotracheal tube may be removed from the trachea and replaced with an appropriately sized tracheostomy tube. Reconstruction of the innominate artery is rarely required in pediatric patients, but, if it is thought to be necessary, the procedure should be delayed until the mediastinal infection is completely eliminated.

Table 7
Criteria for Anterior Laryngotracheal Decompression

-
1. Failure of extubation on two occasions secondary to subglottic pathology
 2. Weight greater than 1500 g
 3. Absence of ventilator support for 10 days
 4. FI02 less than 30%
 5. Absence of congestive heart failure for 30 days
 6. Absence of acute respiratory tract infection
 7. Absence of antihypertensive medication for 10 days
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Percutaneous dilatational tracheostomy has become quite popular in adult practice since its original description in 1985 (25). There have been very few reports of the use of this technique in children, and the manufacturer's literature specifically indicates that the device is not to be used in pediatric patients (26). The most significant complications of this procedure encountered in adults have been creation of a tracheoesophageal fistula by inadvertent penetration of the posterior wall of the trachea and anterior wall of the esophagus and tracheal stenosis, usually thought to be secondary to distortion and collapse of the cartilage on the proximal border of the stoma. Significant stomal stenosis has been described in two children undergoing percutaneous dilatational tracheostomy, one of whom required a local tracheal resection and the other of whom was successfully treated with laser excision.

Certain forms of acquired and congenital subglottic stenosis in the neonate may be treated by an anterior laryngotracheal decompression (anterior cricoid split), eliminating the need for a tracheostomy. The cricoid split is successful in relieving the airway obstruction in approximately 70% of carefully selected patients (27). Cotton, who initially described the procedure, suggests the criteria for performing anterior cricoid split listed in Table 7 (28). Failure of the cricoid split to relieve subglottic airway obstruction usually indicates a more extensive subglottic stenosis than initially appreciated. Significant circumferential stenosis at the level of the cricoid may not respond to this simple procedure. Failure may also be secondary to airway pathology outside of the subglottic region, usually laryngomalacia. With failure of a cricoid split the patient should receive a tracheostomy, inserted below the level of the split.

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Reoperative Thyroid and Parathyroid Surgery

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REOPERATIVE THYROID SURGERY

Reoperative thyroid surgery is a difficult undertaking, with both diagnostic and technical challenges. Ultimately, a better understanding of the pathophysiology of thyroid disease, advances in preoperative diagnosis and management, and more thorough and evidenced-based resections of malignant disease may eliminate the need for reoperation. In the interim, it is important to understand thyroid diseases associated with reoperation, specific indications for reoperation, options for preoperative work-up, and common complications of reoperation.

Thyroid malignancies account for 3% of all pediatric malignancies, and are the most frequent indication for thyroid surgery in children (1,2). Unlike the adult population, thyroid nodules in children are found to be malignant in 20% of cases. Whereas benign diseases are more prevalent in the pediatric population than malignant disease, surgical resection is rarely used in their treatment. Surgical resection is reserved for very specific indications in benign pediatric thyroid disease, including recurrent acute suppurative thyroiditis (for which a thyroid lobectomy is sometimes indicated); Graves' disease after failure of both medical management and abalative therapy; and airway compression resulting from an enlarged thyroid in Graves' disease (2).

Malignant thyroid neoplasms are the most common reason for thyroidectomy in children. Although thyroid malignancies are less common in children than adults (only 10% of all thyroid malignancies occur in patients under age 21), they are one of the more common carcinomas in the pediatric age group. The annual incidence of primary thyroid malignancies in childhood had steadily increased from the 1960s until the mid-1980s, when the link between thyroid radiation and carcinoma led to the more conservative use of radiation therapy in benign disease (2,3). More recently,

however, the increased incidence of thyroid cancer after prior treatment for other childhood malignancies (especially leukemia and lymphoma) has become evident. These secondary thyroid malignancies (STMs) result from the carcinogenic effect of radiation therapy, the sequelae of immune suppression, and the oncogenic potential of chemotherapy (4). Thyroid malignancies account for 9% of all secondary malignancies.

Most childhood thyroid malignancies (70%) present between the ages of 11–17. The most commonly encountered pediatric thyroid malignancy is papillary thyroid carcinoma, followed by follicular carcinoma, then medullary thyroid cancer (MTC). Papillary thyroid cancer is the most common malignancy among both primary thyroid malignancies and STMs. However, primary thyroid malignancies in children tend to present in a more advanced state than either STMs in children or primary thyroid malignancies in adults. Papillary thyroid cancer tends to be multicentric, spreading early to regional nodes. In most case series, more than 70% of children present with lymph node metastasis at the time of diagnosis (4). Also, larger studies have reported rates of distant metastasis at diagnosis that range from 19–25% (5). The most common site of distant metastasis is the lung. Despite the high incidence of nodal and distant metastasis, the prognosis of papillary thyroid cancer in children is quite favorable. The overall survival rate in children for both papillary thyroid cancer and the more aggressive but less common follicular thyroid cancer is 98% (6).

Also uncommon but much more menacing is MTC, a malignancy of parafollicular C cells that is responsible for 2–5% of pediatric thyroid tumors. MTC may arise spontaneously or as part of a hereditary syndrome including multiple endocrine neoplasia (MEN) type 2A, MEN type 2B, or familial medullary thyroid cancer (FMTC). Not only is MTC often the first presenting tumor in patients with MEN, but it is also the most common cause of death from the disease. Because MTC tends to be lethal, aggressive resection including central compartment lymph node dissection from the thyroid to superior mediastinum and laterally to the carotid sheath is indicated. Fortunately, new techniques have led to the increasingly early identification of MTC in hereditary disorders. Greater attention to MEN disorders in families and screening for receptor tyrosine kinase (RET) protooncogene mutations has allowed for both the identification of hereditary MTC tumors at a smaller and less advanced stage than their sporadic counterparts and for the prevention of MTC through prophylactic thyroidectomy. Genetic testing should begin by age 5 for children with a family history of MEN 2A and as soon after birth as possible for MEN 2B. In children carrying the RET protooncogene mutation, prophylactic elective total thyroidectomy has been shown to dramatically decrease the development of MTC and, given relatively low associated morbidity, is universally recommended for patients with MEN 2A and 2B (7). Although larger studies with longer follow-up periods are needed for establishing definitive guidelines on the surgical treatment of children with MEN 2A and MEN 2B, current data support prophylactic total thyroidectomy prior at or before age 5 in MEN 2A and prior to 1 year of age in MEN 2B (6–9). Other thyroid tumors such as lymphomas, sarcomas, and undifferentiated carcinomas have been reported but rarely occur in children.

The optimal initial resection for thyroid malignancies has remained controversial in the absence of definitive data. Surgical options include total thyroidectomy (complete removal of the thyroid gland), near total thyroidectomy (leaving less than 3–5 g of residual thyroid parenchyma), subtotal thyroidectomy (leaving more than 5 g of residual thyroid parenchyma), and lobectomy plus isthusectomy. An emerging consensus supports total or near total thyroidectomy as the initial resection of choice

for differentiated thyroid malignancies, although some surgeons still perform a more limited resection. Support for total or near total thyroidectomy includes data showing the high rate of occurrence of occult, bilobar malignancies in the pediatric population (as many as 80% of affected children) (10). Also, total or near total thyroidectomy aids in postoperative management by maximizing uptake in any remaining thyroid deposits on postoperative imaging for suspected recurrence, and making subsequent radioiodine ablative therapy more efficient. Still, some argue for less aggressive surgical resection, citing reports showing no statistically significant difference in outcomes between the two approaches (using the need for postoperative radioactive Iodine uptake and the need for further ablative therapy as outcome measures) (11). Also, the favorable prognosis and long life span in children with advanced disease at presentation and the increased risk of recurrent laryngeal nerve injury reported in studies prior to 1990 are used to argue for a more limited resection (12). Despite concerns that more aggressive resections lead to higher rates of complications, especially recurrent laryngeal nerve injury, no significant differences in morbidity occurred in lobectomy versus total/near total thyroidectomy in adult studies (13,14). Because more definitive data in the pediatric population is needed, the safest approach may be a near total thyroidectomy with postoperative I-131 remnant ablation.

Another question surrounding initial resection is the extent of lymph node resection. The Society of Surgical Oncology practice guidelines for major cancer centers recommend total thyroidectomy for thyroid malignancies in adults but does not recommend routine lymph node dissection unless clinically palpable nodes are evident (8). However, given the different behavior of the disease in children, there is a growing consensus in the pediatric literature that the standard initial surgical approach to papillary thyroid carcinoma should be a near total or total thyroidectomy with resection of involved lymph nodes followed by radioiodine therapy.

Specific Indications for Reoperative Thyroid Surgery

Unfortunately, reoperative surgery is often unavoidable in the management of pediatric thyroid malignancies. Although scar tissue formation, inflammation, edema, and distortion of landmarks and tissue planes complicate any reoperative neck procedure, reoperation is indicated for disease recurrence, incomplete excision, and the discovery of previously unsuspected carcinoma following an initial lobectomy for what was thought to be benign disease.

Completion thyroidectomy is necessary in cases of less than near total resection on initial operation or disease recurrence in remaining thyroid tissue. In any resection less than a total thyroidectomy, the possibility of finding unsuspected thyroid carcinoma on permanent section exists. Despite improvements in fine needle aspiration (FNA) and intraoperative frozen section, nodules initially thought to be benign may not be recognized as malignant until examined by permanent pathology. As discussed previously, although total thyroidectomy is advocated by most as standard treatment for well-differentiated thyroid malignancies, some surgeons perform subtotal thyroidectomy as an initial operation, necessitating a completion thyroidectomy for persistent or recurrent disease.

The more common indication for reoperation, however, is disease recurrence. The high rate of recurrence of thyroid malignancies in children makes a thorough preoperative evaluation and postoperative follow-up imperative, especially in children with advanced disease. Papillary thyroid cancer recurrence rates in children ranges from

35–45% (with some reports as high as 47%) (11), with the median time to the development of a clinically evident recurrence averaging 7 months. Recurrence is more common in patients with lymph node involvement at presentation (as high as 69% in one recent study, although other studies have shown a weaker relationship) and patients with multiple nodules by physical exam on initial presentation. Age, sex, fixation of nodules, tenderness, family history, and hyperthyroidism have not been shown to be associated with increased risk of recurrence. And, although recurrence rates in children are high for papillary thyroid cancer, recurrence does not seem to confer an increased risk of mortality in this age group (11). To lessen the need for reoperation, much of the literature supports a strategy of careful preoperative inspection for nodal metastasis on preoperative evaluation prior to initial resection, as well as carrying out a meticulous regional lymph node dissection (11,15).

Exactly what constitutes a meticulous or at least adequate lymph node dissection is not always clear. In studies examining patterns of nodal metastases, papillary thyroid cancer usually metastasizes to paratracheal and paraesophageal nodes as well as the ipsilateral cervical and jugular lymph nodes. Recommendations for lymph node dissection in the initial resection of papillary thyroid cancer include a formal modified regional neck dissection to remove pretracheal and paratracheal nodes as well as ipsilateral upper, middle, and lower jugular nodes and the recurrent laryngeal nerve chain (11). In MTCs, the additional resection of contralateral upper, middle, and lower jugular nodes and the recurrent laryngeal nerve chain is indicated given the higher incidence of contralateral node involvement.

It is important to note that some “recurrent” nodal metastases are actually present at the initial operation but missed intraoperatively. In at least one study, fewer than 50% of papillary thyroid cancer nodal metastases and fewer than 64% of MTC nodal metastases were correctly identified by experienced surgeons intraoperatively (8). Therefore, a systematic and meticulous approach to lymph node dissection may be of benefit in identifying nodal metastases and may lessen the need for reoperation.

Preoperative Assessment

Preoperative evaluation techniques can be valuable for both the diagnosis and localization of a suspected thyroid malignancy or recurrence. FNA is the clear first step for the diagnosis of solid thyroid nodules in adults, but its usefulness in the pediatric population is less clear. Given the high incidence of malignancy in children under age 13 (20%) and an FNA false negative rate of 6%, the current recommendations call for the surgical removal of all thyroid nodules found in this age group (6). In the adolescent age group, the incidence of malignancy is low enough (11%) and the safety of nonoperative management of benign lesions well documented enough that FNA is a useful study. Nodules proven benign by FNA can be followed clinically with serial physical exam and serial ultrasound in this age group. Radioactive thyroid scans with technetium-99 or iodine-123 are also useful for identifying hyperfunctioning nodules, which are usually benign.

Advances in preoperative imaging techniques have lead to the detection and localization of new nodules and cancer recurrences at an earlier stage. High-resolution ultrasound, contrast-enhanced magnetic resonance imaging (MRI), and positron emission tomography (PET) scanning can identify nodules smaller than 1 cm and have given surgeons both the ability and responsibility to identify and resect nodules that cannot be readily identified by palpation. Precise preoperative localization of these subclinical

nodules is especially important in preparation for reoperation in the central neck near the trachea, where adhesions complicate visual identification of small neoplasms (16). Also, preoperative evaluation for metastasis prior to reoperation for suspected or recurrent malignancy is important. A chest radiograph is a routine part of the work-up prior to initial resection of thyroid cancer, but may miss 30–40% of pulmonary metastases (5). The whole body radioiodine scan recommended for all children after initial resection is also useful as a preoperative study prior to reoperation to detect pulmonary metastases missed by chest radiograph. Alternatively, a chest computed tomography (CT) scan can be obtained, but care should be taken to avoid the use of iodinated intravenous contrast agents, which complicate subsequent treatment with radioiodine.

Operative Approach

After a thorough preoperative evaluation, including review of previous pathology and operative reports, and discussion of the risks and benefits of reoperation with the patient and family, care should be taken in planning the reoperative procedure. The timing of reoperation is important. If not undertaken during the first week after initial resection, surgery should not be attempted until 6–8 weeks after the initial operation to allow the acute inflammation to subside (17). The operative approach for a completion thyroidectomy or resection of residual thyroid tissue is similar to the initial resection. Care should be taken to maintain hemostasis, to identify and preserve the recurrent laryngeal nerve and the external branch of the superior laryngeal nerve, and to preserve the parathyroid glands and their blood supply.

After preparation of the patient with the head and neck extended, an incision is made above the sternal notch transversing the distance between the sternocleidomastoid muscles. Platysmal flaps are raised. Approaching the thyroid laterally through the undisturbed plane on the medial side of the sternocleidomastoid with retraction of the muscle laterally allows exposure of the lateral paratracheal space in a plane minimally disturbed by previous dissection (18). The sternohyoid muscle is reflected medially with the carotid sheath reflected laterally. Once the recurrent laryngeal nerve is carefully identified, the strap muscles are divided. Identifying the nerve in a previously undissected area and then following it into the reoperative field is the preferred strategy, because the loss of normal landmarks and tissue planes in the reoperative neck may complicate identification. Another approach used in reoperative surgery is the low anterior approach, which uses the same anterior approach as in the initial resection with lateral retraction of the strap muscles after dividing them at the midline. The recurrent laryngeal nerve is identified low in the neck where previous dissection is minimal and followed into the field (18).

It is important to note that the nerve is most often injured at the Ligament of Berry (the attachment of the thyroid gland at the trachea). Also, there is a great deal of variation in the course of the recurrent laryngeal nerve and thyroid artery and its branches. One important variation is the presence of a nonrecurrent laryngeal nerve, which occurs in 1% of patients, most commonly arising directly from the vagus nerve on the right. Use of intraoperative laryngeal nerve monitoring is helpful in tracing the course of the nerve, and has been shown to be both safe and beneficial in children (19). Extracapsular dissection of the thyroid with preservation of parathyroid fat minimizes the risk of permanent hypoparathyroidism. Also, resected thyroid tissue should be inspected after removal for presence of parathyroid tissue. If a parathyroid gland is

inadvertently removed during resection, or removed *en bloc* with tumor, it should be carefully separated from the specimen and autotransplanted in the sternocleidomastoid muscle or the forearm (once confirmation that it is uninvolved with the tumor is obtained).

Because patterns of lymph node metastases differ between the initial and reoperative patient populations in thyroid malignancies, the recommended lymph node dissections are a bit more extensive. For reoperative papillary thyroid cancers, removal of pretracheal and paratracheal nodes as well as both contralateral and ipsilateral upper, middle, and lower jugular nodes and the recurrent laryngeal nerve chains is advised, because a contralateral lymph node involvement is more common in these patients (11). For reoperative MTC, resection includes all central compartment lymph node nodes from the hyoid to superior mediastinum and laterally to the ipsilateral and contralateral carotid sheath, regardless of the size of the tumor (8). In these extensive lymph node dissections, the removal of multiple lymph nodes in the setting of fibrosis and scarring complicates identification of the parathyroid glands and their vascular pedicles. Again, tissue removed should be inspected for the presence of parathyroid glands, which can then be salvaged and autotransplanted. Most metastatic nodes can be removed through the cervical incision. However, thyroid cancer can rarely metastasize to the aortic pulmonary window necessitating a median sternotomy.

Complications

It is little surprise that uniformly higher complication rates are seen in reoperative thyroid surgery than with initial resection (11,15,17,20). The most common complications of thyroid procedures include recurrent laryngeal nerve injuries, transient hypoparathyroidism, and permanent hypoparathyroidism.

With careful identification of the recurrent laryngeal nerve and use of intraoperative neurological nerve monitoring, rates of transient recurrent laryngeal nerve injury have been kept to 2–5% with a permanent vocal cord paralysis rate of 1–5% (15,17). In fact, a few smaller studies report no recurrent laryngeal nerve injuries among their reoperative patients (15).

Pediatric patients have approximately the same incidence of transient hypocalcemia as adults (23%) (21). Careful identification and preservation of the parathyroids help reduce the incidence of this complication. If parathyroids are inadvertently or intentionally removed with the specimen, one or more glands should be autotransplanted to the sternocleidomastoid muscle or the forearm to reduce the incidence of hypocalcemia. Although data on reoperative complications in children is limited, the rate of permanent hypocalcemia among adults after reoperation is approximately 1% among experienced endocrine surgeons.

Postoperative Care

Caring for patients with thyroid malignancies or benign thyroid disease does not end after surgical resection. Follow-up care is imperative postoperatively, especially given the high rate of recurrence. Ablative radioiodine therapy is a recommended adjunct postoperatively and facilitates the use of subsequent thyroglobulin level monitoring for recurrence. A whole-body radioiodine scan is recommended at 6 weeks postoperatively, and should be followed by a therapeutic dose of radionuclide to ablate residual tissue and treat residual metastatic disease. Special attention should be given to searching for

possible pulmonary metastases. In reoperative patients, monitoring for postoperative complications is especially important given the difficulty of the procedure. Pediatric surgeons must also be prepared to coordinate patient care with a pediatric endocrinologist or pediatric oncologist for benign or malignant disease, respectively.

REOPERATIVE PARATHYROID SURGERY (RPS)

As with reoperative thyroid surgery, RPS is a complicated undertaking that should be entered into with care and meticulous planning. One reason for exercising extra caution is the difference in outcomes between RPS and primary resection. Although primary parathyroid operations have a high success rate with low associated morbidity (at least one large study cites cure rates of 99.5% and a permanent morbidity of less than 1%) (22), reoperative procedures have a lower cure rate with a higher rate of associated morbidity (a 90% cure rate but a rate of at least transient hypocalcemia that is as high as 35–43%) (22). Understanding the etiologies of recurrent or persistent hyperparathyroidism, the causes of failure after initial resection, and the preparation for reexploration is crucial for safe and successful reoperation.

Indications for Reoperative Surgery

Primary hyperparathyroidism usually results from a single adenoma, with initial surgical resection curative in 95% of these cases. Most parathyroid adenomas in children occur in adolescence, with only 12–16% of cases occurring in children under age 10. However, as many as 30–50% of children presenting with primary hyperparathyroidism have an etiology other than a solitary adenoma (23). They, along with virtually all infants with hyperparathyroidism have parathyroid hyperplasia related to an MEN syndrome or isolated familial hyperparathyroidism. The MEN syndromes involving hyperparathyroidism include MEN 1 (parathyroid hyperplasia, pituitary adenoma, pancreatic islet cell neoplasms) and MEN 2A (MTC, pheochromocytoma, and hyperparathyroidism). Isolated familial hyperparathyroidism (also called non-MEN familial hyperparathyroidism) describes a hereditary syndrome of positive family history of hyperparathyroidism, adenomatous changes, and absence of other MEN features. A subset of patients will have primary hyperparathyroidism from four gland hyperplasia, a double adenoma, or, more rarely, carcinoma. The initial approach to surgical treatment varies based on the etiology of the disease. Carcinomas and adenomas are approached initially with exploration and removal of the abnormal gland. However, if parathyroid hyperplasia consistent with a MEN syndrome or isolated familial hyperparathyroidism is encountered, the preferred treatment is either a subtotal parathyroidectomy or total parathyroidectomy with autotransplant of one gland into the sternocleidomastoid muscle.

Reoperation is indicated for persistent hyperparathyroidism (biochemical evidence of hyperparathyroidism immediately postoperatively or in the following 6 months) or recurrent disease (disease that develops more than 6 months after initial resection). Although only 3.2% of adults experience persistent disease and less than 1% have recurrent disease after initial resection, the rate of failed primary operation approaches 20% in the pediatric population (23,24). Most commonly, persistence or recurrence relates to failure of the initial exploration. Given the potential for serious complications in RPS, much attention has been given to examining the reasons for failure

of initial exploration and to developing strategies to increase the success of initial operations (9).

The failures of primary exploration are most often from an abnormal gland residing in a normal anatomic location (accounting for as many as 76% of failed primary explorations) (22). The abnormal gland may be adenomatous or simply a hyperplastic gland not recognized during the initial operation. In fact, as many as 37% of patients undergoing reoperation for persistent primary hyperparathyroidism are found to have hyperplastic glands compared to 15% of patients on initial operation—an indication that unrecognized hyperplasia may be a significant cause of disease recurrence and reoperation. Persistent hyperparathyroidism is quite frequent in patients with MEN 1, MEN 2A, or isolated familial hyperparathyroidism (recurrence rates of 20–100% have been described depending on the duration of follow-up) (25). These diagnoses should be entertained in cases of persistent hyperparathyroidism in children.

A substantial number of initial parathyroid resection failures may also be attributed to glands that do not reside in normal anatomic locations or supernumerary glands. A fifth parathyroid gland is identified in 15% of reoperative patients. For extraanatomic glands, the most common location is the mediastinum followed by intrathyroid glands, carotid sheath glands, anterior tracheal glands, and retroesophageal glands. To reduce the need for reoperation, careful preoperative localization prior to initial resection should be undertaken. An additional rare but significant cause of persistent or recurrent hyperparathyroidism is “parathyromatosis,” or the implantation of adenomatous or hyperplastic parathyroid cells after rupture of parathyroid capsule on initial operation. Because the cells may be disseminated, this complication is notoriously difficult to treat.

Parathyroid carcinoma is extremely rare but has been described in children as young as 12 years. After initial operation for parathyroid carcinoma, residual disease or metastasis may manifest as persistent or recurrent primary hyperparathyroidism. Unfortunately, the disease recurs in 40–70% of patients, with most recurrences occurring at the original site of the malignancy. When metastases are present, the most common sites are regional lymph nodes, bone, lung, and liver. The timing of recurrences is difficult to predict, with some occurring 1 month and some decades after initial resection. Multiple recurrences requiring repeated reoperations are not uncommon (26,27).

Secondary hyperparathyroidism results from multigland hyperplasia caused by end-stage renal disease. As in MEN or isolated familial hyperparathyroidism, the initial resection involves either a subtotal parathyroidectomy (three-and-a-half glands removed) or a total parathyroidectomy with autoimplantation of parathyroid tissue into the brachialis muscle of the forearm. Unlike primary hyperparathyroidism, preoperative imaging and localization is of less benefit prior to initial resection. In a comparison of recurrence rates for secondary hyperparathyroidism after each of the two procedures, a recent metaanalysis found significantly higher recurrence rate (49%) after total parathyroidectomy with autotransplantation than after subtotal parathyroidectomy (17%) (28). However, other sources document much lower and more equal rates of recurrence (10%) for both procedures, especially in areas with high rates of renal transplantation (28). Failure of the initial operation is attributed most often to autograft hyperplasia (49% of reoperative patients), followed by missed supernumerary glands (20%, the majority of missed glands were intrathyroid), remnant hyperplasia (17%) and missed *in situ* hyperplastic gland (7%) (28).

Preoperative Evaluation

Because of high the rate of morbidity, indications for reoperative parathyroid resection are more conservative than for the initial operation. For persistent or recurrent primary hyperparathyroidism, the diagnosis of hyperparathyroidism must be reestablished biochemically, with care taken to rule out other potential causes such as familial hypocalciuric hypercalcemia. Elevated levels of both serum calcium and intact parathyroid hormone (iPTH) should be documented. A careful history and physical should be targeted at excluding other diagnoses including radiation, lithium exposure, or other malignancy (although those most associated with hypercalcemia—lung, ovarian, and pancreatic cancers—are rare in the pediatric population). A review of initial operative notes and pathology is also prudent prior to reoperation, especially for consideration of the particular operative approach. Most commonly, a cervical incision is the incision of choice, especially because the residual disease in more than 90% of patients referred for reoperation is accessible through such an incision. Preoperative localization studies (discussed in the next section) are invaluable for operative planning. Another important reoperative consideration is that patients and/or their parents understand the risks of reoperative surgery—including the higher incidence of recurrent or superior laryngeal nerve injury, hypoparathyroidism, or persistent hyperparathyroidism than in primary parathyroid resections. Finally, a preoperative laryngoscopy should be part of the preoperative work-up to document any cord paralysis from the initial resection (9,22).

In persistent or recurrent secondary hyperparathyroidism, reoperation is also undertaken with caution. Indications for reoperation include ongoing nephrolithiasis, renal impairment, worsening osteopenia, serum Ca levels greater than 3mmol/L, and failure of medical management. Less-clear indications include severe aches/pain, fatigue, and depression. Preoperative evaluation in these patients is determined by the previous operative procedure. Those who underwent total parathyroidectomy with autotransplantation may or may not need a neck exploration, whereas those who underwent subtotal parathyroidectomy will all need a reoperative neck procedure. Just as in primary parathyroid disease, a thorough discussion of the risks of reoperation and a preoperative laryngoscopy are advisable prior to reoperation.

Localization Studies

As mentioned previously, preoperative localization is the standard of care prior to most primary parathyroid resections (with the exception of resection for secondary hyperparathyroidism in adults). The unique challenges of RPS make localization studies an even more crucial part of the preoperative work-up. At a minimum, the fibrosis and distortion of landmarks and tissue planes from the initial operation complicates accurate identification of residual parathyroid tissue. This is especially true when the initial operation involves an extensive bilateral neck exploration, such as in primary resection for secondary hyperparathyroidism. The reoperative surgeon may also be faced with uncertainty about number or functional status of parathyroid glands left from previous operations. Through the use of multiple imaging techniques preoperatively, 80–90% of parathyroid tumors can be localized (24). In fact, reoperative parathyroid resection is best thought of as an operation designed to identify and remove a *preoperatively identified* abnormality (29). Most recent studies recommend at least two concordant

localization studies prior to reoperation (30). Three studies may be necessary for localizing a persistent or recurrent parathyroid adenoma if initial study is not categorically positive (29).

Whereas selection of preoperative localization studies depends on the nature of the initial resection, the almost universally recognized first-line imaging study in parathyroid surgery is sestamibi/technetium 99m scintigraphic scanning. The technique is based on the specificity of the radiopharmaceutical agent technetium 99m sestamibi for mitochondrial membranes, which leads to its concentration in metabolically active tissue. The distribution of sestamibi in images of the chest and neck taken at 15 minutes is compared to the distribution in images taken 3 hours later to look for areas of increased uptake consistent with a hyperfunctioning gland. Preoperative sestamibi scanning has been shown to have a sensitivity of 82–86%, with a specificity ranging from 85–94% and approaching 100% in patients with a solitary adenoma (31). Although the high sensitivity and specificity of preoperative sestamibi scanning have made it a first-line modality of choice, it is more limited in the setting of multiple hyperplastic glands or small adenomas (especially small adenomas located behind the thyroid gland). Dual-tracer subtraction scintigraphy, a technique involving a second radiotracer that accumulates in thyroid but not parathyroid tissue, allows for subtraction of the two images to more clearly distinguish small amounts of abnormal parathyroid tissue in or around the thyroid gland. Combining sestamibi scanning with single-photon emission computed tomography (SPECT) is another strategy that allows for better anatomic localization of nodules, especially in the anterior-posterior plane. Another limitation of sestamibi scanning is that other metabolically active tissues including glands with diffuse hyperplasia, thyroid nodules or carcinoma, or surrounding lymph nodes can confound the scans. However, using simultaneous CT scan or neck ultrasound as a confirmatory study helps overcome these limitations. For the reoperative neck, a minimum of two studies should be used in any case, decreasing the risk of missing additional adenomas or diffuse hyperplasia.

The other classic first-line study prior to parathyroid resection is high-resolution ultrasound. In reviews of various localization studies, ultrasound has been shown to have a sensitivity and specificity ranging from 75–88 and 65–90%, respectively, on initial operation. The presence of a thyroid nodule has been correlated with a higher rate of false negative readings on ultrasound, especially in the reoperative neck. Also, visualizing superior parathyroid glands located medially or deep in the neck or retropharyngeal space is difficult. Overall, however, sensitivity and specificity of high-resolution ultrasound in preparation for reoperation is not significantly less than prior to initial operation (80 and 84%, respectively, for reoperation compared with 88 and 90%, respectively, for initial operation in a recent study with experienced operators) (32). The strengths of ultrasound include its ability to provide more exact anatomical localization than sestamibi and to examine vascular structures and supply in addition to adenoma position. Combining ultrasound and sestamibi scanning for preoperative imaging has been shown to increase the sensitivity of the studies to 94–96% (33,34).

Other imaging techniques prior to operation include CT and MRI. Although previous studies had reported lower success rates for these modalities, recent studies have shown a sensitivity for CT as high as 98%, and as high as 80% for MRI if used prior to initial resection. However, metallic clip artifacts from clips placed during the initial operation can lower the sensitivity of CT to 48–56% (33). Small gland size and

coexisting thyroid disease limit the accuracy of CT/MRI as well. Few studies have compared the techniques on the reoperative neck, and neither CT nor MRI should replace sestamibi scanning and ultrasound as a front-line modality. Also, CT and MRI have been shown to offer no additional benefit in the preoperative setting if sestamibi and ultrasound results are positive. However, these noninvasive techniques are useful when sestamibi scanning and ultrasound are discordant or inconclusive. CT is especially useful for ectopic parathyroid tumors, which can be missed by ultrasound, as well as imaging mediastinal tumors or glands within the tracheoesophageal groove. At least one recent study has shown a significant benefit in combining sestamibi scintigraphy with SPECT and CT (giving a sensitivity of 93 versus 31% for sestamibi scintigraphy-SPECT alone) for localizing ectopic adenomas and in reoperative patients with distorted anatomy from prior operation (35).

More invasive preoperative localization techniques are warranted when noninvasive studies are ambiguous or discordant. Selective venous sampling (SVS), often regarded as a gold-standard localization procedure, is not routinely used preoperatively unless noninvasive studies fail. The technique involves catheterization of multiple veins in the neck and mediastinum with samples sent for measurement of iPTH, and usually requires general anesthesia when performed on children. Gradients in PTH can regionalize and sometimes precisely localize an abnormal gland based on venous drainage patterns. A high vertebral vein gradient predicts a retroesophageal parathyroid gland, whereas a high gradient in the internal thoracic veins predicts an ectopic gland in the thymus. With the recent availability of rapid PTH assays, interventional radiologists who perform selective arteriography can further investigate PTH gradients they find for even more precise anatomic localization in the angiography suite. Recent studies have shown SVS even without arteriography to have a sensitivity as high as 90–94% (36). Given the difficulty of reoperation, the benefits of SVS outweigh the length of the procedure and risk of procedure-related thrombosis or hematoma when other studies have failed to adequately or consistently localize an adenoma (37).

Finally, FNA can be used for preoperative localization as a part of ultrasound-guided biopsy or for measurement of iPTH. FNA with ultrasound-guided biopsy has been reported to have a sensitivity rate of 90% and an accuracy of 82% (22). FNA measurement of iPTH is not routinely indicated unless preoperative localization is difficult, but in such cases can be helpful in distinguishing between pathological parathyroid tissue versus normal thyroid or parathyroid. Measured iPTH levels greater than 1000 pg/mL are predictive of a pathological parathyroid gland with a sensitivity of 87%, whereas levels less than 100 pg/mL are highly predictive of the absence of parathyroid tissue (as in an intrathyroid aspiration) (30).

For secondary hyperparathyroidism that persists or recurs, the choice of preoperative localization techniques depends on the initial operation. For those patients who underwent a total parathyroidectomy with autotransplantation, the Casanova test is the initial test of choice. This technique uses peripheral venous sampling of iPTH in the extremity used for prior autotransplant. If the test shows no source of PTH other than the graft site, the reexploration can be limited to that area (usually forearm), limiting the risks of morbidity for patient and, in some cases, limiting the need for general anesthesia. If the autotransplant is not the only source of PTH, an additional adenoma or engraftment is suspected and further preoperative imaging is needed.

For patients with a previous subtotal parathyroidectomy, sestamibi imaging can help localize the remaining hyperfunctioning gland. As with primary hyperparathyroidism,

the combination of sestamibi scanning and ultrasound increases the success of localizing hyperfunctioning glands prior to reoperation (to as high as 88 from 75%) (38). Performing sestamibi scanning prior to ultrasound guides radiologists to the areas of hyperfunctioning glands, allowing for more exact localization on ultrasound. Missed glands are often small, superior glands, although ectopic or supernumerary glands are possible. CT should be part of the preoperative evaluation if initial studies are not concordant or there is any ambiguity.

Using intraoperative localization modalities as adjuncts may also increase the success of reoperation. Intraoperative ultrasound (IOUS) and sestamibi scanning with gamma probe are commonly used techniques in conjunction with intraoperative PTH monitoring. The additional benefit of these techniques compared with thorough preoperative localization and intraoperative PTH assay is debated in the literature. IOUS has been shown to detect more abnormal parathyroid glands than preoperative ultrasound and shorten operative time by as much as 50%, but has not been shown to significantly affect ultimate outcomes (39). Operator skill and experience may also limit the usefulness of IOUS.

Many surgeons have favored intraoperative sestamibi scanning as a technically easier modality than intraoperative ultrasound. The technique requires the injection of technetium 99m sestamibi injected 2–4 hours prior to making the neck incision. A gamma counter with a 9–14 mm handheld gamma detection probe is used to scan the neck after division of the strap muscles. The neck is probed in all four quadrants at the outset, with further exploration directed at those areas of the neck with the highest radioactivity counts. The count directs the surgeon to the hyperfunctioning gland, which usually has counts elevated as much as 20–50% over background. Once the adenoma is removed, radioactivity in each of the four quadrants should equalize. As with IOUS, intraoperative sestamibi scanning is more successful at localizing adenomas than its preoperative counterpart, especially in the reoperative neck (localizing 91% of hyperfunctioning compared with 64% each by preoperative sestamibi and ultrasound) (24).

Finally, intraoperative parathyroid hormone monitoring (IOPTH) is an extremely informative technique to confirm quantitatively that all hyperfunctioning parathyroid tissue has been removed. A drop in PTH levels of 50% or greater at 20 minutes from parathyroid resection is widely used as confirmation of a successful resection. Some authors have argued for a 70% drop, especially in the reoperative setting to avoid false positive results in patients with multigland disease (22). The use of IOPTH in RPS has been shown to increase the success rate of the procedure from 76 to 94% (40). Of note, studies have found that a decrease in PTH of more than 84% is associated with a high risk of developing hypocalcemia in the postoperative period—making the technique useful for identification of patients who would benefit from closer postoperative monitoring (40) (Table 1).

Reoperative Approach

As described previously for thyroid reoperation, parathyroid reoperation should be undertaken either during the first week after the failed initial procedure or 3 months or more afterwards to avoid the inflammation of the immediate postoperative period. Given to the difficulty of reoperation, the surgeon's goal is a focused, direct, and limited approach to resect the abnormality or abnormalities identified on preoperative imaging.

Table 1
Parathyroid Localization Techniques

<i>Study</i>	<i>Sensitivity</i>	<i>Specificity</i>	<i>Notes</i>
Sestamibi scintigraphy	82–86%	85–94%	
High-resolution US	75–88%	65–90%	
Sestamibi scintigraphy + US	96%	83%	
CT	80–98%	*	Sensitivity drops to 48–56% when metallic clip artifact is present; Older studies report CT sensitivity ranges as low as 13–50%
CT + US	91%	*	
CT + sestamibi scintigraphy	74–90%	*	
CT / SPECT-sestamibi scintigraphy	91–96%	*	
MRI	50–80%	78%	
MRI + US	79%	50%	
MRI + sestamibi scintigraphy	75%	39%	
SVS	90–94%	85%	More invasive technique (<i>see text</i>)
US-guided FNA with iPTH measurement	87%	75%	

Data obtained from the following (adult series): Refs. (31–37)

CT, computed tomography; US, ultrasound; MRI, magnetic resonance imaging; SVS, selective venous sampling; FNA, fine needle aspiration; iPTH, intact parathyroid hormone.

Because most missed adenomas will be accessible through a cervical incision (with the most common site of a missed parathyroid adenoma along the esophagus in the posterior superior mediastinum), a cervical approach is reasonable. After the skin incision, a lateral “back door” approach using the plane lateral to the sternohyoid and sternothyroid muscles and medial to the sternocleidomastoid offers the advantage of avoiding dissection through previously mobilized strap muscles. The plane is carried to the level of the carotid sheath. The thyroid and strap muscles are retracted anteromedially. The pharynx and larynx are rotated into view and the recurrent laryngeal nerve is identified early in the procedure. Exploration continues above the inferior thyroid artery to the prevertebral fascia and should include digital exploration of the retropharyngeal space. Meticulous hemostasis is imperative, because blood-staining may obscure parathyroid tissue in an already difficult operative environment. If the exploration is unsuccessful despite preoperative localization data, other locations such as the thyrothymic ligament, cervical thymus, subcapsular and intraparenchymal thyroid, and carotid sheath should be systematically explored (9,10,41).

When a mediastinal adenoma is suspected, it is generally accessible through a cervical incision. The most common location of a mediastinal adenoma is in the anterior mediastinum associated with thymic tissue. Thymectomy is indicated for a

closely associated adenoma or when the lower parathyroid gland cannot be identified in isolated familial hyperparathyroidism or secondary hyperparathyroidism. To aid in dissection of the thymus, retraction of the thyroid gland allows the surgeon to follow the inferior thyroid vessels to the anterior mediastinum. After the thymus is identified, it is grasped and pulled into the field, then excised.

If necessary, further mediastinal exploration is possible through a “T” incision made by carrying a vertical incision down from the cervical incision. A partial median sternotomy is then made. The thymus is resected and exploration continues to the innominate veins and along the jugular and subclavian veins. A mediastinal drain should be placed and the sternotomy closed. For very well-localized mediastinal adenomas, video-assisted thoracic surgery has been used with success in adults. Also, a limited left thoractomy may be necessary for middle mediastinal adenomas after failed cervical and anterior mediastinal explorations.

For reoperation after total parathyroidectomy with autotransplantation to an extremity (such as a forearm), reoperation can be safely limited to the extremity if the preoperative Casanova testing confirms that the extremity was the source of PTH. Reexploration of the extremity is a much less involved and less technically difficult procedure.

All reoperative parathyroid resections should use IOPTH monitoring as a confirmatory study. A 50–70% decline in PTH from baseline indicates successful reoperation. If PTH levels do not decline after 20 minutes, continued exploration is necessary.

Results of Reoperation

Complications of reoperation include persistent hypercalcemia, which indicates a failure of reoperation and should prompt further workup for missed or engrafted parathyroid tissue or multigland hyperplasia. The major cause for reoperative failure is multigland disease, which accounts for 73% of such failures (22). Permanent hypoparathyroidism affects almost 13% of reoperative patients. Care should be taken postoperatively to distinguish between hungry bone syndrome after reoperation for secondary hyperparathyroidism and true lack of parathyroid function. In hungry bone syndrome, the serum phosphate level is low (<2.5 mg/dL), whereas in hypoparathyroidism it is elevated (>4.5 mg/dL). Although care is taken to identify the recurrent laryngeal nerve, nerve injury still occurs in almost 1% of patients (an improvement from 4% two decades ago) (22). Advances in preoperative and intraoperative localization as well as nerve monitoring have contributed to this improvement.

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5

Complications of Pectus Excavatum and Carinatum Repair

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CONTENTS

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INTRODUCTION

TO A FRIEND WHOSE WORK HAS COME TO NOTHING

Now all the truth is out,
Be secret and take defeat
From any brazen throat,
For how can you compete,
Being honour bred, with one
Who, were it proved he lies,
Were neither shamed in his own
Nor in his neighbours' eyes?
Bred to a harder thing
Than Triumph, turn away
And like a laughing string
Whereon mad fingers play
Amid a place of stone,
Be secret and exult,
Because of all things known
That is the most difficult.

W.B. Yeats (*1*)

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Unlike Yeats' friend, the surgeon whose work has met with need for reoperation cannot only console himself with having made a sturdy effort at the first procedure. Reoperative surgery for chest wall problems demands thorough evaluation of the patient, the previous treatment, and careful planning for revision. In this chapter we will review recurrent pectus excavatum, carinatum, and some less frequent conditions affecting the chest wall.

RECURRENT PECTUS EXCAVATUM

Clinical Presentation

Reoperative pediatric surgery for pectus excavatum almost always involves recurrent chest wall depression after previous repair. Occasionally, the condition complicates surgical treatment for some other condition, such as congenital diaphragmatic hernia or lung resection.

Pectus excavatum can occur or reoccur following any of the operations employed to repair it. Because pectus excavatum is an obvious physical deformity, screening is not required postoperation.

Frequency of recurrence following the open or Ravitch operation was reviewed and summarized by Ellis and colleagues in a 1997 report (2). Recurrence rates were reported to be 2% by Fonkalsrud, 2.4% by Shamberger, 5% by Haller et al. in a contemporary report, 6% by Sanger et al., 10% by Gilbert, 11.8% by Singh, 16% by Pena, and 20.5% by Willital and Meier if no internal supporting bar was used, but only 8.9% if a bar was used (2). Subsequent to Ellis's review, Lacquet in 1998 reported 16% unsatisfactory results in a large series (3). Ellis and colleagues supported the observation of others that recurrence was related to a limited procedure at the first operation. When they operated on recurrences after operation by other surgeons, they found deformed cartilages, which appeared to be untouched. However, it is worth emphasizing that recurrence happens for unclear reasons. Recurrence affects patients of even the most expert surgeons at all centers reporting in the literature.

Two reports cite a higher recurrence in Marfan patients (4,5). Of 28 patients with Marfan syndrome and pectus excavatum in a 1989 Johns Hopkins report, 11 recurred (5). Haller and colleagues recommended delay in repair until skeletal maturity had been achieved, and internal stabilization with an Adkins strut.

Recurrence rates after the Nuss repair are less well established, because the operation only came into general use in 1997, less than 10 years ago as of this writing. Numerous observers have pointed out that until a large cohort of children who have undergone the repair have passed through puberty, reliable recurrence rates will not be available. It is important to distinguish in these patients an early bar shift and recurrence after removal of the bars is done 2 years postoperation. In our series of almost 800 patients with pectus excavatum, bar shift has occurred approximately 13% of the time in the first 100 patients, while the procedure was in evolution; with current methods of securing the bar it is approximately 0.5% (1/245 patients). Park recorded bar displacement in 2.4% of 335 patients (6). Early bar shift presently is often related to some unexpected event such as being tackled by uninformed friends or a violent twisting motion. Late recurrence has occurred in 0.9% (7/794) of patients treated at Children's Hospital of the King's Daughters (CHKD)/Eastern Virginia Medical School (EVMS).

Timing of recurrence is affected by the type of repair. There are two operations in common use: the open or "Ravitch" repair (attributed by Ravitch himself as a variation

of Lincoln Brown's 1939 method) (7), and the minimally invasive or Nuss operation. After open repair, recurrence is reported most frequently in the first 2 years following operation, or else at the age of the pubertal growth spurt (often at age 12 or 13 years) (7). After the Nuss operation, in which stainless steel bars are used to push the sternum to the normal anatomic position, recurrence in the first 6 months is almost always caused by shift in the position of the support bars. The bars are anchored to the soft tissue with numerous sutures, and so if they move more than a small distance, the patient usually experiences severe pain. Apart from this circumstance, recurrence almost never occurs when the supporting bar is in place. This holds true for the cartilage resection (Ravitch) operation as well if a supporting strut is employed to brace the sternum after removing portions of the cartilages connecting it to the ribs. But because the support bar, when it is used at all, is removed 6 weeks to 6 months following a Ravitch repair, recurrence can occur early after that operation. Early recurrence after Ravitch operation is thought by some surgeons to be caused by inadequate removal of cartilages at the initial operation. Late recurrence at the pubertal growth spurt is noted after either repair (2).

Symptoms: When the reoccurrence of the depression in the sternum happens later, it is almost always gradual and painless regardless of which operation was used for the primary repair. The patient or family notes the change in external appearance as the only symptom. Authorities agree that recurrence is associated often with periods of rapid growth at adolescence. Gilbert and Zwiren reported 60% of recurrences were after 12 years of age (8).

In 1996 the occurrence of asphyxiating thoracic dystrophy (ATD) was reported to follow too early and too extensive cartilage resection in operations to treat pectus excavatum (9). In children who underwent operation at less than 5 years of age, and whose cartilages were aggressively removed, the chest in later years failed to grow, and the thoracic cage was constricted to an inadequate size (Fig. 1). Patients with this problem often have significant symptoms of exercise intolerance and shortness of breath with very mild exertion. Radiographs often show a replacement of the normally cartilaginous connection between the sternum and ribs with dense bone (10). This problem has proven very difficult to treat.

Fonkalsrud notes,

When the deformed costal cartilages are removed with preservation of the perichondrial sheaths, the sheaths are often damaged, and the regenerated cartilage is often thin, irregular, and commonly rigid with varying amounts of bone and calcification... If the regenerated cartilage is rigid, the chest essentially becomes a cylinder, with respiratory motions being largely dependent on diaphragmatic excursions, which limits the depth of lung expansion and requires more effort than normal respiration (11).

Cartilage resection must not be too extensive, or ATD may result. If it is inadequate, though, recurrence early is a risk. Most authors at present emphasize the need to free all deformed cartilages and their contralateral partners by a limited resection, but suggest leaving a few millimeters of cartilage on the rib and sternal ends, hoping that this maneuver will encourage regrowth of the cartilage. Haller and associates, who first reported the problem after operations done by others, advise against operation in children younger than 4 years of age, and against removal of five or more pairs of cartilages (11,12).

Diagnostic evaluation depends on the acuity of the recurrence. In the early period postoperation, a chest X-ray with a clear lateral view will identify movement of support bars or Adkins struts. In later recurrence, a computed tomography (CT) scan of the chest will show the area of the sternum involved, and whether the cartilages involved have ossified. Ossification of the costal cartilages, seen radiographically as bright white signal, and pathologically as cancellous bone if the cartilages are resected, sometimes occurs following the Ravitch repair (10) (Fig. 2). This is very useful information in deciding on treatment. If a repeat Ravitch repair is to be done, the ossified cartilages need to be divided or removed in order to mobilize the sternum. If a Nuss operation is to be used, the patient and family need to be aware of the reduced mobility of the chest wall due to the calcified cartilages.

Pulmonary function studies help to clarify the physiologic effect of the recurrent depression. Even in its unoperated state, in which the change of shape of the thoracic bellows is not affected by postoperative scarring, the overwhelming majority of patients with pectus excavatum note exercise intolerance, especially as lack of endurance. Mark Ravitch noted 50 years ago that although some patients have no limitation of activity, "It is more common to hear of a boy who can 'fool around' shooting basketball but not play a game, or play a game of tennis but not a set." (13,14).

"Static" pulmonary function tests including spirometry, which measures the air flow and volume on exhalation, and plethysmography, which measures the volume of the chest cavity taken up with air, show diminutions in unoperated patients with pectus excavatum. We and others have reported spirometry values which are on average 80% or so of predicted values in such patients (15,16). Remembering that the predicted values are mean values for the population, it is not surprising that after operation we and others have found that they are improved by clinically significant amounts (15,16).

We have performed redo pectus operations on 65 patients. In those patients, we found spirometry values diminished below those of unoperated patients, in the vicinity of 70% of predicted mean values (Table 1) (Previously unpublished data).

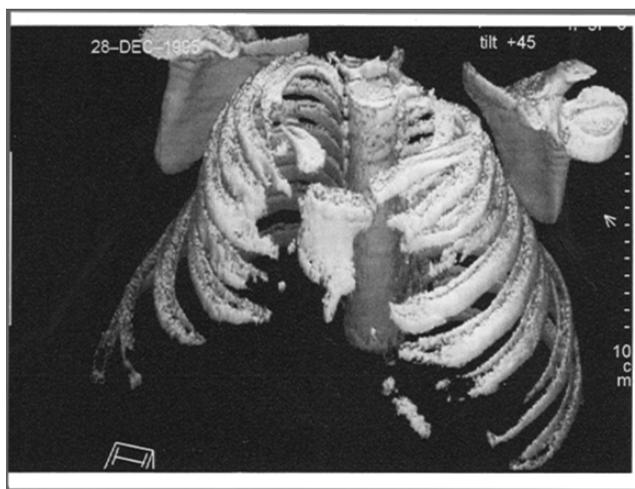


Fig. 1. Reconstruction of a preoperative CT scan in a child with ATD 10 years after pectus repair showing a narrow upper chest cavity secondary to lack of rib growth (36).

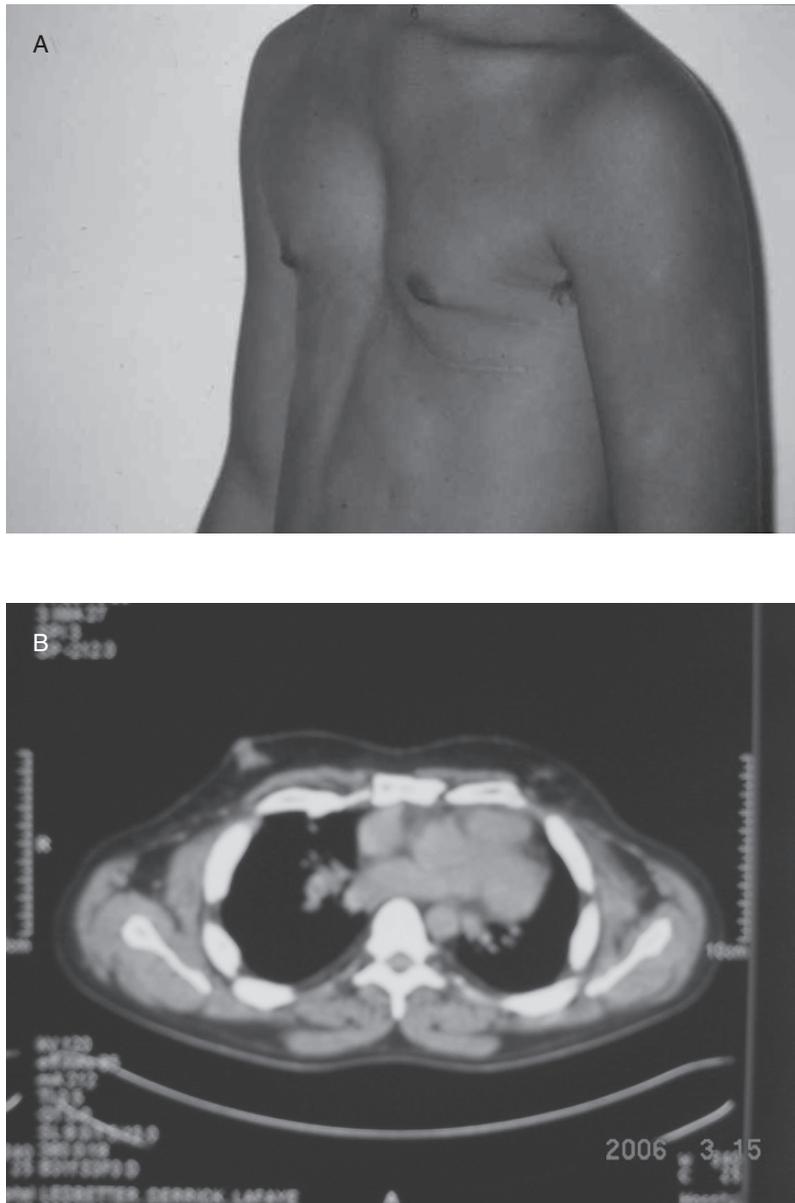


Fig. 2. Clinical photograph and CT scan of patient with recurrent pectus excavatum 17 years after repair, showing ossification of the costal cartilages and a narrow upper chest cavity.

Weber reported on static pulmonary function measurements in 10 patients with ATD. Forced vital capacity (FVC) was $52 \pm 4\%$ of predicted mean values for the population; forced expiratory volume in 1 second (FEV1) $51 \pm 3\%$, forced expiratory flow (FEF) 25–75 was $62 \pm 3.5\%$. Peak expiratory flow was $54 \pm 4\%$ of predicted mean values. Restrictive disease is diagnosed from these values (the ratio of FEV1 to FVC is greater than 80%). In patients with stiff immobile chest walls but normal lung parenchyma, gas exchange is not impaired, but movement of air in and out of the chest is impeded, with spirometry approximately 50% of predicted values (17) in ATD.

Table 1
Pulmonary Functions Studies in Patients Presenting for Redo Repair of Pectus Excavatum,
CHKD/EVMS, Norfolk, Virginia

	<i>Mean % predicted</i>	<i>Standard deviation</i>	<i>Number of patients</i>
FVC	71	17	55
FEV1	70	18	55
FEF 25–25	70	24	53
TLC	85	26	20
MVV	68	20	16
PEF	68	13	14

CHKD, Children's Hospital of the King's Daughters; EVMS, Eastern Virginia Medical School; FVC, forced vital capacity; FEV1, forced expiratory volume in 1 second; FEF, forced expiratory flow; TLC, total lung capacity; MVV, mitral valve closing speed; PEF, peak expiratory flow.

Because most patients with pectus excavatum do not complain of shortness of breath at rest, but only with exertion, exercise pulmonary function tests can be very useful in quantifying the amount of impairment. Malek et al. reported on 21 patients with pectus excavatum and a mean index of 5.1 (normal 2.4; severe pectus is >3.2). They found that maximal oxygen uptake and oxygen pulse were significantly lower than reference values. VO₂ max was 41% of the predicted value! The authors (who are pulmonologists) noted that these findings were not a result of deconditioning; and most patients exhibited cardiovascular limitation, not ventilatory limitation (18). Haller and Loughton reported significantly increased duration of exercise after operation (16).

Evaluation should also include echocardiogram. Mitral valve prolapse (MVP) is reported to occur in from 8–45% of patients with pectus excavatum. In approximately one-half of those, the MVP resolved after surgical treatment for pectus excavatum (20). Compression of the right ventricle by the chest wall is also frequently seen. Echocardiogram was used by Sigalet to follow stroke volume, which improved from 61.6 ± 25 to 77.5 ± 23 mL after primary repair of the chest wall (21). Both of these preconditions may be improved by the chest wall operation (16,22,23).

Body image concerns frequently drive the patient or family to seek correction of the sternal depression. This is as true with recurrence as with initial operation. A body image survey developed with a psychologist expert on the topic (24) may be used both to validate the degree of concern about self-image and to document the postoperative improvement in activity levels. Improvements in disease-specific quality of life have been shown in both children and adults by questioning parents and patients (24,25).

Operative Techniques

Historically, several techniques have been used for repair of recurrent chest depression. The external fixation of the repair introduced in the early 1900s is still used occasionally and needs to be borne in mind as a tool for stabilization (26).

REPAIR OF RECURRENT PECTUS EXCAVATUM BY THE OPEN OR RAVITCH OPERATION

Re-repair by the open operation has been well detailed by several authors. Recently some authors have emphasized that only a small portion of the affected cartilages needs

to be resected; earlier others have emphasized that all of the involved cartilages need to be treated to get a good result (2,11).

The technique of open repair for recurrent pectus excavatum is performed under general endotracheal anesthesia (27), with positive pressure a safety feature to assure lung inflation in case the pleura is entered. Thoracic epidural analgesia can be a useful adjunct in the hands of an anesthesiologist skilled in its use. A mixture of fentanyl and bupivacaine is very effective and can be continued after the operation. Appropriate anesthetic monitoring equipment includes oxygen pulse oximetry and end-tidal carbon dioxide monitoring to assure adequate gas exchange. Prophylactic antibiotics are appropriate; intravenous Ancef® (cefazolin) is a reasonable choice.

Either a transverse inframammary or a vertical midline incision provides adequate exposure. Generally the incision used for the initial repair is utilized. Skin flaps are created. The pectoralis major muscle is detached from its origin on the costal cartilages bilaterally, and the muscle is mobilized out of the way. Similarly, the rectus abdominis muscles are detached. After inspection of the sternum and cartilages to determine which cartilages need to be divided, the perichondrial sheath is incised along the long axis of the cartilage. Then the cartilage must be freed from the perichondrial sheath with a Freer elevator or similar instrument. In a redo case, there is often not a well-defined plane (Fig. 3).

Some length of the cartilage must then be removed at each of the involved deformed cartilages. Most authorities are in agreement that only as much cartilage should be removed as is necessary to allow the ribs and sternum to come into good alignment. Because occurrence of ATD has complicated too early and too extensive operations by this procedure, removing as little cartilage as practical seems prudent. Some authors have proposed that only 3–8 mm of cartilage be removed for a primary repair (11); but sometimes more must be removed to allow a twisted sternum with lengthily deformed cartilages to come into good alignment (2). Robicsek notes,

...the surgeon should not waste time in performing a meticulous 'classic' subperichondrial resection of the cartilages and ribs but should just leave enough perichondrium and periosteum behind to ensure the regeneration of the ribs. For the same reason, a segment of the most lateral portion of the cartilage should be left in continuity with the ribs (28).

Sternal osteotomy may be needed to allow the sternum to come anteriorly. Fracturing the anterior table only with an osteotome is usually sufficient to allow twisting and fracturing the posterior table with one's fingers, allowing the deformed sternum to come into good position (Fig. 4). The process of sternal mobilization in a reoperative field may result in entering the pleural cavity, especially on the right. If so, a chest tube should be considered. Most surgeons then place a stainless steel support behind the sternum, such as an Adkins strut or Lorenz bar. The support bar is secured to the soft tissues with suture or wire (Fig. 5). Occasionally, the support can be placed anterior to the sternum and held in place with sutures. In larger patients, more than one support bar may be necessary.

The pectoralis major muscles are sutured together, and the rectus abdominis sutured to the inferior edge of the pectoralis muscles. Closed suction drains may be placed beneath the muscle layer. Skin closure may be with subcuticular suture.

Support bars may be removed 2–6 months after repair as an outpatient operation. Migration of the supporting strut used for Ravitch repair has been reported, with the bar

coming to rest in the myocardium, thoracic cavity, or peritoneal cavity, and with injury to the heart, lungs, or abdominal viscera (29–34). Removal is thus to be encouraged when it is not needed.

Operation for treatment of acquired ATD or acquired Jeune's syndrome has been pioneered by Dr. Thomas R. Weber. A midline sternotomy is performed, and the pleura is widely opened bilaterally to allow the lung to herniate into the anterior mediastinum. Immediately, this allows pulmonary compliance to improve, with a marked increase in tidal volume with unchanged ventilatory pressures (36).

A subperiosteal lateral osteotomy of all ribs attached to the sternum bilaterally allowed the sternal halves to be separated by 4–8 cm, increasing the mediastinal herniation of lung. Three straight bony rib segments of appropriate length were resected, generally from the lower ribs, and "wedged" and wired to hold the sternal halves apart permanently (Fig. 6).

After operation, all patients remained on a ventilator for 3–7 days. Because the heart is no longer covered entirely by bone, patients have been advised to wear a chest protector for sports after operation. Patients report tremendous subjective improvement in breathing, but pulmonary function studies did not show great improvement. Body image was a concern after operation in approximately 70% of patients (36).

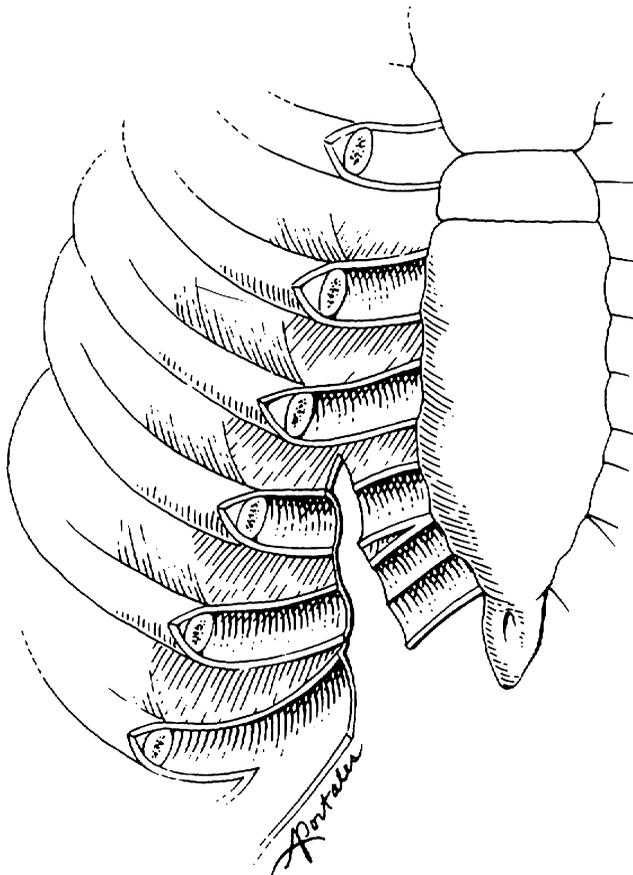


Fig. 3. Division of the right fifth, sixth, and seventh perichondrial bundles and intercostal muscles (2).

REPAIR OF RECURRENT PECTUS EXCAVATUM BY THE NUSS OPERATION

Minimally invasive repair (Nuss procedure) redo operation is accomplished with similar anesthetic and perioperative considerations as the Ravitch operation: General endotracheal anesthesia, epidural analgesia supplement, full anesthetic monitoring, perioperative antibiotics, and bladder catheterization (37). Allergy to nickel or other metal components of the pectus bar should be sought by application of a skin patch test (38). In the case of allergy to stainless steel, a titanium bar can be made to order at the factory. Because titanium must be highly polished to prevent tissue ingrowth to the bar, and because bending the bar scratches it, and because titanium is very stiff and hard to bend, the bar must be preconfigured (bent) at the factory. This is done by using computer-assisted design/computer-assisted manufacturing (CAD/CAM) technology to shape the bar (typically 25–40 cm long) to conform to the shape on the patient's CT scan (which image is a few centimeters across) (Walter Lorenz Surgical, Inc., Jacksonville, FL).

Repair is performed throughout small bilateral midaxillary transverse incisions, subcutaneous tunneling, and intrathoracic placement of a stainless steel or titanium bar (39). The surgical technique is similar to the technique described for primary repairs with the addition of thoracoscopic lysis of adhesions as required to gain adequate visualization.

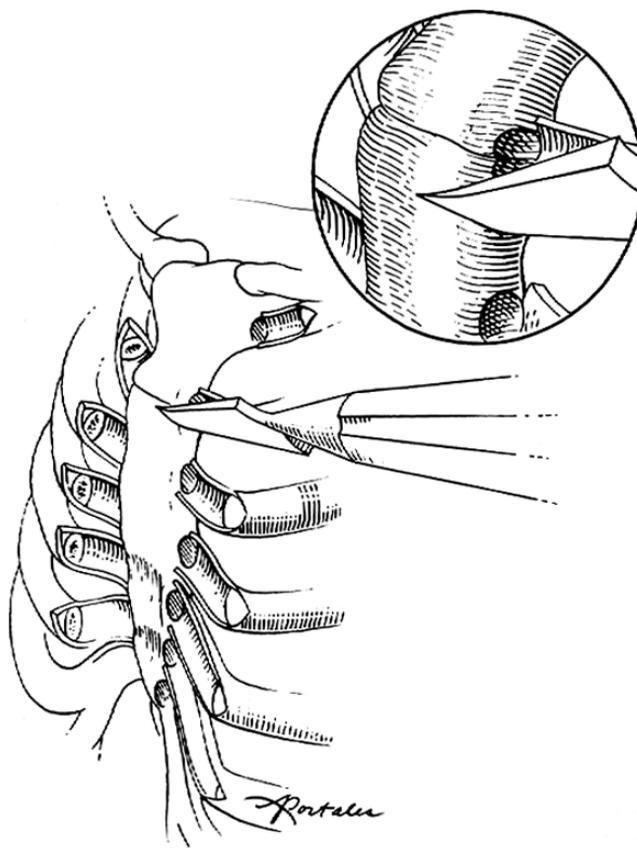


Fig. 4. Division of the anterior table of the sternum leaving the posterior table intact (2).

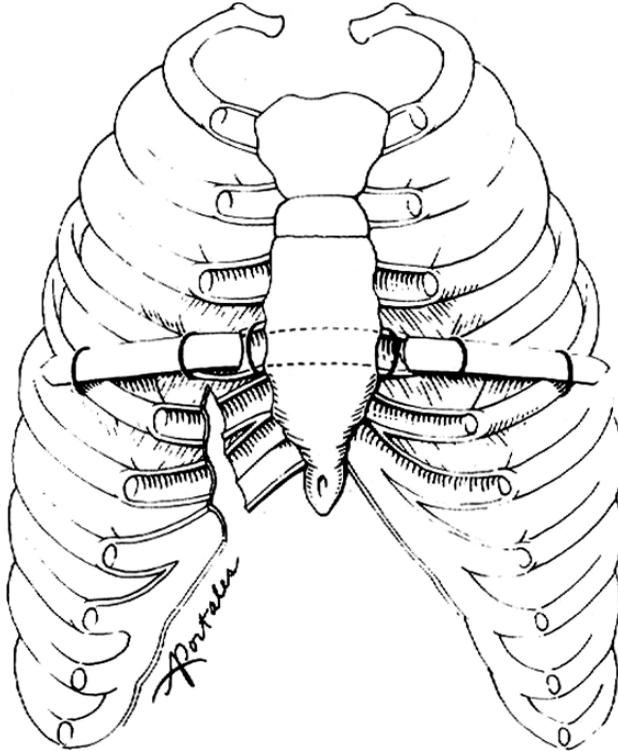


Fig. 5. Bar secured in place behind the sternum and in front of the ribs (2).

The width of the chest is measured in the operating room prior to surgery, and the correct length steel bar is selected and bent to conform to the curvature of the anterior chest wall. A small transverse incision 2 or 3 cm long is made in each lateral chest wall between the anterior and posterior axillary lines. Subcutaneous tunnels are raised.

A site for entering the chest medial to the pectus ridge is selected (Fig. 7). In a redo patient, it is crucial that the substantial pressure the bar exerts on the chest wall to oppose the pressure of the sternum is borne by the ribs, not the intercostal muscles. If the site of entry into the thorax is too lateral, no rib surface is available for load bearing, and the intercostal muscles will tear because they cannot support that much weight.

A long clamp is used to enter the right hemithorax at this spot under direct visualization through a thoracoscope. The long introducer is passed through this tunnel, posterior to the sternum and anterior to the heart, to emerge through the contralateral intercostals space (Fig. 8). An umbilical tape is tied to the end of the introducer, and the umbilical tape is then pulled back through the tract. Once the umbilical tape is across the tract, the introducer is removed and the umbilical tape is transferred to the steel bar. Using the umbilical tape for traction, the steel bar is pulled through the tract with the convexity facing posteriorly, and then the umbilical tape is removed. Once the bar is in position, it is rotated using the bar flipper so that the convexity faces anteriorly, thereby raising the sternum and chest wall into normal position. One end of the bar is secured to a lateral stabilizer using steel wire, and each stabilizer is secured to the musculature of the chest wall with absorbable suture. The bar should be secured to the ribs with a heavy absorbable suture such as polydioxanone suture (PDS) under

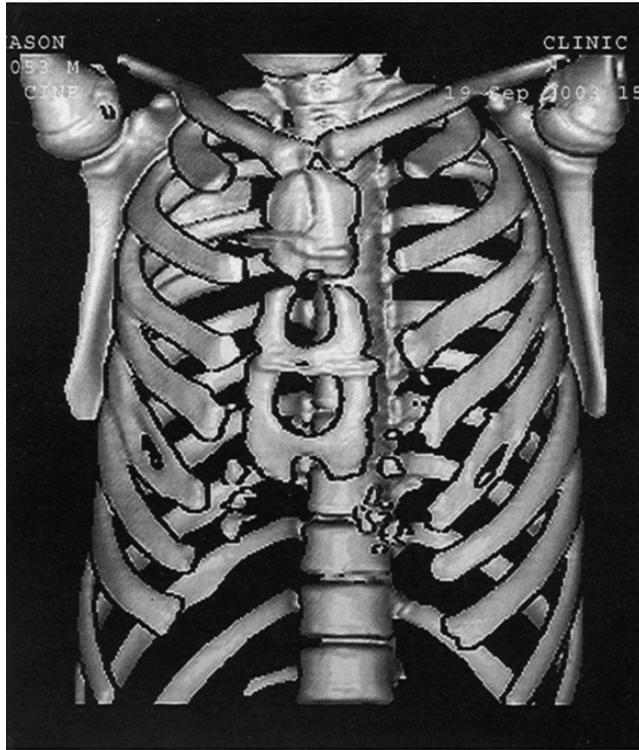


Fig. 6. Reconstruction of a CT scan performed 1 year postoperatively in a child with ATD showing complete healing of three rib grafts separating the sternal halves (36).

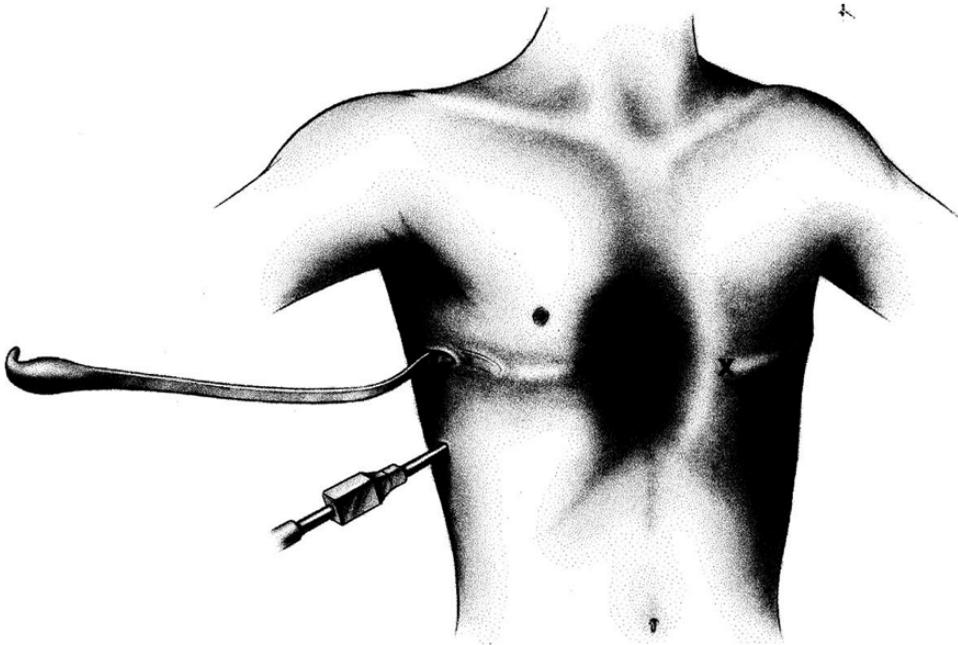


Fig. 7. Sites for entry of the curved dissector through the chest wall into the pleural space are marked with an "X" and are medial to the pectus ridge (39).

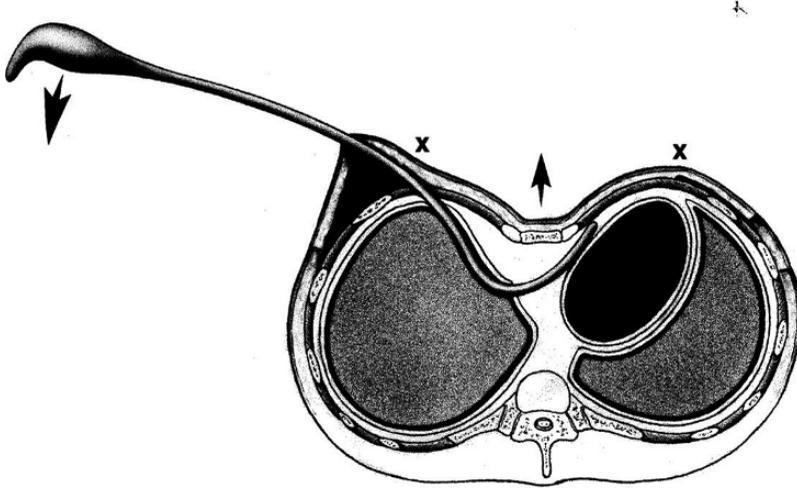


Fig. 8. The pectus bar is flipped over, pushing the sternum anteriorly (39).

direct vision with an endo-catch device (Fig. 9). Before closing the incisions, positive end-expiratory pressure is added to prevent pleural air trapping, then each wound is closed in layers.

There are several factors to consider for redo operations. In patients with an early recurrence, for example in the first 2 weeks after operation, the bar has usually flipped from its initial and desired position to one in which the sternum is not pushed anteriorly as much

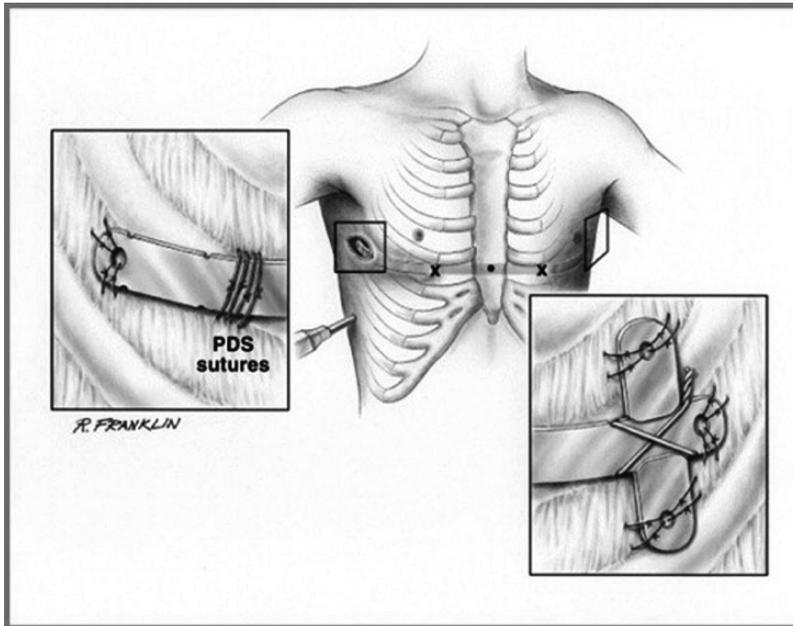


Fig. 9. The bar is secured in place by wrapping heavy absorbable sutures around the bar and the adjacent ribs with thoracoscopic guidance; then the stabilizer and bar are sutured to the muscle of the chest wall with absorbable sutures (39).

as desired. When this situation is recognized, it is important to put the bar or bars back where they belong. A group in Norway reported erosion of the arch of the aorta from a bar that had flipped to a position putting pressure on the aortic wall (40). Generally repositioning can be done through the initial incisions, even when bar displacement occurs more than 1 month after the operation (6,37,38,41). Thoracoscopy can be considered as needed. On-table radiography confirms the correct position of the bars.

It can be difficult at times to tell whether the bar has shifted in the first few days after operation. After repair a cross-table lateral taken in the recovery room can help with making decisions regarding need to reoperate later.

Usually redo operations are in older patients. Two bars should be considered to support the sternum, rather than a single bar. Two bars will not be needed in every instance, but one should have a low threshold to use two. The chest is often more rigid in redo ops, and two bars distribute the pressure over a broader area and bear less force on each bar. This may give a better result.

If two bars will be used, the upper bar is often the better one to put in first. Often, the deepest part of the depression is at the lower sternum or xiphoid process, where the lower bar will be placed. Tunneling between the pericardium and sternum may be easier cephalad to that spot, where the severity of the depression is less.

After passing the first dissector, it should be left in place to support the sternum anteriorly before tunneling with a second dissector at the inferior bar's site. This greatly improves visualization for tunneling at the deeper part of the chest depression.

Sometimes it can be difficult to see the tip of the dissector because of the deep chest depression. To improve visualization, besides changing from a 0° to 30° scope, it may help things to put the scope in the same opening in the chest as the dissector went through. A group in Japan uses this approach routinely with good results.

A bone hook can also be placed under the xiphoid and used to pull the xiphoid toward the ceiling of the operating room. A similar incision to that used for a transdiaphragmatic pericardial window gives access to the back of the sternum. The operator can sweep the tissue between the sternum away by blunt dissection and help guide the dissector across by palpation through this approach (43). Loss of insufflated gas is to be expected when this approach is used.

After the dissector is passed through successfully, the stiff chest of a redo patient should be slowly but firmly pulled toward the operating room ceiling by pressure from the surgeon and assistant (Fig. 10). If this is not done, the bar may be twisted into a slightly helical shape when it is flipped over by resistance from the chest wall. We have had one sternal fracture on a redo patient, and the elevation of the sternum should be done gently but firmly.

A sternal saw should be in the room, and the team prepared to use it. We have had one patient suffer an arrhythmia leading to full cardiac arrest as the dissector was passed across the mediastinum. In his very rigid chest, pressure from the dissector may have produced bradycardia and then arrest. Sternotomy and open defibrillation were needed for resuscitation. We have had only one such event in almost 800 Nuss operations at our facility.

If the patient has numerous intrapleural adhesions, which can occur in considerable amount even after Ravitch repair, one should have a low threshold to leave a chest tube in place, because small air leaks may result from the dissection.

In reoperative patients, the bars should be especially well secured to the ribs with heavy PDS sutures taken doubled back so two strands go around each rib.

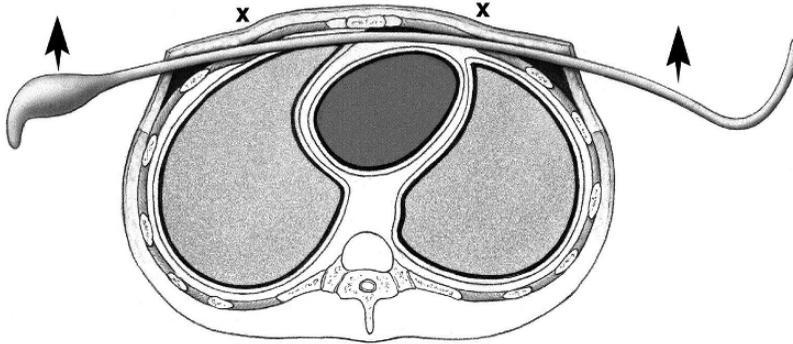


Fig. 10. Before turning the bar the sternum should be elevated gently but firmly by pulling the dissector anteriorly with the surgeon's and assistant's hands close to the site of entry into the skin (39).

Obtaining a chest radiograph on the operating table wise in redo patients, because the potential for unrecognized residual air collections (pneumothorax) is greater.

Thoracic epidural catheters are even more useful in redo patients because significant postop pain is to be anticipated (27).

Ostlie et al. have confirmed our findings that redo correction can be done safely with minimal blood loss and short operating time in patients who have undergone prior unsatisfactory open repair of pectus excavatum (41).

RECURRENT PECTUS CARINATUM

After open operation:

Pectus carinatum in reoperative surgery occurs after treatment with open or Ravitch repair. That operation has a low but nonzero recurrence rate. The frequency of recurrence must be sufficiently low that we are unable to uncover good figures on frequency, timing, or symptoms of recurrence. Lacquet reported in 272 patients with pectus

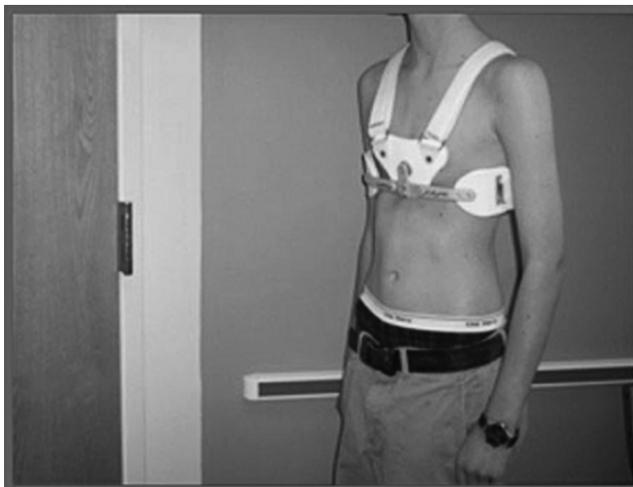


Fig. 11. Orthotic device used for correction of pectus carinatum by external compression.

carinatum, “Clinical results... were always good for protrusion deformities if enough deformed cartilages were resected.” Correia de Matos and colleagues report 24 patients with pectus carinatum with no recurrences (42). Some authors have suggested that recurrence can be minimized by resecting cartilages bilaterally, not just on the side of the sternum where they are visibly deformed. Shamberger and Welch reported that of 152 patients with pectus carinatum, “three required revision with additional unilateral lower cartilage resection for persistent malformation of the costal arch. All patients ultimately had a satisfactory result” (44).

After treatment with bracing:

Pectus carinatum has been treated with externally applied orthotics. The bracing has resulted in marked improvement in up to 90% of patients in whom it was applied (45,46) (Fig. 11). Recent improvements in the design of the brace have made it much smaller and more comfortable, which has dramatically improved compliance. But use of orthotics to treat recurrent pectus carinatum has not yet been reported (Fig. 12).

LESS FREQUENT CONDITIONS

Congenital Chest Wall Problems

Pectus excavatum has been reported to occur in a fraction of patients who undergo neonatal repair of sternal cleft primarily. Most of these patients have not required intervention (47).

Poland’s syndrome is reported to infrequently recur after surgical repair. Columbani states that this is likely because of the late age at operation (48) (generally after the teenage growth spurt). Reconstruction often involves repair of the chest wall defect and muscle transfer or free flap for coverage. Problems with nonunion of rib grafts and/or breakdown of the prosthetic repair can lead to lung herniation. These problems are sufficiently infrequent that treatment must be individualized (48). Remarkably, termination of pregnancy at 22-weeks gestation has recently been reported following antenatal diagnosis of Poland’s syndrome by ultrasound (49). Fokin and Robicsek report 20 operative cases without complication. They suggest a staged repair in younger patients but a single stage in adults, including muscle flaps and breast implants in female patients (50).

Repair of pentralogy of Cantrell has infrequently required reoperation, but should be managed by the same principles one uses for other recurrent defects, either thoracic or abdominal wall (48).

Cardiac Surgery and Pectus Excavatum

Several reports have described successful combined cardiac operations and open repair of pectus excavatum (51–54). These have included coronary artery revascularization, aortic graft placement, and repair of tetralogy of Fallot (53,55).

Long after open operation to correct pectus excavatum, successful sternotomy has been performed following Ravitch operation for pectus operation (54) and after sternal turnover operation (53).

Combined minimally invasive repair and repair of atrial septal defect and closure of patent ductus arteriosus has been reported recently (56). More importantly, there may be advantage to performing minimally invasive repair in patients with Marfan syndrome before skeletal maturity; then if aortic root dilatation occurs later

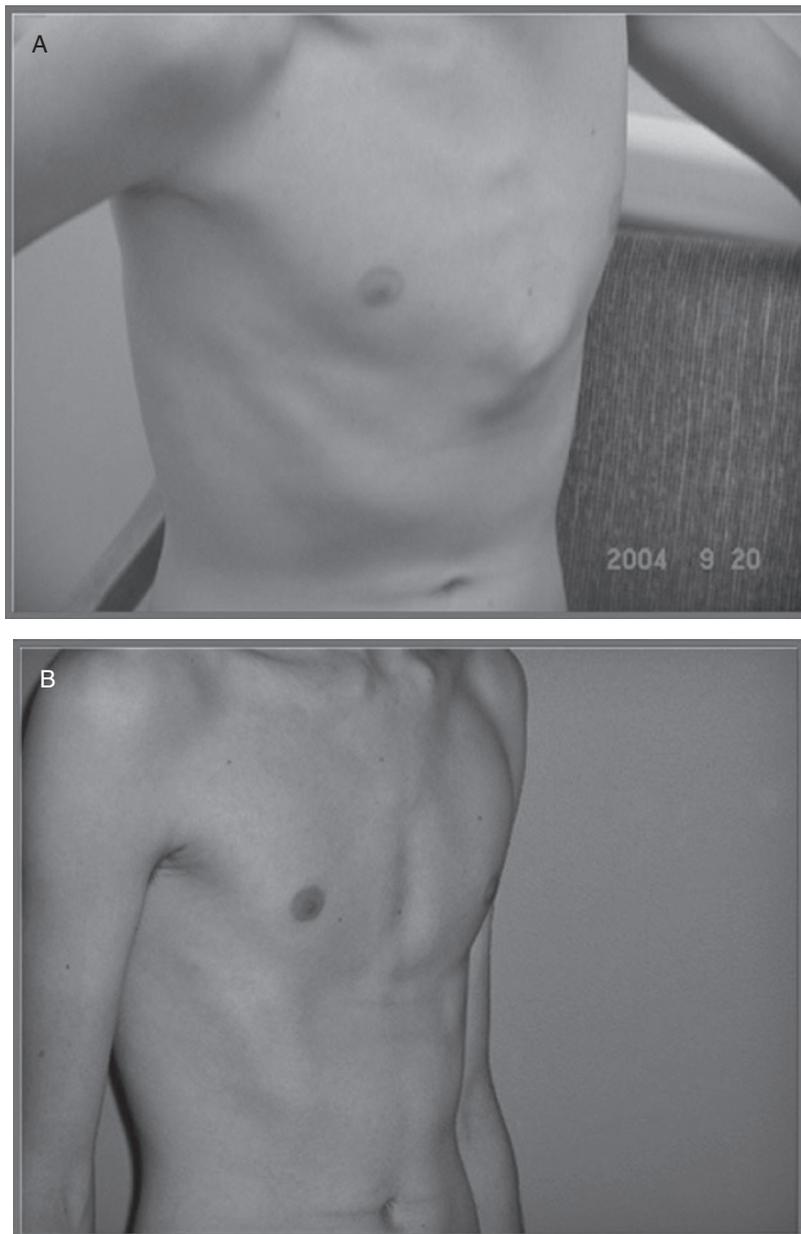


Fig. 12. Photographs of a patient before and after treatment with orthotic bracing for pectus carinatum.

in life, the repair of the aorta is simplified. If the pectus recurs, one could do both ops at the same time.

Unusual Associations With Recurrent Pectus Excavatum

Floating sternum, an unusual entity in which the costal cartilages failed to heal after resection, was treated at Johns Hopkins in seven patients following open operation

for pectus excavatum. Operation to stabilize the sternum entailed mobilization of the sternum and placement of two retrosternal support bars (57).

Pectus excavatum has been reported recently after chest wall resection for breast cancer. Management must be individualized to account for physiologic effects of the chest wall on cardiopulmonary function, oncologic considerations, age of the patient, and pliability of the chest wall. In patients under approximately 30 years of age, the Nuss should be considered. In older patients, the traditional open operation may have advantages (58–63).

Chest Wall Defects Following Resections

Chest wall resection for sarcoma or other tumors can result in deformity, which requires chest wall reconstruction (28). Infection of the sternum following sternotomy can require debridement or resection of the sternum. Rib graft harvest can result in deformity. Rib resection at childhood posterolateral thoracotomy is seldom performed now, but asymmetry of the chest and scoliosis were reported in later years in some patients when it was frequently used. It is difficult to produce fracture of the ribs by blunt trauma in children. However, after major blunt trauma, pseudoarticulation of injured ribs can occur. If the deformity is severe, reconstruction may be necessary (28). Use of prosthetic materials such as Gore-Tex® can be helpful. Defects are often covered with muscle flaps such as latissimus dorsi. Neither of these conditions occur with sufficient frequency to draw conclusions from the literature regarding approach beyond adhering to general chest wall repair principles.

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6

Complications of CDH Repair and Recurrent CDH

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OVERVIEW/INTRODUCTION

This chapter is dedicated to discussing those complications and conditions that require further surgical intervention in children who have survived the initial insult of congenital diaphragmatic hernia (CDH). The complexity of patients born with CDH creates a challenge to every surgeon who must bring these infants to the operating theater for the initial repair. In this instance, the operation is relatively straightforward, but the physiology is treacherous. Conversely, when situations arise requiring a subsequent procedure, the physiology is often, but not always, more settled, but the procedure is usually much more difficult. With increased survival has come increased morbidity and, not surprisingly, subsequent operations have become fairly common. In one report, 42% of CDH survivors required at least one additional operation. The need for reoperation is often predictable, as are the common indications (1,2). First and foremost is a recurrence of the hernia. The next most common indication is either feeding intolerance or gastroesophageal reflux (GER) or both, necessitating placement of a gastrostomy tube with or without fundoplication. Then come a variety of situations not unique to CDH, such as development of an incisional hernia, an intestinal obstruction secondary to adhesions or a persistent hydro- or chylothorax requiring drainage. There are several other conditions that are closely associated with CDH such as chest wall deformities, cardiac defects, undescended testicles, scoliosis, and others. However, because these really represent associated primary defects in which the operative fields

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are usually pristine, they will not be discussed fully in this chapter. Finally, there is the rare neonate that, having been successfully weaned from extracorporeal membrane oxygenation (ECMO), experiences a second insult requiring a second course of ECMO.

The aim of the chapter is to assist in diagnosis and guide the decision pathways when determining the need to return to the operating room in this unique patient cohort.

REHERNIATION

Incidence

The reported incidence of reherniation after a CDH repair has varied from 2–22%, but has been reported to be as high as 80% in a select group, where a patch repair and the use of ECMO therapy were concomitantly employed (3–8). Although the reason for such a wide discrepancy is unclear, it is almost certainly influenced by the size of the initial defect, the method of initial reconstruction, and the length of follow-up. It is not surprising that small defects, amenable to a relatively tension-free primary repair, are less likely to disrupt than larger defects, which require placement of a prosthetic patch. In a report by Van Meurs et al., 40% of CDH survivors with a patch repair eventually had a recurrence (7). Moss et al reported that within 3 years of initial repair, nearly 50% of patients who underwent a patch repair had developed a reherniation (8). Moss also noted that the majority of reherniations were owing to the patch pulling away from the posterior thoracic wall (8).

Presentation

A diaphragmatic hernia can reoccur at anytime, but the majority present within the first 24 months of life (6,9–11). Moss and colleagues found a bimodal peak incidence of recurrence, the first being between 1 and 3 months of age and the second between 10 and 36 months (8). Saltzman et al. reported that the age at time of recurrence ranged from 2 to 48 months, with the average being approximately 14 months (6). We too have noted that most reherniations occur prior to 24 months of age, and have tailored our surveillance program accordingly. We believe that the incidence of patch failure is probably extremely high, with most, if not all, repairs eventually disrupting. However, in many instances there is sufficient scar tissue formation to either prevent reherniation or to result in only a small degree of reherniation that may not be radiologically evident or clinically symptomatic. In fact, reherniation is often only discovered on routine chest radiographs, which is why we recommend regular surveillance chest radiographs monthly for the first 6 months, every 3 months until the age of 24 months, then at 30 and 36 months, and every 2–3 years until age 10 in those infants who underwent a patch repair initially (6). The surveillance is initially similar for primary repairs; however, in our experience the incidence of recurrence drops precipitously after 6 months, so we do not obtain annual chest x-rays (CXR) for surveillance purposes alone after that time.

Patients who present with symptoms related to a reherniation generally fall into two categories: those with pulmonary symptoms and those with gastrointestinal symptoms (8). Pulmonary symptoms are rare beyond the neonatal period but may present as tachypnea, persistent cough, development of aspiration pneumonia, and/or recurrent wheezing requiring increased use of bronchodilators. Respiratory distress in an older child is an ominous sign as tachypnea is usually a physiologic response to metabolic acidosis rather than pulmonary compromise. Development of any of these symptoms

should trigger a search for an occult recurrence. The gastrointestinal symptoms of reherniation are akin to those of a late-presenting primary CDH (12,13). They may include progressive oral aversion, new or increased feeding intolerance, increased vomiting or worsening GER, abdominal distension, or abdominal pain. Although these symptoms are not unique to reherniation, and are also seen with adhesive intestinal obstruction, the tempo is often much more indolent, unless the two conditions co-exist. Many of these symptoms are also associated with GER. However, it should be fairly simple to distinguish between these entities.

Diagnostic Tools

The diagnosis of recurrent CDH should be suspected based on history and physical exam, and can often be confirmed with a single chest radiograph. Examples of various reherniations evident on plain radiographs are demonstrated in Figs. 1–3. If the CXR shows evidence of a recurrence, adjunctive radiological studies will be necessary to help better understand the anatomy as regards the location of reherniation and the amount of abdominal contents involved. The studies of value are an upper gastrointestinal (UGI) with small bowel follow through, contrast enema, and/or contrast-enhanced CT scan of

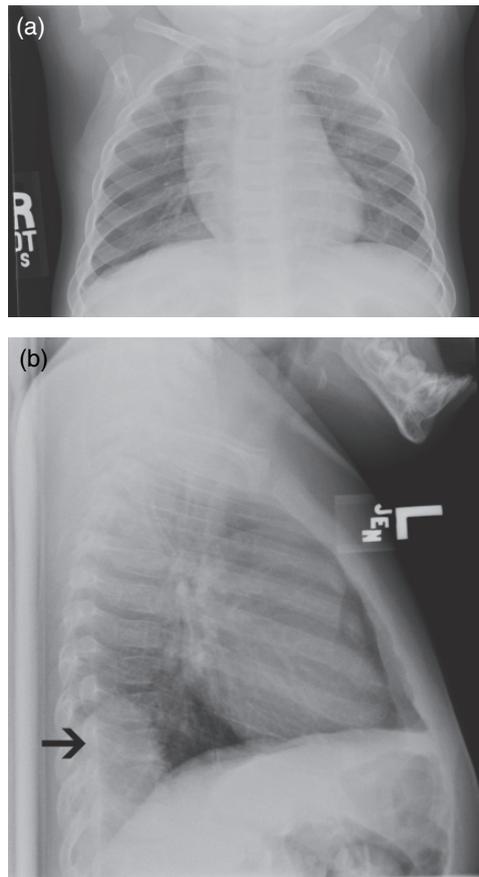


Fig. 1. (A) AP view right CDH recurrence 11 months. Unable to see recurrence. (B) Lateral view right CDH recurrence 11 months. Can see the recurrence marked with arrow.

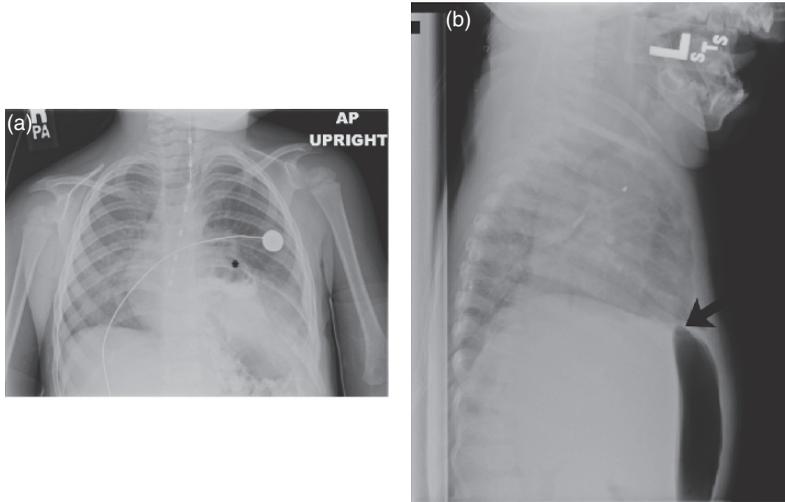


Fig. 2. (A) Recurrent left CDH with bowel in chest. Note the pH probe. The asterisk marks bowel in the chest—the tell-tale feature of the recurrence. (B) Lateral view recurrent left CDH. Arrow marks bowel in chest. Note again the pH probe and the presence of contrast in the herniated bowel located posteriorly.

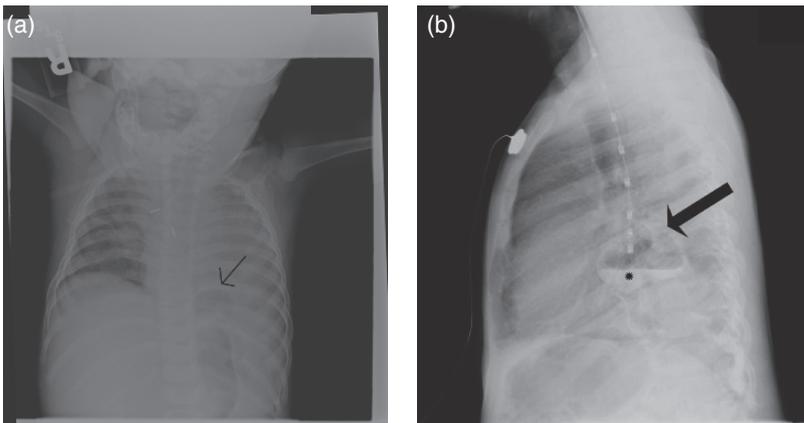


Fig. 3. (A) AP image paraesophageal hernia. Arrow marks recurrence. (B) Lateral view paraesophageal hernia. Arrow marks recurrence.

the chest and abdomen. In cases where reherniation is suspected on clinical grounds, but the chest film is equivocal, obtaining one of these contrast studies usually makes the diagnosis clear. With the advent of faster CT scans which have eliminated the need for sedation, CT has become our preferred diagnostic tool (Figs. 4A and B).

The Surgical Approach

Once it has been established that a reherniation has occurred, patients can be divided into symptomatic and asymptomatic groups. We believe that all recurrences will eventually necessitate repair. However, few are urgent, and deciding when the repair is to take place should be based on the size of the recurrence and the severity of the associated symptoms. The symptomatic patient should be repaired as soon as it is

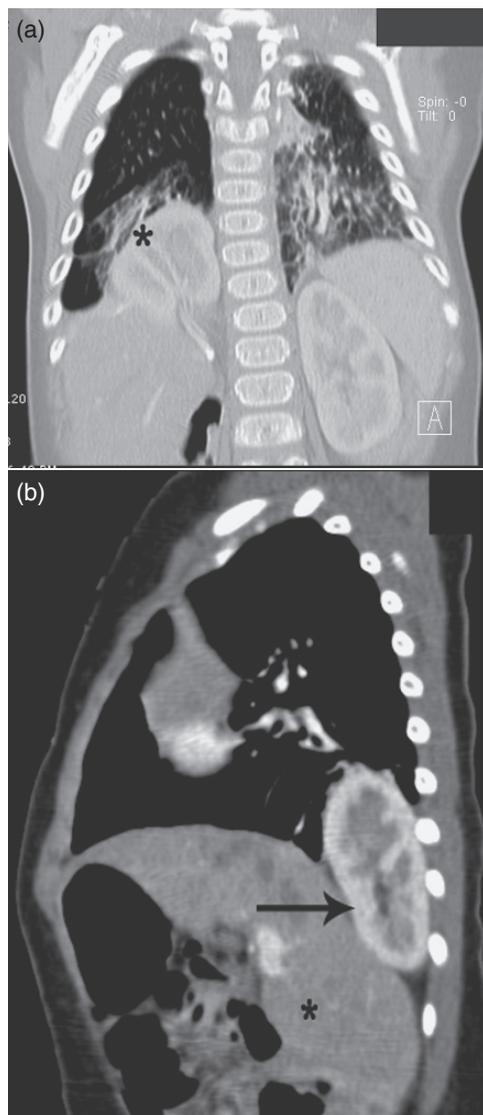


Fig. 4. (A) Coronal CT reconstruction right CDH recurrence. Arrow marks reherniated kidney on right. (B) Lateral CT reconstruction right CDH recurrence. Arrow marks kidney in thorax and asterisk marks liver.

feasible, as should large herniations. But the asymptomatic patient with a small defect may be treated expectantly for several months, or in some cases years, as long as they are carefully monitored. The justification for delaying repair in asymptomatic patients includes allowing time for improvement in pulmonary and nutritional parameters. In addition, from a practical standpoint, repairing or replacing a patch simply resets the clock until the next recurrence. So allowing the asymptomatic child to grow might allow for a more secure repair later and fewer repairs in total.

A recurrent CDH can be approached via either the abdomen or the chest. Theoretically, the repair can be performed either via open or minimally invasive technique, but as a practical matter, the latter is probably only appropriate for the most minimal

of defects. The choice of approach may be influenced by other circumstances such as whether a fundoplication and/or gastrostomy tube will also be performed at the time of the repair, or whether an incisional hernia is present.

The transthoracic approach is favored by some, as it is more likely to provide a virgin operative field in patients who initially underwent a transabdominal repair. But if the child has evidence of pulmonary hypertension or inadequate lung reserve, the transthoracic approach may not be reasonable. Furthermore, many recurrence sites are posteromedial, and we have found this difficult to address via the chest. If the patient also needs a fundoplication, the transthoracic approach may not be ideal. We usually prefer the transabdominal approach which is often a bit more challenging and time consuming but, allows us better visualization of the defect itself, and the organs and structures that must be preserved. It also provides ready access to perform a fundoplication and/or gastrostomy tube should one be needed. The laparoscopic approach is likely to be very difficult and in most cases should probably be avoided unless the initial repair was transthoracic.

Repair Techniques

In order to plan the most appropriate repair technique, the mode of failure of the initial repair must be ascertained. In our experience there are four primary modes of failure: (1) failure of a primary repair; (2) failure of the medial aspect of the repair; (3) recurrence at the posterior-lateral margin; and (4) development of a paraesophageal hernia. Anterior and lateral failures are rare, probably because the anterior lip of diaphragm seems to be the thickest muscle, and laterally, owing to the absence of any diaphragm at this location sutures are usually placed around ribs. The location of failure is important, as it will dictate subtle but important differences in timing and technique.

In the case of a failed primary repair, the cause of recurrence is often a high-tension initial repair in which the sutures simply could not hold the tissue together. In these cases, another primary repair is usually impossible and a new prosthetic patch should be placed. The new patch may be domed or reefed to allow room for the child to grow. A novel dome technique was described by Loff and coworkers in 2005. They utilized a flat polytetrafluoroethylene (PTFE) patch that was folded into a cone shape and held together by a few simple stitches. The cone was then secured to the rim of the remaining diaphragm with simple stitches, such that the rough portion of the patch abutted the diaphragmatic rim (14). Other techniques for pleating a patch with absorbable sutures intended to release in response to tension have also been described (8). We have tried both techniques several times but do not have long enough follow-up to comment on their utility. We tend to favor Gore-Tex[®] patches (PTFE); however, a study by Grethel and colleagues reported no difference in outcomes or complication between Surgisis[®] (intestinal submucosal patch) and Gore-Tex[®] patch repairs (11).

When the initial patch repair fails, it may do so in the early postoperative period, or weeks, months, or years after the initial repair. An early recurrence usually denotes some technical error and the patch can usually be simply replaced or resutured. With a late medial recurrence, when scar tissue has had ample time to form, our preferred approach is to augment the existing patch by placing a new medial portion rather than to replace it. In this situation, the spleen, now well adhered to the lateral aspect of the diaphragm and prior patch, does not need to be mobilized, and in fact should not, as long as the posterior-lateral aspect of the repair is intact. To do so is not only unnecessary; it significantly increases the difficulty of the dissection, and not

uncommonly will result in a splenectomy. When repairing medial defects in a recurrent left-sided hernia, we augment the patch using a “C” or crescent shaped patch nestled around the GE junction. Medial defects on the right are difficult because the inferior vena cava (IVC) comprises most of the medial wall. The reoccurrence is almost always posterior in these situations. In this instance, a secure posterior stitch placed around the rib medially, and an anterior medial suture into muscle or around the rib, will usually prevent herniation, principally because the liver will protect the weak medial wall. We continue to observe some small right medial recurrences in which only a small portion of liver has herniated, as it is often just as secure as a patch.

When the patch has torn away from the posterior-lateral aspect of the thoracic wall, it is usually because of trusting the integrity of an inadequate rim of diaphragm at the initial surgery or mistakenly placing posterior sutures into the retroperitoneal fascia and/or Gerotta’s fascia rather than into diaphragmatic muscle or around a rib. This is probably the most common mechanism of failure we have seen and can be avoided by being sure that the posterior sutures are placed into the thoracic wall. In this scenario, it is common for the kidney to herniate into the chest, thereby creating a “retroperitoneal” herniation and not a true reherniation. In fact, at reoperation it is not unusual to find the patch repair, itself, intact and the kidney herniated posterior to it. We prefer to repair this mode of recurrence by dissecting free the remnant diaphragm and patch and re-anchoring, augmenting, or replacing the patch with suture around the ribs. If a lateral recurrence is present in a right-sided defect, the liver will likely be scarred in place medially, and, as with the spleen on the left side, one should not need to mobilize the liver. The patch should be augmented rather than replaced.

The final mode of “recurrence” that we have observed, which always occurs late, is the paraesophageal hernia. This occurs when the child “outgrows” the original patch and the patch does not tear away from the lateral chest wall. In this situation, the thin rim of medial diaphragm gets pulled laterally by the inflexible patch, resulting in a new paraesophageal hernia and worsening GER. Immediate repair is seldom necessary and timing should be guided by symptoms. Again, because of the large amount of scar tissue present at the time of repair, we advise augmenting the medial portion, rather than replacing the entire patch. We also feel that it is prudent, if not mandatory, to perform a fundoplication procedure at the same time as the process will invariably re-occur.

A variety of novel approaches to repairing a recurrent hernia both with prosthetic patches and with muscle flaps have been described. One type of “patch” repair utilizes a Bard Mesh plug, similar to those used in inguinal hernia repairs (6). Saltzman and coworkers described this technique in 2001, employing a transthoracic approach for placement of the mesh plug in five patients with a posterior-medial recurrence (6). The average follow-up was approximately 14 months (range was 1–33 months), with no recurrences. This approach may be reasonable in small reherniations. Several groups have described various muscle flaps. Sydorak and colleagues reported the use of a reversed latissimus dorsi flap (LDF) in seven patients, three with a recurrence and four who were considered to have significant chest wall excursion restrictions secondary to a patch repair (15). The flap was rotated, with the neurovascular bundle intact, into the thorax via a thoracotomy. To assist in the function of the LDF, the thoracodorsal nerve was anastomosed to the phrenic nerve (15). In four of the seven children, there was evidence of physiologic diaphragmatic function (15). Despite attempts to improve upon the repair of a recurrent hernia, placing an additional prosthetic patch remains

the mainstay of therapy. The “Holy Grail” of CDH repair is a patch that can grow and remodel with the child. Recent large animal data suggest that CDH repair with autologous, engineered tendon patches is possible. These grafts can be made available at birth, as they can be engineered in parallel to gestation, from cells normally present in, and harvested from, the amniotic fluid (16,17). A proposed protocol for the first clinical trial of this novel therapeutic concept is currently under review by the Food and Drug Administration (FDA). However, until pre-engineered bioprosthetic patches become a clinical reality, we will continue to rely on the synthetic devices currently available.

GER

Etiology

Physiologic GER is common to many infants and thus can pose a challenge in recognizing the transition to a pathologic process in patients with CDH. In the first several weeks of life, GER, also called “spitting-up,” is common (18,19). As many as 60–65% of infants with GER are symptom-free and the GER has resolved by 18–24 months of age without any medical interventions or therapy (20). When symptoms persist or the GER is so severe that complications ensue, then it is considered pathologic and the child is diagnosed as having gastroesophageal reflux disease (GERD). The North American Society for Pediatric Gastroenterology and Nutrition estimated that approximately 7% of children ages 3–9 years old have GERD, and Nelson et al. reported a 5% incidence of daily reflux in 10- to 12-month-old infants (21,22). It wasn’t until the 1990’s that the literature began to recognize GERD as an entity associated with increased morbidity in patients with CDH (23–26). The incidence of GERD in patients with CDH ranges from 20–72%, depending on the series (23,27).

The current concepts regarding the etiology of GERD revolve around the function of the lower esophageal sphincter (LES), the competency of which is a coordination between the stomach, esophagus, and diaphragm. Lack of competency at any level leads to the development of GERD (28,29). In children with CDH, several mechanisms have been proposed to explain the higher rates of GERD observed in this subset of patients. Stolar et al. noted ectatic esophagi, with evidence of dysmotility, in most of the CDH patients with GERD, suggesting that perturbation of esophageal peristalsis may, in addition to the diaphragm, be an important component in GERD development (26,30). Kieffer et al., who reported a 62% incidence of GERD in their CDH survivors, proposed that fetal swallowing and kinking of the esophagus caused by bowel contents in the chest may play a role in the development of an abnormal esophagus (27). It has also been postulated that operative disruption of the angle of His may also play a role in promoting GERD by creating an obtuse angle as a result of the repair. Similar observations have been made in patients with hiatal hernias who demonstrated an increase incidence of GERD (19). As seen in patients with hiatal hernias and in patients who have had a tracheo-esophageal fistula repair, the GE junction may also be located abnormally high, thereby allowing the negative intrathoracic pressure created with respiration to promote GERD (19,31). With the altered diaphragmatic structure in CDH, the state of the diaphragmatic crus and the surrounding muscle undoubtedly also play a role in the development of GERD, preventing the diaphragm from maintaining an appropriate LES pressure (27). Finally, delayed gastric emptying, often seen in patients with CDH, will aggravate any pre-existing GERD (18).

Although the reasons why GERD is so prevalent in CDH are fairly well understood, identifying predictors of its development have not been easy. Koot et al. prospectively followed 31 CDH survivors and found that the rate of GER decreased over time from 52% at 6 months to 35% at 12 months, but were unable to identify any single, clear predictor as to which child would progress to GERD (23). The study looked at duration of mechanical ventilation, primary versus patch repair, preoperative position of the stomach, presence or absence of a hernia sac, and gestational age at birth. Other studies have suggested predictive factors, but these have all been retrospective. Kieffer et al. found that an intrathoracic location of the stomach through the hernia defect was correlated with an increased chance of developing GER (27). The CDH Study Group, in 2006, reported that 72% of patients with agenesis of the diaphragm went on to develop GERD versus only 42% of patients who had some remnant of diaphragm (32). Other studies have correlated the use of a patch and/or ECMO as predictors of subsequent GERD (5,12,33). Despite the lack of consensus regarding which predictive factors best identify the CDH survivors who will develop GERD, the end result is unchanged—GERD is a common and significant co-morbidity for this group of children.

Diagnosis

A certain degree of GER exists in most patients with CDH, so in our institution most are treated empirically with proton-pump inhibitors such as omeprazole, or histamine-2 blockers like ranitidine, and sometimes prokinetic agents such as metoclopramide before any formal evaluation is pursued. However, clinical signs of persistent reflux despite optimal medical therapy should prompt a more formal evaluation for GERD. The symptoms are varied and often involve more than just the GI system (Table 1). The most common complaint is continued emesis, which is present in 90% of the infants and children with GERD (20). Additional GI symptoms may include feeding intolerance, whether it is oral or via a gastrostomy tube, regurgitation and rumination, and weight loss. Pulmonary symptoms are also common and particularly worrisome

Table 1
Signs and Symptoms of Gastroesophageal Reflux Disease

Vomiting
Rumination
Dysphagia
Feeding intolerance
Halitosis
Failure to thrive
Recurrent aspiration pneumonia
Bronchitis exacerbation
Chronic cough
Recurrent sinus infections
Recurrent otitis media
Irritability
Apnea
Acute life-threatening event
Anemia

in patients with CDH who already have lungs compromised by pulmonary hypoplasia (4,10,34). These symptoms may include recurrent bronchitis, lobar pneumonia, worsening bronchopulmonary dysplasia, chronic cough, and persistent wheezing despite the use of nebulizers, recurrent otitis media, sinusitis, and occasionally apnea leading to sudden infant death or an acute life-threatening event (18,21). In patients with CDH, it is often difficult to assess whether pulmonary symptoms are caused by GERD or just part of their primary pulmonary disease (35). Further complicating the ability to diagnose GERD in patients with CDH is the fact that many patients with CDH display some degree of feeding difficulty. However, continued loss of weight or a drop down of a percentile on the appropriate growth curve are clear signs of GERD that has failed medical therapy (4,12,21,34).

Once a child is suspected of having GERD, the diagnostic toolbox includes noninvasive radiologic studies such as an upper GI series (UGI), to more invasive techniques such as endoscopy and esophageal biopsy. However, the major components of the evaluation include an UGI and 24-hour pH monitoring, which is considered the “gold-standard” for evaluating GERD (18,20,36–38). The UGI can provide valuable information about the anatomy of the esophagus, the height of refluxed contents, and can also evaluate gastric emptying. As already noted, Stolar et al. found a strong correlation between GERD and the presence of an ectatic esophagus, and it is well documented that patients with tracheo-esophageal fistula repairs and with hiatal hernias, where the LES is above the diaphragm, are all at high risk of having GERD (19,26,30,31). The UGI has a positive predictive value for GER of approximately 80–82%, but the sensitivity and specificity are inconsistent, from 31–86% and 21–83%, respectively (21). A nuclear medicine study that has been reported to be useful involves feeding the infant radio-labeled juice and looking for tracer in the lung fields and in the mediastinum to quantify the severity of reflux (18,20,21,39). A test that we use frequently is the 24-hour pH probe and sleep study, as we believe that it is very effective at recording reflux events and any associated respiratory compromise such as apnea (20,21,37,38). The study entails the securing of a pH probe at the junction of the middle and distal one-third on the esophagus with the tip of the probe located 2–3 cm above the LES. The number and duration of reflux events ($\text{pH} < 4.0$), are recorded and reconciled with the patient’s position and presence or absence of symptoms at the time of the event (23,40,41). A similar study to the 24-hour pH probe is a 24-hour impedance probe, which is placed into the stomach and has sensors along its length to assess the pH at various points in the esophagus as well as the stomach. This allows the examiner to assess not only acidic, but also nonacidic reflux events. This is a newer technique, but there is concern as to whether the probe passing into the stomach might affect the usefulness of the data obtained because it is stenting the LES open, and “standard values” are not yet clear (42). Finally, the most invasive test is esophagoscopy, which allows direct visualization of the esophagus with biopsies. This is generally reserved for cases in which the results of other tests are inconclusive.

Management of GERD in CDH

Nonoperative management consists of thickening oral feeds, often with cereal, small frequent meals, sleeping on a wedge, decreased exposure to secondhand smoke, and pharmacologic therapies such as histamine-2 blockers, proton-pump inhibitors, and prokinetic agents (18,20,34,40). Stolar et al. reported that in his CDH cohort with GERD, all responded to medical therapy and did not require a surgical antireflux

procedure (26). However, this has not been our experience. We observed that approximately 15–20% of our patients with CDH will fail nonoperative management for GERD and will require surgical intervention (34). In other studies, it has been reported that 9–23% of CDH survivors will need an antireflux procedure (23,27). It is important to recognize that unlike normal infants with physiologic GER who will improve with time, infants with CDH and GERD usually worsen. This is especially true of patients with patch repairs, which predispose to development of paraesophageal hernias as the child grows.

Surgical Approach

The surgical management consists of a fundoplication with or without gastropexy. In the planning of the operation for the patient with CDH, there are several factors to be considered. First is to assess if there is evidence of reherniation or development of a paraesophageal hernia. Second, it has been our experience that postoperatively most children will not take adequate calories orally and thus a gastrostomy tube is a useful adjunct as a route to provide adequate caloric intake (12). We almost always place a gastrostomy in our patients with CDH during fundoplication. Finally, whereas in most children a laparoscopic approach is favoured, in the majority of patients with CDH this would be difficult if the CDH repair was done via a transabdominal approach so we most often will employ an open approach.

Repair Technique

There are four major types of fundoplications: Nissen, Thal, Boix-Ochoa, and Toupet. The Nissen fundoplication is the only one in which the stomach is completely wrapped around the esophagus (360° wrap) (44). The Nissen fundoplication is also the most common type of fundoplication used in both adults and children (18). The other three procedures are all variations of a partial wrap. The Boix-Ochoa and Thal fundoplications are both anterior, 180° wraps with the major difference the fixation of the stomach to the left diaphragmatic undersurface with the Boix-Ochoa technique. The Toupet wrap is similar to the Boix-Ochoa technique in the fixation of the stomach to the diaphragm with three to five single stitches, but the Toupet is a posterior 270° wrap. Although the data is sparse in the pediatric literature, in adults a Nissen fundoplication is the procedure of choice unless there is evidence of abnormal esophageal motility, and then a partial wrap is the procedure of choice (45). A recent European study evaluated the efficacy and complication rates between laparoscopic Nissen, Toupet, and Thal fundoplications and found no difference in the rates of dysphagia or other complications, suggesting that each technique was equally efficacious in alleviating GERD (46). In light of the paucity of data, we tend to favor a Nissen fundoplication or a Toupet wrap if there is a suggestion of poor esophageal motility, with a modification we have coined “Gulliver’s Travels.” As described by Hendren in 1980, in both techniques the crus of the diaphragm, once freely dissected, are brought closer with a simple stitch. The esophagus is then sutured to the diaphragmatic hiatus with a series of five (4-0 or 5-0) proline sutures. The wrap is made around an esophageal bougie, the length of which is based on the size of the patient ery. We also modify the suturing of the stomach by using multiple rows of fine interrupted stitches placed from stomach to esophagus instead of the standard three. Finally, we employ multiple sutures to affix the stomach to the diaphragm or patch. Consequently, the wrap, like “Gulliver,” is

held together by up to 40 small sutures which individually have very little integrity but collectively are quite strong (47). It is seldom necessary to mobilize the spleen to gain access to the medial portion of the diaphragm as long as it was appropriately positioned at the initial operation.

This technique has the advantage of distributing the tension across multiple tether points, with no single suture being critical to the integrity of the wrap. It has the disadvantage of not being practical to perform laparoscopically. In our experience, subsequent failure of the wrap in patients with CDH has been rare utilizing this technique.

GASTROSTOMY TUBE

Indications

Nutrition, even in patients without reflux, is a large concern for the CDH survivors. Several series have reported that between 20 and 56% of all patients with CDH fall below the 25th percentile for weight, despite aggressive nutritional support (1,3,12,33). The reason for this is probably multifactorial. Patients with CDH, especially in infancy, have an increased work of breathing and require the use of accessory skeletal and abdominal muscles to maximize ventilation to compensate for the lack of a fully functional diaphragm. This is especially true for those with a patch repair. Many of these patients also have prolonged intubations and have some degree of oral aversion, and some have difficulty swallowing. Poor oral intake may also be a sign of severe GERD (4,7,34). It has been our policy to defer placement of a gastrostomy tube as long as the patient remains on his or her growth curve and as long as they are above the fifth percentile. However, in our experience, as least 25% of patients will fail and will need supplemental nutrition via a gastrostomy tube (12). We have found a strong correlation between the need for a gastrostomy and the use of a prosthetic patch (5). The length of intubation and the need for home oxygen also correlate with poor oral intake and feeding intolerance (39).

Surgical Approach

There are several methods of placing a gastrostomy tube in this population. Three basic options exist: open placement, laparoscopic placement, and percutaneous endoscopic placement (PEG). Each of these techniques has advantages and disadvantages. The majority of children have the CDH repaired via a transabdominal approach, and there are likely to be extensive adhesions. Placing a PEG would carry a significant risk of injuring colon or small bowel in the patient with CDH, because malrotation and an underdeveloped abdominal cavity predispose the small bowel to occupy every available space, including over the liver and anterior to the stomach. A new technique coined "SLiC" was recently reported by Ponsky and colleagues as an alternative for patients in whom a traditional PEG is not a suitable option. This technique uses endoscopy to insulate the stomach, and a finder-needle is then passed through the anterior abdominal wall into the stomach while being visualized by the endoscopist. After the local anesthetic is injected, a silastic tissue expander prosthesis (STEP) trocar is inserted directly into the stomach while being visualized (48). This avoids pulling the gastrostomy tube down the esophagus, but offers no particular advantage when the concern is bowel injury.

A laparoscopic approach, for the same reasons as the PEG, is often not a favorable option. However, if the patient did not have a transabdominal CDH repair, then the laparoscopic approach may be a viable option if an antireflux procedure is also to be performed. Because most patients have had a transabdominal repair of their diaphragmatic defect, an open gastrostomy tube is still the most common approach for us. We usually employ the Stamm technique, in which the feeding tube is placed into the anterior gastric wall near the greater curvature of the stomach and secured by two purse-string sutures. The stomach is then fixed to the anterior abdominal wall surrounding the tube exit site (49). It has been our experience that most patients will no longer need the gastrostomy tube by the time they reach school age. Tube removal and closure of a gastrocutaneous fistula in patients with CDH follow the same algorithm as any other patient.

ECMO RECANNULATION

Indications

Approximately one-third of our CDH survivors have had ECMO as part of their treatment course. Meehan and colleagues analysis of the Extracorporeal Life Support Organization (ELSO) data through 2002 reported that 1.25% of 16,450 patients in the ELSO database required more than one ECMO run. Indications for a second ECMO run were pulmonary in nature in 82% of the cases. Patients with CDH accounted for 62.5% of those cases (50). Indications for a second ECMO run may include sepsis, worsening pulmonary hypertension, and right heart failure. In order to justify a second ECMO run there should be an identifiable, correctable, acute insult that has occurred to an otherwise recovering patient. Failure to maintain adequate gas exchange, after successful ECMO decannulation (the dwindles) is not a reasonable indication.

Technique

The challenges associated with recannulation vary depending on the technique used (veno-arterial [VA] versus veno-venous [VV]), how the involved vessels were dealt with during the first decannulation, and how much time has passed since the initial ECMO run. In general, it is usually fairly straightforward to recannulate through the previously utilized vessel or vessels within the first several days after the initial ECMO run. Care should be taken to backbleed the vessels to assure patency before placing new cannulas. Embolectomy catheters are useful if thrombus is present, and occasionally Hegar dilators can be helpful if the vessel is scarred or has become stenotic. The catheters, dilators, and cannulas must be passed carefully, as arterial dissection is a very real risk.

Although we have been successful in recannulating vessels as late as one month after the initial run, it is probably unwise to attempt this beyond two weeks unless the vessels were reconstructed and are known to be patent. Beyond this period of time, the transthoracic approach is preferable, especially in neonates in whom the femoral approach is impractical.

Complications

Analysis of the ELSO data also shows that the rate of complications increases by 20.6% from the first to second course of ECMO (50). These complications include

mechanical, (i.e., difficulty with the cannula placement and achieving adequate flow rates), a higher incidence of neurological deficits, and a much higher incidence of multisystem organ dysfunction or outright failure. Consequently, very careful consideration must be given to the patient's realistic chances of recovery prior to embarking on a second run.

MISCELLANEOUS

Chylothorax

Chylothorax is reported to occur in up to 10% of patients with CDH and, when present, is quite difficult to eradicate and significantly complicates postoperative management (51–53). It appears to be more common in patients with a sac than without one and may be caused by disruption of lymphatic channels during sac resection (52,54). The initial treatment of a chylothorax is always nonoperative, which is successful in nearly all cases (52). The placement of a chest tube is warranted in cases where the effusion is causing clinically significant respiratory compromise. These can often be done under ultrasound guidance to achieve optimal placement. In addition to chest tube drainage, enteral feeds are either stopped completely or the formula switched to one containing medium chain fatty acids only. Beghetti et al. reported that dietary management and drainage were effective in 80% of the pediatric patients in their series (55). Recent studies also favor the addition of Octreotide, a somatostatin analog, to diminish chylous drainage if the chylothorax is not diminished with dietary changes alone (56,57). Octreotide may be administered as a continuous intravenous infusion, or in divided doses via subcutaneous injections. We have found it to be very useful in cases where dietary manipulations alone failed to resolve this problem.

If all nonoperative management fails, the last option is to attempt surgical ligation of the thoracic duct. In some cases of congenital chylothorax, pleurodesis is also attempted, but because of the pulmonary hypoplasia present in the patient with CDH, this is unlikely to work. Unfortunately there is no clear consensus in the literature as to how long nonoperative management should be employed, or when to abandon it in favor of surgical options. However, the success rate of nonoperative management has been extremely high in our experience.

Small Bowel Obstruction

When CDH survivors present with abdominal pain, constipation, abdominal distention, emesis, and/or other signs and symptoms of a bowel obstruction, the first step is to obtain plain films to look for a recurrence of the CDH. The patient who has a bowel obstruction, regardless of etiology, should have a nasogastric tube placed for decompression, intravenous fluids administered, and serum chemistries evaluated for signs of dehydration and metabolic acidemia. Lund and colleagues reported that the incidence of bowel obstruction requiring operative intervention in this population was slightly less than 20% (58). The principle causes of obstruction in their series were adhesions, midgut volvulus, gastric volvulus, and recurrent CDH. Operative decision making and treatment do not differ from those in any other pediatric patient other than to determine whether the obstruction is caused by reherniation.

Chest Wall and Spinal Deformities

The most common chest wall deformity in this population is pectus excavatum. It has been our experience that it does not require operative repair in the majority of cases. There are few cases where the pectus deformity is so severe that there is an element of respiratory compromise owing to restriction by the deformity (*see* Fig. 5). Each patient needs to be assessed on an individual basis with pulmonary function tests, cardiac echocardiograph, evaluation of chest wall mechanics, and anatomical evaluation utilizing chest CT. The selection of operative approach does not differ from

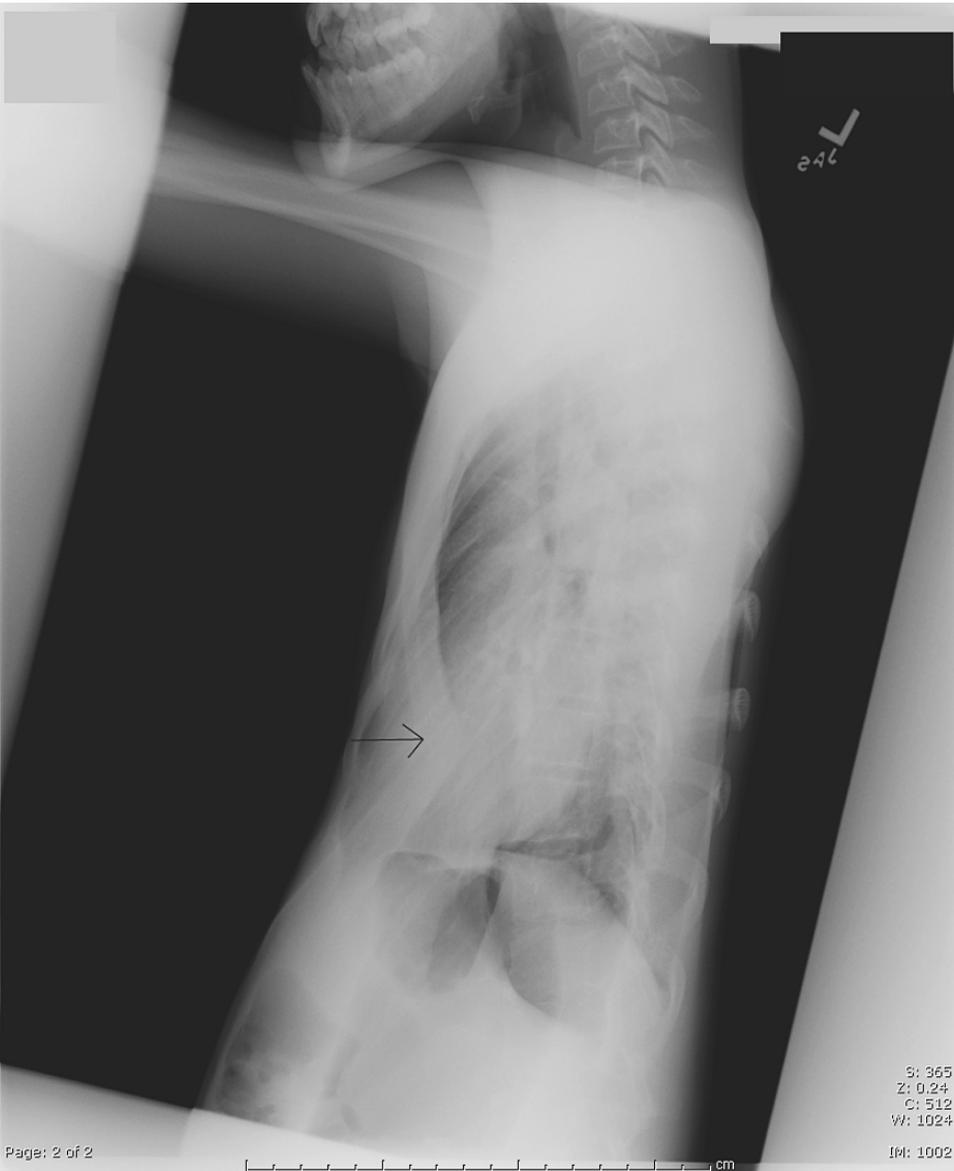


Fig. 5. Pectus. Lateral view of pectus, with arrow marking inferior edge of sternum.

any other patient with pectus with the exception that placement of a transthoracic bar may be complicated by pulmonary adhesions.

Scoliosis is the other common skeletal deformity in this population. We recommend assessing the spine at each visit during childhood. A plain chest radiograph can be of great assistance in identifying mild scoliosis. Although common in patients with CDH, very few of our patients have required spinal correction.

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7

Reoperation for Jeune's Syndrome

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CLINICAL PRESENTATION OF JEUNE SYNDROME

Jeune syndrome is an autosomal recessive disorder that is characterized by very small thoracic volumes, which can lead to progressive respiratory insufficiency and death (1,2). In addition to being anatomically small, the short, broad ribs are horizontally oriented, resulting in a very stiff chest wall. Jeune syndrome is a form of dwarfism, so the patients are also small overall, but most severely so in the thorax (Fig. 1). There are parenchymal lung changes associated with Jeune syndrome, consisting of patchy atelectasis and bronchial crowding, but it is not clear whether these changes are secondary to chronic crowding of normal lung or are a primary characteristic of the disease (3). There is an association with renal and hepatic disorders; although, in our experience, most of the time the disease is limited to the skeleton. They appear to be mentally unaffected, and most patients are quite bright. Other asphyxiating thoracic dystrophies (ATDs) can look similar from a skeletal perspective, but have other associations, such as Barne's syndrome, which is associated with laryngeal stenosis.

Jeune syndrome is actually a spectrum of disease, and over the years we have encountered all degrees of severity. On the mild side of the spectrum, we have seen adult patients with the stigmata of the disease who are structurally very small, but



Fig. 1. Picture of patient with Jeune syndrome, illustrating small “pinched in” thorax.

are able to carry out the normal activities of daily living and even some mild athletic activities. At the other end, we have seen infants requiring significant mechanical ventilation from the moment of birth, and who developed progressive respiratory insufficiency and died after a brief time. In some instances, significant tracheomalacia or bronchomalacia can compound the respiratory challenge.

The term “acquired Jeune syndrome” has been encountered from time to time, and is a very different situation with a different etiology. This was described by Haller (4) as the result of early and aggressive repair of pectus excavatum, such that the rib growth plates are affected and result in a small thorax relative to the rest of the body. However, the ribs are normally oriented with normal obliquity, and the degree of impairment is usually rather mild compared to classic Jeune syndrome.

DIAGNOSTIC EVALUATION

To completely evaluate a patient with Jeune syndrome, a number of studies are required. In addition to the history and physical examination, a routine chest roentgenogram will provide a baseline for future examinations, and rule out any acute infiltrative process that could acutely influence findings in the more sophisticated studies.

High-resolution computed tomography (HRCT), accomplished under deep sedation with volume and respiratory control to obtain motionless images at end-inspiration and end-expiration, can provide valuable information (5). Morphologic analysis will demonstrate the typical “clover leaf” deformity of the chest characteristic of Jeune syndrome (Fig. 2).

Measurements of the trachea and bronchi in inspiration and expiration, and their ratios, can provide some objective measurement related to the existence of tracheomalacia or bronchomalacia. Volumetric measurements of pulmonary parenchyma can also be made in inspiration and expiration, providing a measure of vital capacity.



Fig. 2. CT image of the thorax of a patient with Jeune syndrome, illustrating “clover leaf” deformity.

Infant pulmonary function studies using the specialized raised volume rapid thoracic compression and body plethysmographic techniques described by Castile (6) can provide a basis for long-term follow-up and evaluation. All studies are expressed in percentages of expected values. Because of the dwarfism, sitting height is used to generate expected “normal” values for comparison.

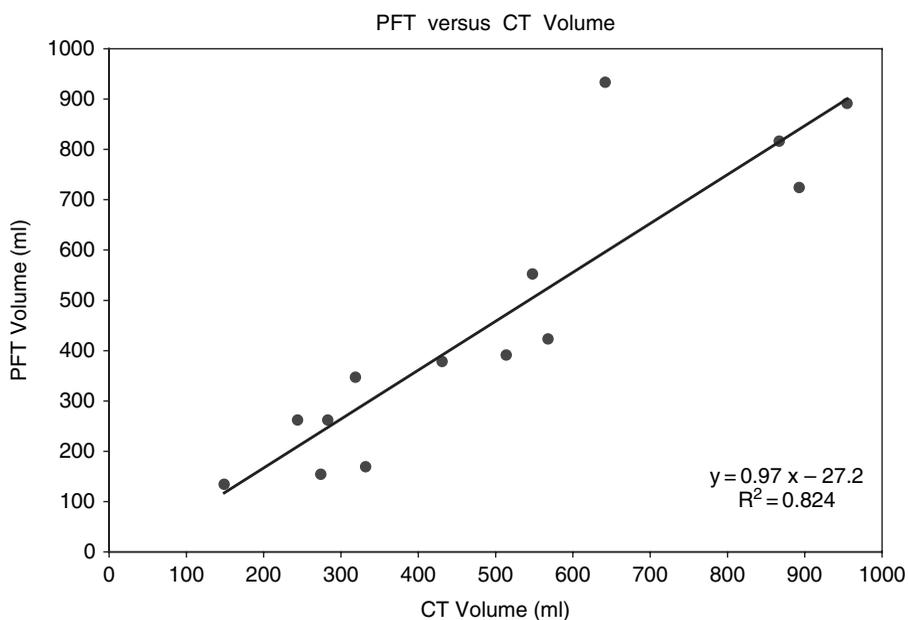


Fig. 3. PFTs, total lung capacity as measured by pulmonary function tests; CT volume, total lung capacity as measured by CT. All values are in milliliters.

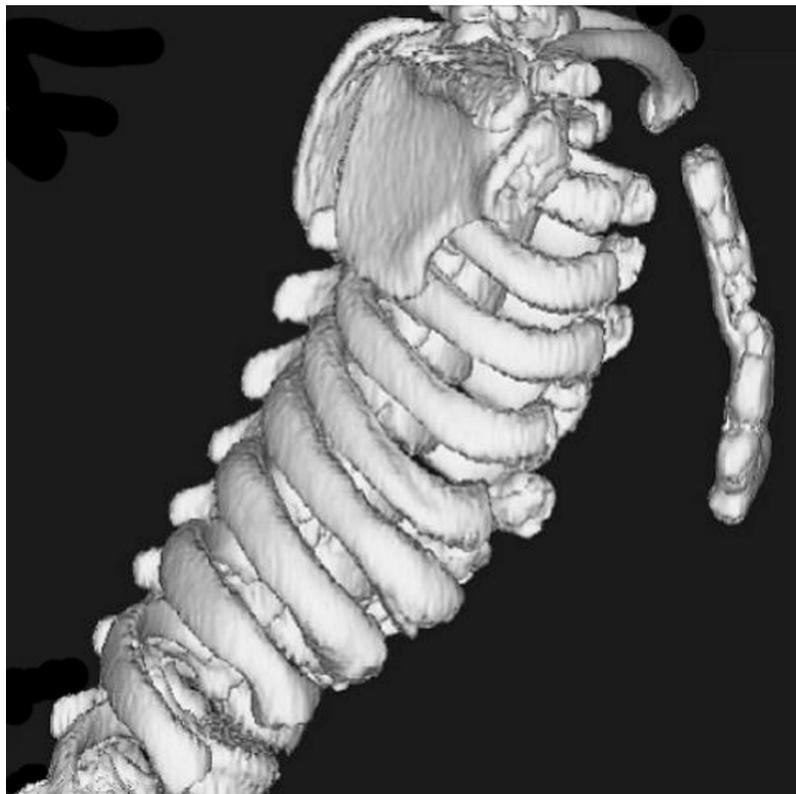


Fig. 4. 3D reconstruction of a CT scan of the bony thorax of a patient with Jeune syndrome.

Comparison of the radiological methods with infant pulmonary function studies' methods of measuring vital capacity shows a remarkably close correlation in our hands (Fig. 3).

3D reconstruction of tomographic images of the pulmonary parenchyma can be useful to document areas of atelectasis, and to quantitate areas of lung discreetly compressed by dents of cartilage that is not visualized well by CT. In addition, 3D-CT reconstructions of the bony thorax can provide useful landmarks to the surgeon when planning thoracic expansion (Fig. 4).

In follow up, after expansion, the reconstruction nicely demonstrates the results along with new bone formation from liberated periosteum (7) (Fig. 5).

Our evaluation also includes an echocardiogram to rule out pulmonary hypertension; although, perhaps surprisingly, we have yet to find any significant examples in our series.

INDICATIONS FOR OPERATION

At our current state of development, we are only recommending surgery for patients with symptomatic Jeune syndrome. Early in the series, we encountered some patients with very marginal pulmonary reserves, who spent most of the winter in the hospital. Expansion resulted in significant clinical improvement. More recently the majority of indications have been infants with Jeune syndrome who have continually required



Fig. 5. 3D reconstruction of a CT scan of the bony thorax of a patient with Jeune syndrome who has undergone bilateral expansions. Arrows point to the new bone formation from the liberated periosteum.

mechanical ventilation. The settings are frequently fairly minimal, but there has been no ability to make any progress on weaning prior to expansion. Most have had a tracheostomy and a gastrostomy tube placed for management.

OPERATIVE TECHNIQUE

Our preferred surgical approach is lateral thoracic expansion (LTE) (8,9). LTE is performed first on the side most severely affected. We have done this under the theory that, in the event of a complication, the better lung would be available to sustain the child. In reality, we have not experienced respiratory or ventilatory complications in our series. We believe the patient should be at least 6–9 months of age to allow sufficient bone size and strength to support an expansion. We position the child in a lateral thoracotomy position with the side to be expanded on top.

We expose an area of chest wall that contains a six-rib segment in the posterolateral area of the thorax. Rib is liberated from the surrounding periosteum, and osteotomies are performed: the top and bottom ribs are divided in the center of the field, whereas staggered osteotomies are performed on the middle four, making sure the anterior osteotomies are far enough behind the costochondral junction to preserve it so that the forward displacement caused by the expansion can be transmitted all the way out to the sternum (Fig. 6). The residual chest wall, including the periosteum, is then also divided in a staggered fashion, with the staggers opposite to those on the bony thorax. Bringing the longer anterior section of one rib to the longer posterior section of an adjacent rib creates the expansion, which is held in position by a titanium mini-plate and screws (Fig. 7). Early in our experience we used 1.5-mm plates, believing they would be strong enough struts to provide support until new bone formed. However,

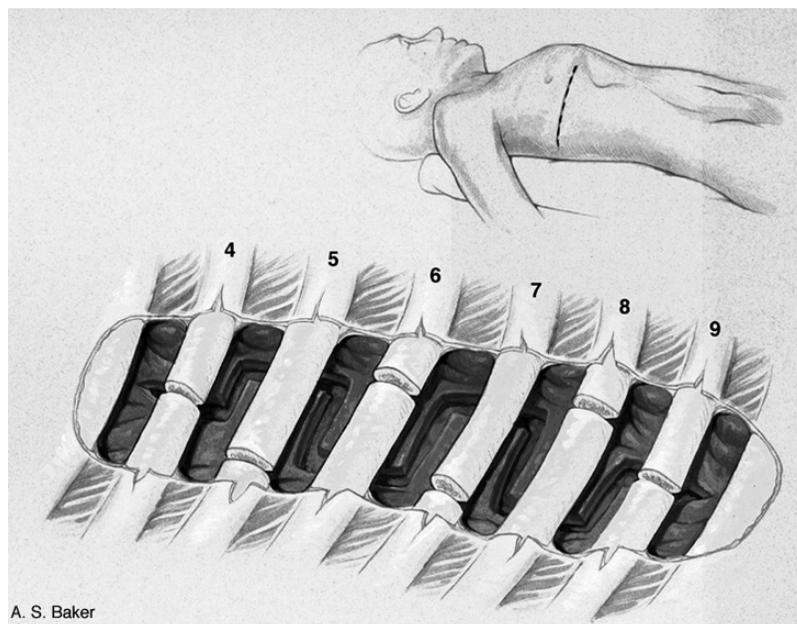


Fig. 6. Illustration of LTE with osteotomies performed and underlying chest wall divided.

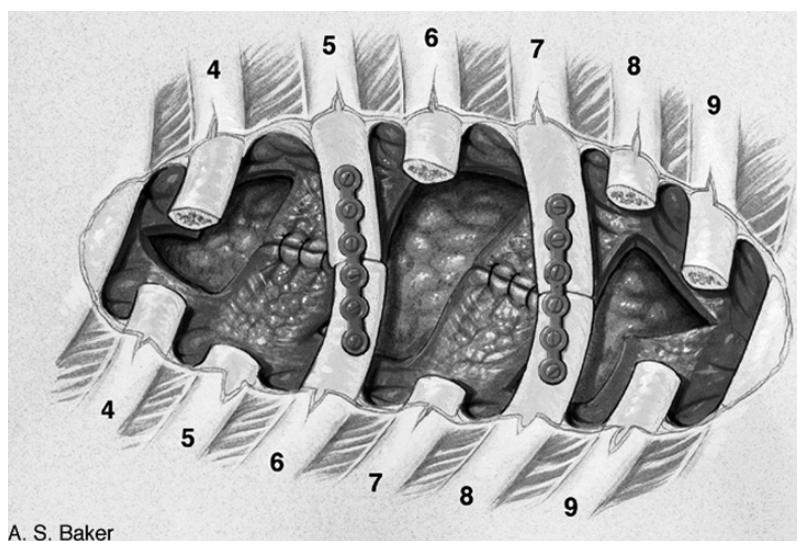


Fig. 7. Illustration of LTE with expansion completed and plated.

strut fractures occurred with a frequency too great to ignore, probably from repeated bending and torque at every breath on the relatively rigid metal. Since we began using bigger heavier plates (Mandibular Modular Fixation System, 2.4/3.0 locking reconstruction plate; Synthes Systems, Paoli, PA) 6 years ago, we have not experienced a strut fracture.

Liberated periosteum is then tacked across the empty space related to the short rib sections. This is tacked to the external surface of the expansion section, to maximize internal space. Muscles are reapproximated over the expansion and the incision closed.

POSTOPERATIVE MANAGEMENT

For the first day or two, the patient is heavily sedated and pain managed by narcotics. After day 3, pain does not seem to be much of an issue, so we lighten up on the sedation and go back toward baseline ventilation. Our strategy is to maintain full ventilation for 4–6 weeks, while the periosteum regenerates new rib. We have demonstrated (7) that the ribs will heal across the plate, despite the absence of surrounding periosteum; and also that the liberated periosteum will generate new rib by 6 weeks. During this time of new rib formation, we want to maintain it in the most expanded position to maximize ultimate internal thoracic volume.

Most of our patients come from outside our service area, and are ready to go back to where they came from in approximately 1 week, by which time they are stable and at baseline nutritional and respiratory status. We have arbitrarily waited 4–6 months for complete healing and new rib formation before bringing the patient back to expand the other side. Once an additional 4–6 weeks has elapsed to allow full healing of the second side, a very slow wean can be commenced.

RESULTS OF LATERAL THORACIC EXPANSION (LTE)

Clinical results have been encouraging (9). Updating our last report, we now have a series of 27 LTE procedures in 18 patients. There has been no early mortality. There were two late mortalities early in the series, both related to progressive tracheomalacia or bronchomalacia. We now screen for those problems, and would avoid operation in that setting. Another patient died at home from acute airway obstruction after accidental decannulation. Autopsy revealed a large, previously unrecognized tracheal granuloma as the most likely source of the mortality. A fourth late mortality was a patient who was doing very well who was found dead in bed, and we have no confirmed cause of death. Of the last two patients, both had pulmonary function studies suggesting they were doing very well, and may have ultimately gotten off ventilation over time. Virtually all other patients have had clinical improvement, mostly in the form of decreased requirement for mechanical ventilation. In some cases, ventilation is no longer required, with one patient actually having the tracheostomy removed. Others can be managed off of the ventilator for significant parts of the day, and are ventilated at night, resulting in a significant improvement in lifestyle.

Serial pulmonary function studies repeated 4–6 months following LTE show an increase in total lung capacity (Fig. 8). Taking into account the overall growth that has occurred in the meantime, we estimate that 7–15% increase in capacity is accounted for by the procedure (10), and the remainder by growth in general. However, over time, the increase in somatic growth appears to be greater than the increase in total lung capacity, so that, when expressed as “percent of expected total lung capacity,” there appears to be a slight decrease over time (Fig. 9). Although this could be an artifact of applying a normal growth curve to the Jeunes population, our impression in long-term follow-up is that this reflects a reality: Although the LTE procedure results in clinical improvement in the short run, the fundamental problem of Jeune syndrome,

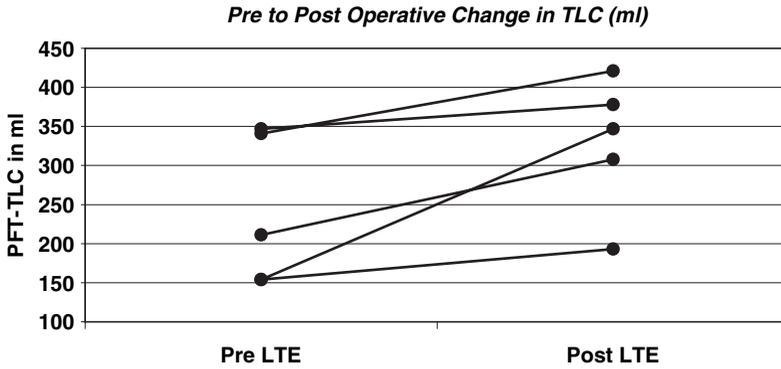


Fig. 8. Effect of LTE on TLC. TLC, total lung capacity in milliliters. PFT-TLC, total lung capacity as measured by pulmonary function tests.

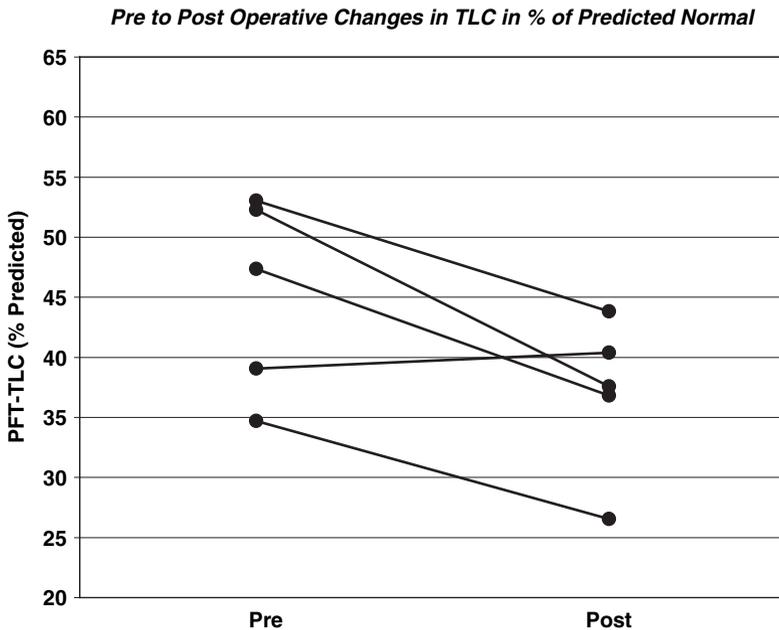


Fig. 9. Change in TLC before and after LTE expressed as percentage of predicted normal. CT-TLC, total lung capacity as measured by CT.

that growth of the thorax does not keep up with that of the body, is not altered. Only long-term follow-up will tell us if the benefits of the procedure will last, and, if they don't, if reoperation is effective or even possible.

REOPERATIVE OR LONG-TERM SURGICAL MANAGEMENT

Strut Fractures

As mentioned earlier, we noted that fracturing of the titanium struts was a long-term problem until we changed to the larger struts. Most of the time the fractures occurred

late, after the ribs had healed and new rib had formed. Therefore, the fractures did not have any effect on the integrity of the expansion. Reoperation was only done on a couple of occasions when the fracture occurred in such a way that one of the fragments was pushing into the skin from the chest wall, causing discomfort and prompting removal of the fragment. The lone exception was one case where the strut fracture occurred in the early postoperative period, and resulted in collapse of the expansion. In that case we reoperated, restoring the expansion and using the larger struts. That patient went on to do well. After going to the larger struts, and as we went for larger expansions, another problem appeared: greenstick fractures behind the strut. These on occasion have been an apparent source of pain and were treated by going to total ventilatory support for a few weeks until healing occurs. The problem has not occurred since we have gone to routine elective full ventilation for 4–6 weeks after the procedure.

Outgrowth of the Expansion

As of this writing, we have yet to reexpand a side already expanded. In a few instances where the patient's ventilatory requirements seemed to be increasing rather than decreasing, we noted they had gained considerable body weight, and total calorie reduction seemed to take care of the problem. With all of that said, the data in Figure 9, as well as our clinical observations, suggest that, given enough time, somatic growth may once again outstrip pulmonary function. Theoretically, it would be possible to reexpand an already operated site, but that will have to be individualized. Finding an area to expand outside of the original expansion may be the largest challenge. Alternatively, use of external, expandable titanium rods in the cranio-caudal axis as described by Campbell et al. (11,12), may possibly provide additional capacity.

LONG-TERM ISSUES

Creation of Synostosis

The LTE procedure in essence creates a synostosis over a significant portion of the chest wall. Although the space is enlarged, and internal volume is increased, this is at the expense of chest wall mobility. Ribs are fused and the intercostal muscles rendered immobile. As previously mentioned, the underlying pathology of Jeune syndrome are the short, flat, horizontally oriented ribs resulting in no chest wall mobility; consequently, they are totally diaphragmatic breathers. Therefore, there is no penalty for a loss of mobility that they never had. This would not be the case in acquired Jeune syndrome, where rib orientation and chest wall mobility are more normal. In that setting the increase in volume from an LTE may be offset by the loss of mobility; hence, our reluctance to offer LTE for those patients.

Tracheal Granulomata

In our recent experience, the incidence of tracheal granulomata is approximately 25%, which is higher than other populations with chronic tracheostomies. As mentioned previously, that condition was likely responsible for one of our late mortalities. As a result, we have recommended periodic screening for these lesions both above

and below the tracheostomy stoma. If found, the granulomata can be removed electively.

SUMMARY AND SPECULATION

LTE is a relatively safe and effective palliation for symptomatic patients with ATDs. Improvement in respiratory symptoms can be expected in the short and medium term. Some patients may be able to totally overcome ventilator dependence, others may lessen dependence, allowing times of separation from the machine to play or be more mobile. Significant improvement in quality of life can be expected.

However, there are potential long-term problems that require continual follow-up, preferably by an interested and involved local pediatric pulmonologist. Although clinical results look good for the first decade, it is possible that, given more time and growth, the LTE may not be a permanent solution. Our longitudinal studies confirm that the underlying pathology of Jeune syndrome has not been affected and that thoracic growth will not keep up with somatic growth, and respiratory insufficiency could decline with time. At that point, reexpansion, or the use of alternative techniques may be indicated.

We have yet to recommend the LTE as a prophylactic procedure for patients with milder forms of Jeune syndrome. These patients have the clinical stigmata of the disease, but not to the degree that they have important respiratory insufficiency. Theoretically, expansion in the first 4–6 years of life could result in an increase in the total number of alveoli in their lungs, as opposed to expansion later in life. When done after 4–6 years of age, one can only expect some recruitment of atelectatic alveoli reducing ventilation/perfusion mismatch, but not increasing the actual population of alveoli. Those extra alveoli might represent a long-term benefit to patients. However, at the present time we do not believe we have sufficient understanding of the natural course of the disease, or the long-term outcome of surgical expansion, to make that recommendation.

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8

Reoperation for Benign Breast Disorder

Denise B. Klinkner, MD
and Marjorie J. Arca, MD

CONTENTS

RECURRENT SOLID BREAST MASSES
RECURRENT BREAST CYSTIC DISEASE
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Pediatric breast disease covers a broad spectrum of pathology. Fortunately, benign masses, such as fibroadenomas, gynecomastia, and macromastia, account for the majority of diseases (1). Thus, operations on the pediatric breast occur infrequently (2). Reoperations may be required for new lesions or recurrence of previous disease. Although reoperative pediatric breast cases are few in number, each must be undertaken carefully, minimizing injury to the breast bud. Damage to the breast bud may result in irreversible cosmetic changes and the need for reconstruction.

As in adult patients, pediatric patients may develop solid or cystic disease. Recurrent masses are typically of the same nature as the original disease. The most common malignant mass in the breast of a child or adolescent is a metastatic lesion (3). Rhabdomyosarcoma (4), Hodgkin disease, and non-Hodgkin lymphoma (5) may present as primary breast lesions, and thus recurrent breast malignancies. Only 74 cases of breast carcinoma in the pediatric population have been reported from 1888 to 1977 (6).

RECURRENT SOLID BREAST MASSES

Clinical Presentation

Solid breast masses may be noted upon palpation by the patient, the patient's caregiver, by the physician, or on follow-up imaging. High-risk patients should be carefully followed long term, because outcomes improve with early detection (7). Risk factors for breast malignancies include positive family history of inherited breast disease (BRCA1 and BRCA2), history of benign disease associated with premalignant changes (i.e., fibrocystic changes with atypia), history of other malignancies, or previous irradiation to the neck and chest areas.

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The most common recurrent solid masses in the pediatric breast are fibroadenomas, which present as smooth, mobile lesions. These lesions may recur at the site of the previously excised lesion, or in another location within the same breast or the contralateral breast. Up to 15% of patients with fibroadenomas will develop multiple lesions. Rapidly growing juvenile or giant fibroadenomas warrant further intervention, as their large size may cause significant breast distortion.

Patients with resected cystosarcoma phyllodes tumor may develop recurrence in 20% of cases (8). These patients will present with a palpable mass at the surgical site or a physical change of the scar. The recurrence of a phyllodes tumor may suggest a sarcomatous degeneration of the lesion. Aggressive treatment is required, often requiring a mastectomy to provide clear margins in some patients.

Diagnostic Evaluation

Ultrasound remains the mainstay of diagnostic options in the young breast (9). Ultrasound is typically painless and allows for definition of borders and/or mass infiltration. However, ultrasound cannot distinguish between fibroadenoma and phyllodes tumor, requiring instead histologic evaluation. In preadolescent children, a biopsy should be considered with extreme caution because the developing breast bud may be irreparably harmed, even with a needle aspirate. Furthermore, these patients usually require sedation in order to safely perform interventions at the time of ultrasonography.

In the adolescent, fine needle aspiration (FNA) and core biopsies may be performed under ultrasound guidance, perhaps eliminating a need to resect (10). FNA has identified metastatic disease to the breast in the pediatric population, with rhabdomyosarcoma and acute myeloid leukemia most commonly detected (11). FNA cannot distinguish between fibroadenomas and phyllodes tumors (12). Core biopsy may provide additional information in the assessment of the breast mass (13). Mammography is reserved for those greater than 35 years of age, as the dense breast tissue in young children limits visibility (14).

Magnetic resonance imaging (MRI) with gadolinium-based enhancement has drawn recent interest as a modality to study breast disease. However, findings have been inconsistent and therefore limiting for the routine use in children and adolescents. For example, age and menstrual cycle may affect the parenchymal contrast medium enhancement on MRI (15). Because these variables cannot be controlled, reading the images consistently has been difficult. In women with a high risk of breast cancer, breast MRI does have a high sensitivity (88%) but only moderate specificity (67%) (16). In addition, MRI cannot adequately distinguish between fibroadenomas and phyllodes tumors (17). Thus, MRI does not obviate the need for tissue diagnosis. The use of MRI in the pediatric and adolescent population for breast disease has not been validated.

Operative Techniques

Excisional biopsies are usually performed using a circumareolar incision; if the mass is distant from the areola, an incision directly overlying the mass may be performed. If the location of the recurrent or second mass is amenable to use of the previous incision, the same incision may be utilized. In the event of malignancy, the original incision site should be excised with the tissue. Alternatively, confirmed benign fibroadenomas may be treated with cryotherapy under ultrasound guidance, thus avoiding multiple incisions and possible breast distortion (18).

Masses found with ultrasound examination only or masses in large breasts may be localized using ultrasound to place wire(s) preoperatively. After the skin incision is made, the wire is brought into the incision. A wide margin around the mass and the tip of the wire ensures complete excision. Use of a knife or cautery allows safe and complete resection of the wire. The *ex vivo* tissue sample is then sent for mammography to confirm the removal of the target tissue.

Because up to 25% of phyllodes tumors can be malignant, any recurrent disease mandates additional resection. The axilla must be carefully examined preoperatively, as lymph node dissection would be necessary only in clinically palpable disease. Breast conservation is recommended in the pediatric population (19). Mastectomy is appropriate in patients whose tumor fails wide local excision. Sarcomas appearing in previously irradiated fields likewise mandate aggressive surgical therapy. Patients treated with chemotherapy alone (14 patients) had a 0% 5-year survival, as opposed to 39% survival after surgical resection (28 patients) (20).

Although rare, secretory carcinoma of the male breast affects the pediatric population. Treatment has included lumpectomy to mastectomy with lymph node biopsy in the male patients. de Bree et al. reported lymph node involvement in three of 10 patients undergoing lymphadenectomy, suggesting mastectomy with lymphadenectomy as the first choice in treatment (21). In recurrent disease, this would be the operation of choice.

Patients requiring extensive resection may benefit from preoperative assessments with plastic or reconstructive surgeons (22). Immediate reconstruction has been deemed possible in older women with breast cancer (23). Postpubertal young women may be candidates, whereas adolescents may require multiple staging procedures or use of prosthetic devices until puberty has completed (24). Zion et al. reported a greater need for reoperation in patients undergoing reconstruction with implants compared to patients with mastectomy alone (25). Data comparing mastectomy and implants to tissue reconstruction have not been reported.

Long-Term Issues

Children and adolescents with benign recurrent lesions such as fibroadenomas should be taught to perform self-breast examinations. In addition, a breast examination should be performed as part of the regular yearly physical examination. If the breast is nodular, making the examination difficult, ultrasonography may be used to follow a mass serially. If a single focus of atypical ductal hyperplasia is present and is completely removed, judicious observation may be applied. Johnson has reported a single case of a 16-year-old patient who presented with multiple foci of atypical ductal hyperplasia, who was managed with prophylactic mastectomy and immediate prosthetic reconstruction (26). Adolescents who present with breast carcinoma should be screened for the BRCA genes.

Although uncommon in the pediatric population, younger women tend to have more aggressive breast cancer. Thus, systemic adjuvant chemotherapy is strongly advised in all young women with breast carcinoma. There are no concrete recommendations regarding the use of endocrine methods (i.e., ovarian ablation) in treatment of breast cancer in these women (27). Patients who test positive for *BRCA1* or *BRCA2* may opt for bilateral prophylactic mastectomies (28). This may be delayed until age 35 or childbirth has been completed (29). Young patients must be included in the decision-making process for both genetic testing and operations.

RECURRENT BREAST CYSTIC DISEASE

Clinical Presentation

Fibrocystic breast changes commonly occur in the adolescent population. Physical examination findings may reveal discrete breast cysts or diffuse small lumps throughout. Cystic changes may be simple or complicated by infection or papillomatosis (30). These fluid-filled masses may coincide with the patient's menstrual cycle. Eliciting this information from the history may obviate the need for additional operative interventions.

Diagnostic Evaluation

In the event of previous benign cyst disease, a FNA may be performed in the clinical setting. The fluid should be sent for cytopathological examination if bloody. Fibrocystic changes are classified histologically into three categories: nonproliferative changes, proliferative changes without atypia, and proliferative changes with atypia. Patients with proliferative changes and/or atypia have a higher risk for future malignancies. If the cyst fails to resolve, ultrasound examination may guide the drainage of the cyst or provide evidence of malignant disease. As with solid masses, ultrasound is the modality of choice and may guide additional evaluation (30). Cysts with irregular walls, septations or mixed cystic-solid masses, a single dominant lump that is present for several months, or recurrent cyst after FNA likely require excisional biopsy to accurately assess the pathology (31,32). Surgical evaluation combined with ultrasound and core biopsy provides a safe means of diagnosis in patients younger than 30 years of age (33).

Operative Techniques

Benign cysts may be removed at the patient's request or in the event of persistence. Cysts do not require extensive margins. Thus, simple cyst excision is possible. In fully developed females, a periareolar or incision along the lines of Langer will provide a cosmetic outcome. If the recurrent cyst is near the previous cyst, the same incision may be used.

Long-Term Issues

Although no specific data are available in adolescents, the malignancy risks in adults are well-described. Proliferative fibrocystic disease (which has been defined as moderate or florid hyperplasia, sclerosing adenosis, or papilloma with a fibrovascular core) has been associated with a 1.5- to twofold increased risk of developing breast cancer. The most substantial increase in risk of breast cancer is observed in patients with atypical or lobular hyperplasia; this is associated with a 4.4-fold increase in cancer risk, which increases to 9-fold with a positive family history (34). Recent data suggest gross cystic disease itself is not a significant risk factor for breast cancer (35).

Screening guidelines for patients with a history of atypia on breast biopsy are still evolving. In adults, current recommendations are for monthly self-breast examinations, yearly physician examinations, and yearly mammograms. No data indicate that the additive radiation from mammograms increases the risk of breast cancer. Aside from annual mammograms, these recommendations should be followed in children and adolescents.

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9

Abdominal Wall Disruption

Steven Teich, MD
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Postoperative disruption of the abdominal wall, either by dehiscence or evisceration, is a rare occurrence with an incidence of 0.4–3% (1–6). The low rate of wound disruption is believed to be the result of advances in postoperative care, use of broad-spectrum antibiotics, and a better understanding of the deleterious effects of systemic illness and malnutrition on wound healing (1–6). The mortality rate in adults who suffer from wound disruption approaches 15%, usually owing to their underlying associated condition (7,8).

In children, four clinical studies from 1953 to the present have addressed the problem of wound dehiscence and evisceration, all of which are retrospective in nature. Gross and Ferguson demonstrated a decline in the evisceration and mortality rates at the Boston Children's Hospital from 1931 to 1950. By the last 5 years of their study, the evisceration rate was 0.39% and the mortality rate was 27% (9). In 1972, Campbell and Swenson examined the dehiscence rate of paramedian, midline, and transverse incisions (10). Their results suggested that transverse or grid-iron muscle splitting incisions were preferable to longitudinal (midline or paramedian) incisions (dehiscence

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rate 1.6% versus 0.1%). These results were confirmed by Waldhausen and Davies, who found that transverse incisions had a dehiscence rate of 0.25% versus a dehiscence rate of 1.75% in vertical incisions (11). Furthermore, vertical incisions were much more likely to dehiscence in patients under 1 year of age, especially when other risk factors for disruption were present. These risk factors include preexisting disease, infection, age (very young and very old), immunosuppression, method of closure, nutritional status, and presence of a drain or stoma. They recommended that a transverse incision be the incision of choice for infants less than 1 year of age unless there are no other risk factors present or there is a clear exposure advantage for another type of incision. Ventral incisions should be used primarily in older children. Among the adult population, there is currently little evidence that the orientation of the incision plays a significant role in abdominal wound dehiscence. The nature of the operation itself with its risk of wound infection, the technique of wound closure, and postoperative factors appear to be more critical to proper wound healing.

The technique of wound closure for children is also controversial. Kiely and Spitz found no difference in wound infection rate, dehiscence, and incisional hernia between layered and mass closure of abdominal wounds in infants and children (12). Both transverse and vertical incisions were included in their study and with both types of closure.

Many predisposing factors that put an adult at risk of wound dehiscence do not play a role in children, such as advanced age and poor vascular supply. However, other factors such as preexisting pulmonary disease, malnutrition, and immunosuppression are probably relevant in pediatric patients. Preoperative pulmonary disease and subsequent postoperative respiratory complications may play a role in the development of wound failure in infants and children, but their role in healing of operative incisions has not been investigated by clinical studies. In adults, pulmonary complications are the most common event preceding postoperative wound dehiscence (13–15).

The importance of adequate nutrition in wound healing was first reported more than 65 years ago with several reports documenting the association between hypoproteinemia and wound disruption (1,16,17). Protein deficiency is rare in the United States today but its prevalence among surgical patients who have serious underlying diseases remains an ongoing problem. One study in adult patients documented hypoproteinemia in 85% of patients who had wound dehiscence (18,19). Therefore, protein deficiency should be corrected prior to reoperative surgery. When enteral nutrition is not possible, total parental nutrition should be instituted until malnutrition and hypoproteinemia have been corrected prior to elective reoperative surgery.

INTRAOPERATIVE PREVENTION

Perhaps the most important measure for prevention of wound disruption following reoperative laparotomy is avoidance of bowel injury and subsequent contamination of the peritoneal cavity. Several studies have shown that wound infection is present in 70–80% of patients who experience wound dehiscence (20–23). Infection interferes with the normal healing processes. Infected wounds contain less collagen and the collagen is not as highly cross-linked as in a healthy wound. The detrimental effect of wound infection on tissue strength is extremely important, because this weakness sets the stage for postoperative wound herniation and dehiscence.

The use of intraabdominal drains following reoperative laparotomy remains controversial. Valid indications for drain placement include: (1) anticipated leakage from an

adjacent organ such as the gallbladder bed; (2) an isolated abscess cavity with an attempt to collapse it; and (3) a questionable intestinal anastomosis caused by tension or compromised tissue at the suture line (24). Closed suction drainage is preferable to a Penrose drain because the open drainage system can serve as an entry route for pathogens.

The route by which drains are brought through the abdominal wall is important to the risk of subsequent wound infection. After draining an intra-abdominal abscess, the drain should not be brought through the incision, because this technique increases the likelihood of a wound infection (7,12,25). The drain should be brought through a separate, more dependent opening in the abdominal wall, as far away from the operative incision as possible. A stoma should not be brought through the incision, because this location also increases the risk of wound infection and disruption (12).

FASCIAL CLOSURE

Whereas different suture materials for closure of the abdominal fascia have been studied extensively in adults, there are no parallel studies in children. The suture materials that have been developed over the last 20–30 years strive to achieve the principles of the “ideal” suture: (1) maintain adequate strength until the tissue is completely healed; (2) disappear after healing to minimize patient discomfort and suture granulomas; (3) have a low risk of promoting wound infection or inflammation; and (4) handle easily and tie securely with minimal difficulty.

Permanent monofilament sutures such as polypropylene and nylon have decreased the incidence of wound dehiscence and incisional hernia dramatically (26–29). Although these sutures have achieved the same extremely low dehiscence rate as stainless steel, both have a propensity for incisional discomfort and suture granuloma formation (30,31). These complications are particularly common in thin patients with minimal subcutaneous fat, including children and lean adolescents. It is unclear if “long-term” absorbable sutures such as polydioxanone (PDS) have a lower incidence of wound disruption or simply alter the time curve of wound failure so that an incisional hernia is more likely to occur rather than a wound dehiscence (5,31). Furthermore, sinus formation may result from absorbable as well as nonabsorbable sutures. We do not recommend absorbable sutures for wounds at high risk for dehiscence. Monofilament polypropylene or nylon may lead to occasional chronic suture sinuses, but this problem can be minimized by using a continuous closure and burying the knots at both ends of the incision.

TECHNIQUE OF CLOSURE

The technique of abdominal wound closure is the most important factor in the prevention of postoperative wound dehiscence. Several important principles have been elucidated over the past 30 years. Numerous clinical trials have confirmed that it is unnecessary to close the peritoneum (19,32,33). Sutures that enter the peritoneal cavity cause a foreign body reaction and increase the potential for adhesions and possible bowel obstruction. Fascial closure should be performed via a preperitoneal technique without penetrating the peritoneum.

Mass closure of the fascia is superior to a layered closure because it imparts greater wound strength. Several studies have demonstrated a decrease in wound dehiscence with mass closure (26,34–38). In addition to providing a less secure wound, layered

closure is more time-consuming and significantly increases the amount of foreign material within the wound.

Animal studies have confirmed that a continuous fascial closure in comparison to interrupted closure better distributes wound tension along the length of the incision to accommodate increased stress at any point (26,39). Fascial tearing caused by wound tension is considered to be the primary cause of wound dehiscence in most patients.

RETENTION SUTURES

Retention sutures are placed to hold the fascial edges together and reduce shearing on the suture line until adequate healing has taken place. Patients who have one or more risk factors for wound dehiscence may be excellent candidates for retention sutures. "Conventional" retention sutures may fail because they only indirectly appose the fascial edges and allow deep separation of the fascial edges while the skin remains intact (40). Therefore, several important technical points must be followed in placing retention sutures: (1) the sutures should be placed 4–5 cm lateral to the incision, traverse all layers of the abdominal wall except the perineum, and cross the midline beneath the mass fascial closure just under the posterior sheath superficial to the peritoneum; (2) each suture must enter the skin closer to the incision than to the point where it traverses the ipsilateral posterior fascia to allow approximation of the fascia without compression and potential necrosis of the overlying skin; and (3) if the fascia is closed with the abdomen distended, the retention sutures must be checked periodically in the postoperative period to ensure that tension is maintained (Fig. 1). The retention sutures can be removed 14–21 days after the operation.

PRESENTATION AND MANAGEMENT OF DEHISCENCE

Because the mortality rate after wound dehiscence is approximately 15%, early recognition and treatment are critical (7,8,33,34,41). Wound dehiscence that occurs before the fifth postoperative day is usually caused by an error in technique. The typical wound dehiscence occurs between the seventh and eighth postoperative days. It usually follows progressive abdominal distention secondary to ileus or after severe vomiting or coughing. An alternate presentation is the drainage of serosanguinous fluid from the wound. When the skin sutures are removed viscera is visible in the wound.

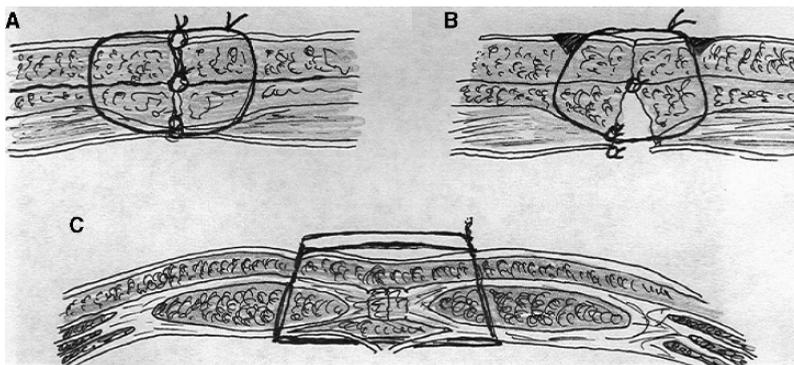


Fig. 1. Placement of retention sutures. (A) Improperly placed retention sutures; (B) Resulting fascial separation; (C) Properly placed retention sutures.

The initial management of dehiscence includes prompt coverage of the exposed intestine with warm saline dressings. The dressings should be kept moist to avoid desiccation of the bowel and minimize the loss of fluids and heat. In infants the use of a sterile bowel bag is preferable, because wet dressings may lead to hypothermia. The stomach should be decompressed with a nasogastric tube. Intravenous fluids should be given at a rate to compensate for maintenance fluid requirements and fluid losses owing to drainage from the open abdomen, and broad-spectrum antibiotics should be instituted.

Once the patient is anesthetized, the entire wound should be reopened and the fascial closure inspected to determine the cause of the dehiscence. Any devitalized tissue should be debrided. A formal exploratory laparotomy should then be performed to assess for an intraabdominal abscess or other abnormalities. We recommend irrigation of the peritoneal cavity with warm saline solution. Fascial closure should be performed using a heavy nonabsorbable suture in a continuous fashion. Retention sutures should also be utilized. Postoperatively, the patient should remain on nasogastric decompression and total parental nutrition should be instituted. Vigorous postoperative pulmonary toilet must be achieved in conjunction with adequate pain control.

THE OPEN ABDOMEN

Successful management of the open abdomen remains a formidable challenge for pediatric surgeons. The mortality rate after laparotomy in adult patients with severe intraabdominal injuries and a subsequent open abdomen is 26% (42). Treatment of the open abdomen is costly and the hospitalization is lengthy. Short-term complications include evisceration, peritonitis, third-space fluid losses, reperfusion syndrome, and prolonged ileus. Long-term sequelae are substantial and include enterocutaneous fistula, incisional hernia, intraabdominal abscess, unstable wounds prone to breakdown, and cosmetic issues. Banwell and Teot have proposed a classification system for abdominal wounds that stratifies them according to increasingly difficult clinical management requirements (43). Table 1 outlines this classification paradigm.

MANAGEMENT OF THE OPEN ABDOMEN

The open abdominal wound may involve the skin only or there may be a concomitant muscle and fascial defect. In wounds left open for more than 72 hours, there may be secondary retraction of the fascia, making delayed primary closure more difficult. Temporary abdominal wound closure is desirable when tension-free primary closure of the abdominal wall is impractical or when planned reexploration is necessary. The principles of management of an open abdominal wound include: contain the intraabdominal viscera, protect the viscera from mechanical injury, prevent bowel

Table 1
Classification of Open Abdominal Wounds

<i>Classification</i>	<i>Wound Type</i>	
I	Superficial Skin defect only	Prosthetic material status
II	Deep Exposed bowel and omentum	a. Absent
III	Complex Presence of intraabdominal sepsis	b. Present
IV	Complex Presence of enterocutaneous fistula	

desiccation, actively remove wound and visceral exudate, minimize abdominal wall tissue damage, prevent peritoneal contamination, minimize complications, and expedite hospital discharge.

The secondary goal of open wound management is to facilitate closure of the abdominal wall. This can be achieved by delayed primary closure or by promoting granulation tissue with subsequent skin graft. Closure of the abdominal wall may involve skin only or skin and fascia. If the fascia cannot be closed primarily, a number of strategies have been described for adult patients that can be modified to achieve temporary closure in a difficult pediatric abdominal wound. These include the Bogota bag (a modified plastic irrigation bag (44,45)), the Wittmann patch (a Velcro-like device to sequentially reduce the abdominal wall defect (46)), and prosthetic mesh (absorbable [e.g., polyglactin] or nonabsorbable [e.g., Gore-Tex, polypropylene, and silastic] (47)). Pediatric surgeons experienced in the use of the pre-made silastic spring-loaded silo for nonreducible gastroschisis can apply this device in the open abdominal wound of a neonate or small child. Nonabsorbable mesh increases the rate of enterocutaneous fistula formation, whereas absorbable mesh is prone to late incisional hernia. A zipper-type closure has been described to allow planned multiple reoperations (47).

VACUUM-ASSISTED CLOSURE (VAC) OF OPEN WOUNDS

In 1933 Fleischmann et al. first described the concept of controlled subatmospheric pressure in the treatment of open and infected wounds (48). The VAC system consists of a polyurethane ether sponge with pore sizes of 400–600 μm , which is cut to fit over the open wound. An adhesive dressing is placed over the sponge with a drainage tube connected to an adjustable vacuum pump (49). Subatmospheric pressure of -125 mmHg, or less for neonates and small infants, is then applied to the wound (Fig. 2). Multiple studies have demonstrated an increase in granulation tissue formation and decreased time for healing in VAC-treated wounds (50–52). The negative pressure generated by the VAC system has been postulated to accelerate wound healing by reducing edema, improving local blood flow, and removing extracellular fluid to create an optimal environment for wound healing.

Teich et al. reported a series of 51 pediatric patients ranging in age from neonates to teenagers with complex wounds that were successfully treated with the VAC negative pressure system (53). The wounds included pilonidal abscesses, sacral and extremity ulcers, traumatic soft tissue wounds, and extensive tissue loss from the abdominal wall, perineum, and axilla. Multiple other series and case reports document the efficacy of the VAC system in pediatric patients for treating a wide variety of complex wounds (54). Markley et al. reported a series of six pediatric patients who had the vacuum packing (Vac-Pac) technique for abdominal wound closure following damage-control laparotomy (55). Five of the six patients survived and there were no abdominal complications related to the Vac-Pac treatment.

TECHNIQUES TO AID ABDOMINAL REOPERATION

Prior to performing reoperative abdominal surgery several essential decisions must be made by the pediatric surgeon. A preoperative bowel preparation should be employed when there is no bowel obstruction and a lengthy, complex intra-abdominal reoperation is planned. This decreases the contamination risk from planned or unplanned enterotomies. Preoperative bowel decompression with a nasogastric tube can aid with entering

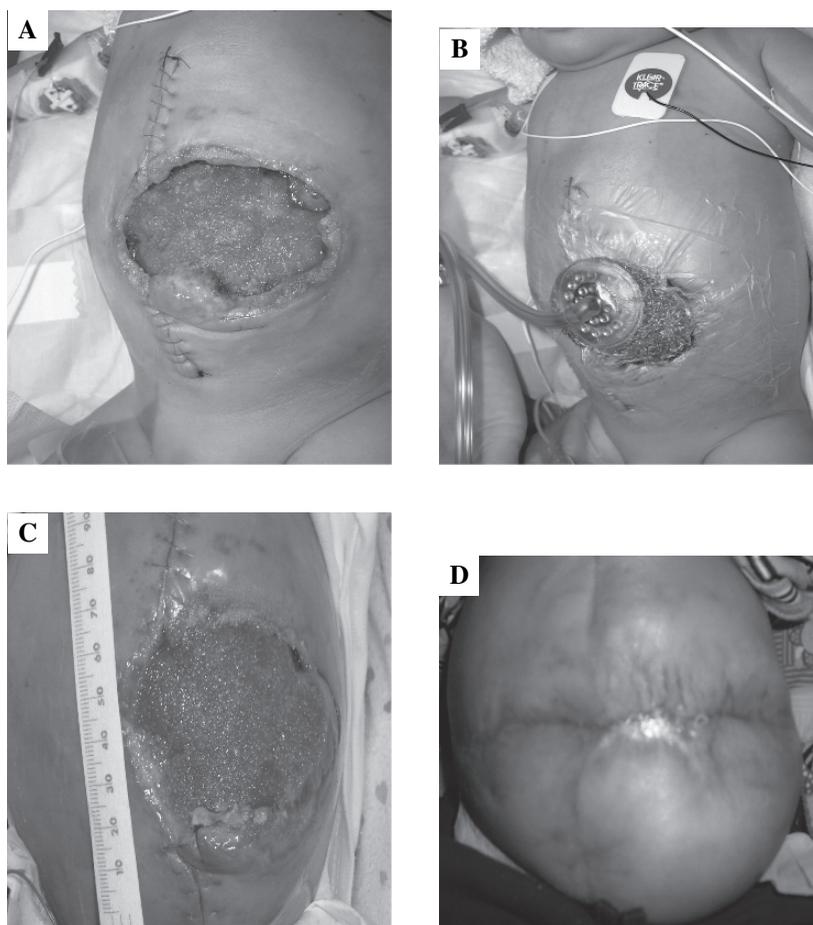


Fig. 2. Wound vac on open abdominal wound.

an abdomen with adhesions and also provide better operative exposure. Consideration should be given to preoperative marking of the abdominal wall for potential stoma(s), optimal patient positioning on the operating table, and the best surgical incision to achieve wide exposure.

INTRAOPERATIVE AIDS

The intraoperative assistance of other specialty services, such as gastroenterology, urology, and radiology, may be helpful in selected patients with complex, multi-organ problems. Intraoperative endoscopy can be extremely valuable when searching for a small gastrointestinal lesion. The surgeon can aid the endoscopist in guiding an endoscope through the duodenum into the jejunum with subsequent visualization of the entire small bowel lumen. This technique is invaluable when searching for an occult site of gastrointestinal bleeding. Intraoperative colonoscopy is also helpful for evaluating lower gastrointestinal hemorrhage and for identifying the intrapelvic end of a short Hartmann's pouch that is obscured by dense adhesions. Intraoperative ultrasonography can be used during reoperative open and laparoscopic abdominal

procedures. It can identify lesions within the liver that cannot be palpated or visualized, and assist in characterizing unanticipated findings such as a mass within the pancreas (56). Dye substances, such as methylene blue or dilute povidone-iodine, may be helpful in testing the integrity of an anastomosis (57). Tumors, inflammatory lesions, and dense inflammation within the pelvis may obscure and involve the ureters. In these situations, preoperative cystoscopic insertion of lighted ureteral catheters may be helpful in delineating the course of the ureters (58).

TECHNIQUES FOR SECONDARY CLOSURE OF THE DISRUPTED ABDOMINAL WALL

Management of surgically created abdominal wall defects has changed significantly over the past 25 to 30 years. Tension-free repair is now considered to be the treatment of choice; therefore, primary closure should only be utilized to repair small ventral hernias (59,60). Synthetic mesh, readily available in most operating rooms permits closure of defects as great as $25 \times 33 \text{ cm}^2$ in size. The mesh can be implanted by either open or laparoscopic techniques. However, its use is fraught with many potential complications including rejection, infection, wound seromas, enterocutaneous fistulas, and adhesions. Large sheets of mesh may lead to decreased abdominal wall compliance and increased abdominal wall rigidity.

Component separation (CS) is a technique of abdominal wall closure that has gained popularity since it was first reported in 1990 (61) (Fig. 3). Several clinical studies have demonstrated the utility of CS in the repair of ventral wall defects with acceptable results after abdominal compartment syndrome, in infected wounds, and in intractable defects (62–64). CS is effective because the technique includes mobilization of vascularized myofascial flaps that are brought together in the midline. Native vascularized tissue is extremely important in an infected environment and has a greater potential for healing than prosthetic material. In an intractable hernia, CS leads to

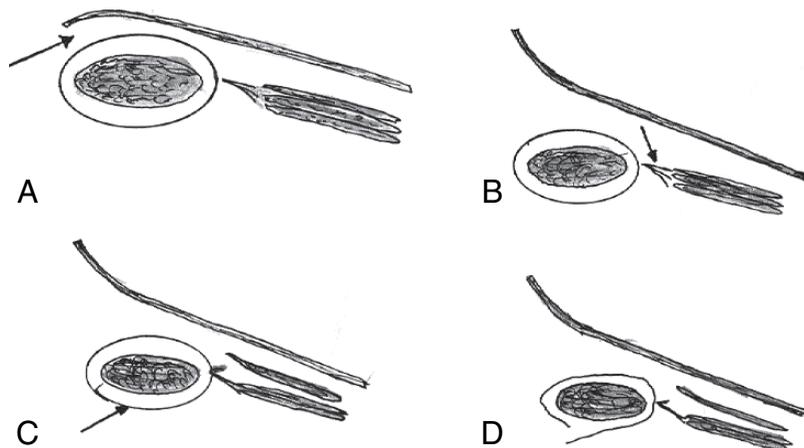


Fig. 3. Abdominal wall component separation. (A) Elevation of skin flap off anterior abdominal wall. (B) Incision of external oblique fascia at junction with rectus abdominus. External oblique dissected off internal oblique laterally. (C) Incision of posterior rectus sheath, which is dissected off rectus muscle. (D) Completed flap.

the reapproximation of native tissue under minimal tension, which allows for more secure healing. If a mesh is required, the use of the CS technique allows for a smaller size mesh to be placed and increases the likelihood of healing and a better long-term functional result.

ABDOMINAL COMPARTMENT SYNDROME

Abdominal compartment syndrome (ACS) is defined as multi-organ system dysfunction involving the renal, pulmonary, and cardiovascular systems secondary to an acute, sustained elevation of intra-abdominal pressure (IAP). ACS was first described by Baggot in 1951, but only in the last 25 years has it become a widely recognized life-threatening problem that is potentially treatable by abdominal decompression (65). Pediatric surgeons have long been aware of the deleterious effects of increased IAP because of our experience with the treatment of congenital abdominal wall defects. Pediatric surgeons were the first general surgeons to utilize prosthetic materials for the temporary closure of abdominal wall defects (66). Furthermore, pediatric surgeons utilized measurements of IAP as a guide to safe silo reduction and closure of the abdominal wall (67–69). Recent clinical reports have documented the occurrence of ACS in children beyond the neonatal period (70–72).

The most common cause of ACS is severe blunt abdominal trauma with secondary hemorrhage and coagulopathy (73–75). It is more likely to develop with combined abdominal and pelvic trauma (74). Other causes of ACS in children include tight packing used for tamponade of uncontrolled abdominal hemorrhage, after fluid resuscitation for severe burns, extraperitoneal injuries leading to the use of high-end expiratory pressures with high intrathoracic pressures, and a high Injury Severity Score (73,76–79). ACS should be suspected in a critically ill patient who develops a tense abdomen over a short period of time. Typically the child exhibits elevated intra-abdominal and peak airway pressures, decreased cardiac output, reduced renal function, and hypoxia and hypercarbia with progressively increasing airway pressures. IAP must be measured to confirm the diagnosis. This is most commonly performed by measuring urinary bladder pressure, an indirect measure of IAP. Bladder pressure is determined by instilling 1 mL/kg body weight of saline into the Foley catheter and connecting the end of the catheter to a pressure transducer or manometer via a three-way stopcock (80). The symphysis pubis is the zero reference point. A urinary bladder pressure greater than 25 mmHg requires immediate abdominal decompression, although children may develop ACS with a pressure as low as 15 mmHg (73,81–84). The decision to decompress the abdomen in a child is not determined solely by a specific pressure measurement but by concomitant physiological parameters, such as progressive renal failure or cardiopulmonary insufficiency (73). Abdominal computed tomography (CT) may suggest increased IAP in patients at risk for ACS by narrowing of the inferior vena cava (IVC), rounded appearance of the abdomen on axial CT, direct compression of the kidney parenchyma, bowel wall enhancement and thickening, and bowel dilatation (85–87).

The only treatment for ACS is immediate decompression with the use of prosthetic mesh or the placement of a wound vac for negative pressure wound therapy (NPWT). In high-risk surgical patients, a prophylactic wound vac should be placed over the open abdomen at the end of the surgical procedure (Fig. 4). NPWT applies constant and uniform negative pressure to the abdominal wall. It allows the fluid losses to be quantified and replaced more precisely. Closure rates of 71–92% have been reported

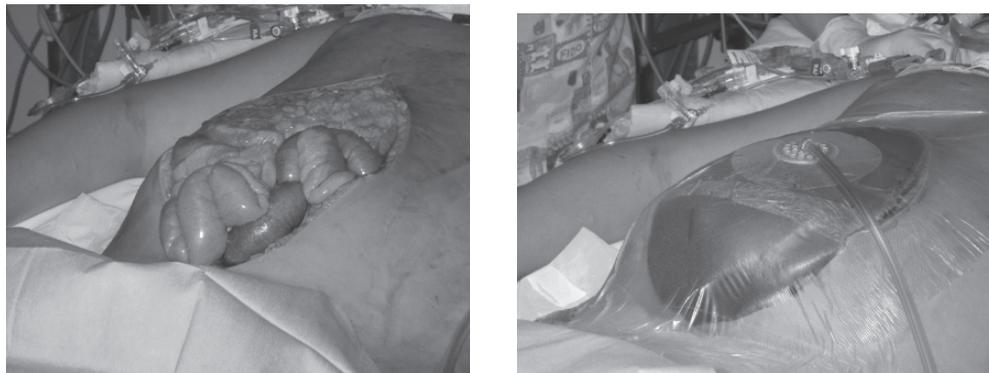


Fig. 4. Wound vac on open abdomen.

in multiple series of patients with open abdomens treated with NPWT (88–90). The mean closure time varied from 10 to 21 days, depending on the etiology of the open abdomen.

TREATMENT OF INCISIONAL HERNIAS

Wound dehiscence is the most severe and rare sequelae of failure of healing of the abdominal wall after laparotomy. Incisional hernia, however, remains a relatively common late complication of faulty healing of the abdominal wall. In multiple series of adult patients, the incisional hernia rate varies from 1–14% of all laparotomies (91–93). Recurrence rates after incisional hernia repair vary from 10–50% (94–98). The incidence of incisional hernias and the recurrence rate after incisional hernia repair in the pediatric population is unknown, because no clinical series has ever been published!

An incisional hernia usually presents as an asymptomatic bulge over the incision or immediately adjacent to it. More than one-half of all incisional hernias in adults are noticeable within the first two years after the operation, but a high percentage can occur years later (99–101). Over time incisional hernias enlarge and may become painful. Several causative factors are thought to increase the risk for development of an incisional hernia both in adults and children. These factors include a prolonged initial operation, long or multiple incisions, reoperation, obesity, postoperative hematomas, increased dead space, wound drainage, layered closure, absorbable sutures, inadequate fascial fixation of sutures, and excessive wound tension (102). Systemic considerations in children include sepsis, malnutrition, malignancy, chemotherapy, liver or kidney disease, paralytic ileus, and BPD (bronchopulmonary dysplasia). In one series of adult patients, one-half of all incisional hernias were associated with emergency procedures, infected wounds, or postoperative evisceration (103).

Except in the case of a traumatic incisional hernia or an obvious technical error (e.g., a broken suture or an unclosed trochar site) most incisional hernias result from defects in wound healing (104). This basic principle helps to explain the following observations about incisional hernias: (1) a large percentage of incisional hernias present a considerable time following the previous operation; (2) the cumulative incidence follows a linear time course; (3) repeating a previously unsuccessful closure technique has a high risk of failure; (4) the entire incision should be reinforced regardless of the

intraoperative findings; (5) patients with a proven defect in collagen metabolism have a higher incidence of incisional hernias; (6) the use of absorbable materials increases the failure rate; and (7) a successful mesh repair requires significant mesh overlap of the hernia defect.

Reinforcement of the repaired incisional hernia by mesh allows for the ingrowth of fibrous tissue into the prosthetic mesh with a resulting scar-mesh compound (104). The amount of mesh material influences the intensity of the scar formation but not the quality of the scar. Therefore, mesh fixation by fibrosis will only prevent recurrence by having a wide overlap underneath healthy tissue. In reported clinical series, there is a trend toward using larger mesh prostheses (104,105).

The only nonoperative alternative for treatment of an incisional hernia is the use of an abdominal binder, which may be applicable in selected patients who are not immediate candidates for operative repair. We advocate early repair, because most incisional hernias tend to increase in size over time and cause parental and patient anxiety. Mesh should be used routinely for any defect over 3 cm in older children and may be required for smaller defects in infants and young children (100,101).

Table 2
Techniques for Incisional Hernia Repair

<i>Type of Incisional Hernia Repair</i>	<i>Technique</i>	<i>Recurrence Rate</i>
Primary repair only	● Fascial defects <5 cm in diameter	Up to 50% (97,106–109)
Primary repair with relaxing incision/component separation	See Fig. 1	10% (102,104) (10% significant skin and wound problems)
Primary repair with inlay mesh	● Hernia sac excised ● Interrupted or continuous permanent sutures to fascial edges	10–20% (109)
Primary repair with retro-rectus mesh	● Hernia sac left intact as buffer ● Mesh fixated to muscle layer above	<10% (110,111) (Mesh infection/removal rate 5–12%)
Primary repair with intraperitoneal underlay mesh	● Allows for largest underlay of mesh ● Can be performed open or laparoscopically ● Fixation to fascial edges or to lateral posterior abdominal wall	<5% (112)
Primary repair with other mesh techniques	● Onlay mesh only ● Sandwich technique with mesh placed both onlay and either retro-rectus or intraperitoneal ● Cuff technique with placement of mesh around muscle on each side of defect with closure of mesh-reinforced edges	Unknown

In most cases, incisional hernia repair is an elective operation. Every attempt should be made to minimize the influence of risk factors that lead to recurrence. The skin should be in good condition with no areas of superficial excoriation. A preoperative bowel preparation should be performed for the repair of large abdominal wall defects to minimize postoperative tension on the abdominal wall closure, and prophylactic antibiotics should be utilized whenever use of a mesh material is contemplated. Significant oozing may result if there is a need for extensive dissection, warranting preoperative investigation for an unsuspected bleeding abnormality.

Many different techniques for incisional hernia repair have been published in the literature (*see* Table 2). They vary from primary closure only to primary closure with mesh placement in various positions. Some techniques have a documented series of patients whereas others are only reported in the literature without evidence as to their effectiveness. With the use of mesh materials, the recurrence rate for incisional hernia repair in adults has decreased to 5–10% or less in various series. Current investigations include the best approach for incisional hernia repair (open versus laparoscopic) and the best means of fixation (partial versus full-thickness abdominal wall/fascia) to stabilize the mesh during tissue ingrowth.

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10

Reoperation after Esophageal Atresia Repair and Other Esophageal Conditions

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Reoperative surgery of the esophagus in children is challenging because of the unique position and surgical anatomy of the esophagus, the general complications that can occur in esophageal surgery, and finally the specific clinical problems that require surgical attention in children.

ANATOMY

The esophagus is a muscular tube that allows food to pass from the pharynx to the stomach. The esophagus begins in a median position, which shifts slightly to the right of the spine in the midthoracic portion. The lower esophagus crosses over the vertebral column so that the esophagogastric junction is on the left. The upper esophagus is in direct contact with the membranous part of the trachea. The azygos vein crosses anterior to the midthoracic esophagus to enter the superior vena cava. The esophagus is divided into three anatomic portions. The cervical esophagus begins at the level of the cricoid cartilage and includes the upper esophageal sphincter. The longest segment of the esophagus is the thoracic esophagus, which extends from the sternal notch to the diaphragmatic hiatus. The third portion is the abdominal esophagus, which is the short segment below the hiatus extending to the gastroesophageal junction. The central portion of the esophagus is contained within loose areolar tissue in the posterior mediastinum, which allows for esophageal motility to accommodate effortless swallowing. The fascial compartment surrounding the esophagus extends longitudinally into the pretracheal space and deep cervical fascia superiorly and into the retroperitoneum inferiorly. Surgical procedures of the esophagus must respect its

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vascular supply: the cervical esophagus receives its blood supply from the thyrocervical arteries; the mid esophagus is supplied directly from the aorta at segmental levels; and the lower esophagus is supplied from the cardiac branches of the left gastric artery (1).

GENERAL COMPLICATIONS IN ESOPHAGEAL SURGERY

Reoperative esophageal surgery is required for complications of surgery and for symptoms that develop following uncomplicated surgery. In common with surgeries involving other areas of the gastrointestinal (GI) tract, operations on the esophagus can be complicated by stricture, leakage, or dilation (focal or diffuse). Strictures result from circumferential healing fibrosis and are exacerbated by acid reflux or poor blood supply. The result of a stricture is the inability to swallow, the impaction of food, and potentially aspiration. Esophageal leakage can occur following an anastomosis and is most hazardous in the thoracic portion of the esophagus. The thoracic portion is also the most frequent area of leakage, perhaps owing to the lack of serosa. Leakage can also occur following instrumentation of the esophagus either by esophagoscopy or with esophageal dilatation. Leakage from the esophagus can lead to severe mediastinitis with extension of the infection along tissue planes into the neck or retroperitoneum.

SPECIFIC CONDITIONS REQUIRING SURGERY OF THE ESOPHAGUS IN CHILDREN

Tracheoesophageal Fistula and Esophageal Atresia

Problems in the formation of the esophagus leading to esophageal atresia and various forms of tracheoesophageal fistula mandate surgical repair, usually within the first few days of life. The treatment of these problems is challenging because of the age and size of the patient and because of the relatively high frequency of associated congenital anomalies. Upon repair of the tracheoesophageal fistula, patients can require reoperation for recurrence of the fistula, anastomotic leakage, anastomotic stricture, or severe gastroesophageal reflux. The contributions of individual factors to the incidence of these problems is poorly defined but includes: surgeon experience; patient-related factors such as age, birth weight, and type of defect; and technical factors such as choice of suture material, surgical approach (open versus thoracoscopic), and type of anastomosis (single-layer versus double-layer).

Both the incidence of these complications and the frequency of reoperations are difficult to estimate. A review of several large series of patients treated for tracheoesophageal fistula, with over 1170 patients combined, shows a marked variability in the frequency of postoperative problems (2–8). Recurrence of the tracheoesophageal fistula was the least common complication, occurring in 1–10% of patients. Although one study showed “no discernible anastomotic leaks” (2), most other series included a leakage rate of 8–19%. The incidence of stricture formation varied most widely of the complications, perhaps owing to the intensity of surveillance for this problem, ranging from 6–52%. The percentage of patients in these studies that had gastroesophageal reflux severe enough to require fundoplication also varied widely, ranging from 7–44%. The wide range likely reflects controversies about surgical versus medical treatment of reflux in this situation.

RECURRENT TRACHEOESOPHAGEAL FISTULA

Recurrence of the fistula may be caused by inflammation or contamination at the surgical site or perhaps by poor wound healing. Most patients present with chronic cough or with recurrent episodes of pneumonia. Although three-dimensional computed tomography (CT) has been described to demonstrate recurrence (9), the diagnostic evaluation usually requires a prone esophagogram with direct instillation of radiographic contrast in the esophagus through a nasogastric tube (10). Endoscopy can be helpful for diagnosis, but is less sensitive and more invasive than contrast radiography. The value of endoscopy is found in the intraoperative localization of fistulae to direct the treatment of this complication. Patients with recurrent tracheoesophageal fistulae must be differentiated from those who have both proximal and distal fistulas in whom the proximal fistula was missed at the initial surgery.

Reoperation for a recurrent fistula usually includes interposition of adjacent soft tissue such as pleura, intercostal muscle, or pericardium between the divided ends of the fistula (11). The repeat thorotomy is technically challenging for several reasons: fibrosis and inflammatory tissue obscure tissue planes; the esophagus or membranous trachea can be injured, as can the vagus or recurrent laryngeal nerves; and recurrence continues to be a risk. Because of these difficulties, other operative approaches have been described, including median sternotomy with interposition of a sternocleidomastoid muscle flap (12) or transtracheal fistula closure via an anterior cervical approach (13). Another approach to avoid many of these technical difficulties is with an endoscopic closure of the recurrent fistula. A variety of agents have been reported including: fibrin glue; methyl methacrylate; n-butyl-z-cyanoacrylate and aethoxysclerol; Nd:YAG laser; or insulated wire electrocautery (14–22). No ideal agent has been found because all have a significant risk of recurrence (23). Recurrent tracheoesophageal fistula is attended by significant morbidity, and extreme cases of multiple recurrences may require esophageal replacement (24).

ANASTOMOTIC LEAKS AFTER TRACHEOESOPHAGEAL FISTULA REPAIR

The causes of anastomotic leakage after tracheoesophageal fistula repair are not fully known but have been associated with the use of braided silk sutures compared to monofilament sutures (absorbable or nonabsorbable) (25). Anastomotic leakage usually becomes obvious in the first few days after surgery or after commencement of feedings. A drainage tube is routinely left near the anastomosis at the initial surgery to help detect and control any leakage. An esophageal swallow is performed 7–10 days following surgery and, if leakage is not detected, feedings are commenced. If no leakage is noted after 24 hours of feeding, the drainage tube is removed. When present, anastomotic leakage is most often treated nonoperatively with tube thoracostomy. Patients are given nothing by mouth, maintained on parenteral nutrition, and given intravenous antibiotics; most patients will respond to these measures (26,27).

Although the majority of postoperative leaks are minor, approximately one-fourth involve a major anastomotic disruption. Some major disruptions will be found with the routine postoperative contrast swallow but most will be evident earlier, manifested clinically by a persistent pleural fluid collection or pneumothorax (25). Patients with a major dehiscence or those with progressive sepsis will usually require reoperation. Options at reoperation include: primary repair of small areas of dehiscence if the tissues appear healthy; repair with pleural or intercostal muscle reinforcement; improved drainage of the leak; or, in the more severe situations, cervical esophagostomy and

closure of the distal esophagus with plans for subsequent esophageal replacement (25,26). The long-term prognosis for patients with a leak after tracheoesophageal fistula repair includes a higher risk of stricture formation (25).

ANASTOMOTIC STRICTURE AFTER TRACHEOESOPHAGEAL FISTULA REPAIR

Perhaps the most common reason for reoperation following repair of esophageal atresia is anastomotic stricture. Several factors have been associated with an increased risk of stricture formation including: patients who have had an anastomotic leak; patients with gastroesophageal reflux; or anastomoses with braided silk sutures rather than with polyglycolic acid and polypropylene sutures (28). Strictures usually become evident 1–6 months postrepair or sometimes with transition to solid foods. Dysphagia caused by a postoperative stricture usually has an abrupt onset and patients may even develop difficulty with salivary secretions. The diagnosis is confirmed by contrast esophagography (Fig. 1A).

Strictures have traditionally been treated by bougienage or more recently by balloon dilatation; greater than 95% will respond to dilatation. (29–31) (Fig. 1B and C). In the minority of patients who require multiple dilatations, investigation for gastroesophageal reflux is imperative. Patients with reflux should receive aggressive medical treatment, often including gastric fundoplication. Strictures that are unresponsive to dilatation and fundoplication will require local resection and reanastomosis or possibly esophageal replacement (32,33).

GASTROESOPHAGEAL REFLUX AFTER TRACHEOESOPHAGEAL FISTULA REPAIR

Gastroesophageal reflux is common following tracheoesophageal fistula repair and is often difficult to control because tension in the lower esophagus to create the anastomosis can distort the gastroesophageal junction, leading to incompetence. Contributing factors to the severity of reflux in these patients may include dysmotility of the distal esophageal segment and, in some patients, delayed gastric emptying (34,35). Because approximately one-fourth of children with tracheoesophageal fistula repair will suffer gastroesophageal reflux disease severe enough to require fundoplication, surgeons must maintain close follow-up of these patients.

Many patients are placed on medical therapy for reflux immediately after tracheoesophageal fistula repair, and this is continued for at least 6 months. Patients with severe reflux, and consequently the majority of patients requiring fundoplication, will often exhibit those symptoms in the first 6 months of life. Symptoms may include vomiting, failure to thrive, anastomotic stricture formation, or pulmonary problems such as reactive airway disease, recurrent pneumonia, or evidence of aspiration. Diagnostic evaluation should include an upper GI study to look for anastomotic stricture as well as to determine if there is any pathology that could exacerbate reflux, such as duodenal stenosis or malrotation. An esophageal probe for 24-hour pH monitoring is helpful in defining the reflux severity.

The operative management of severe gastroesophageal reflux is approached with either open or laparoscopic surgery. Controversy exists over the use of a complete (e.g., Nissen) versus a partial (e.g., Toupet, Thal) fundoplication. Because dysmotility of the distal esophageal segment is almost universal in patients who have had esophageal atresia, partial fundoplication may diminish the risk of postfundoplication dysphagia. All approaches and types of repair appear to have a higher failure rate in this population of patients than in otherwise normal children treated for severe reflux (36–39).

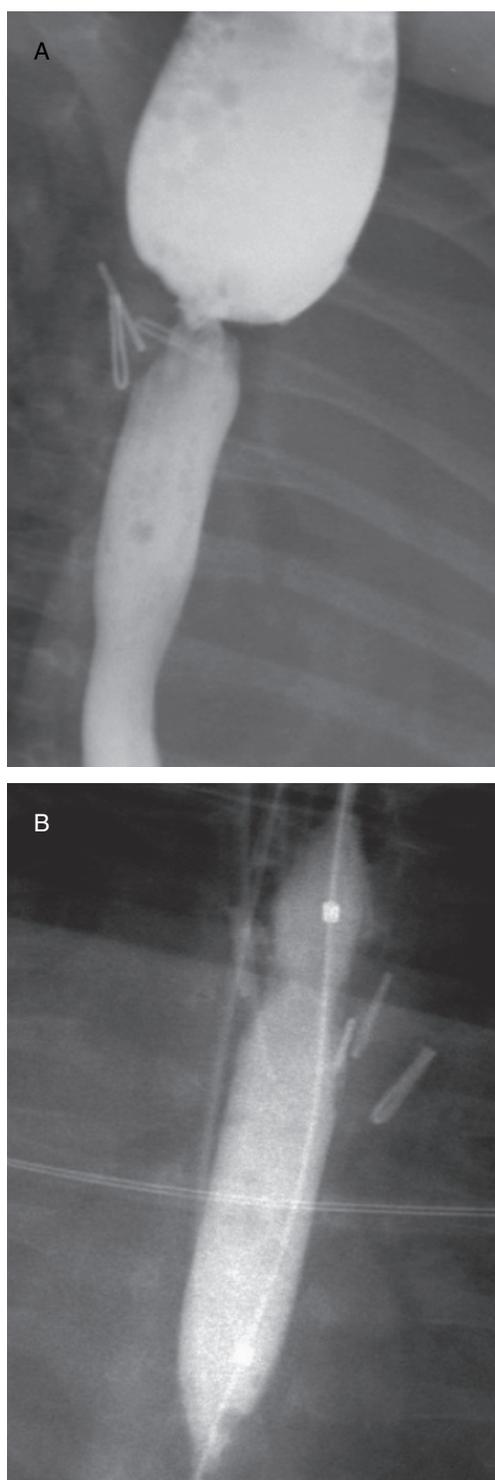


Fig. 1. (A) Esophageal stricture following thoracoscopic repair of tracheoesophageal fistula. (B) Balloon dilatation of anastomotic stricture. (C) Esophagogram following successful balloon dilatation.



Fig. 1. (Continued).

Concern has been developing about the long-term implications of gastroesophageal reflux following tracheoesophageal fistula repair. Perhaps one-third of these patients will have chronic reflux symptoms and evidence of reflux esophagitis. Some evidence suggests an increased risk for the development of Barrett's esophagus (40,41). One long-term study of patients with esophageal atresia showed esophageal inflammation in 51% of patients and Barrett's esophagus in 6% of patients (42). Although further study is needed to define these risks, and symptomatic improvement has been reported with age, patients should continue to be followed for the evaluation and treatment of reflux symptoms (43).

Recurrent Reflux-Induced Esophageal Stricture

Reflux-induced esophageal strictures may become symptomatic with food impaction or may be found simply in the evaluation of patients with gastroesophageal reflux symptoms. Strictures are documented either by contrast radiography or endoscopy (Fig. 2A). Most of these strictures are amenable to treatment with bougienage or balloon dilatation (44). Fundoplication to control the reflux is important to help prevent recurrence. Most recurrent strictures can be treated successfully with dilatation and fundoplication, but some patients will require additional dilatation (45,46). The recurrence rate of strictures may be decreased with the topical application of mitomycin C to the esophageal mucosa by endoscopy (47,48). Additional surgical options for those who do not respond to dilatation and fundoplication include local stricturoplasty (Fig. 2B) or repair of the esophagus using an intestinal or gastric patch (32,33,49). In the few patients who do not respond to the above procedures, the ultimate treatment is esophageal replacement with intestinal or gastric interposition (50,51).

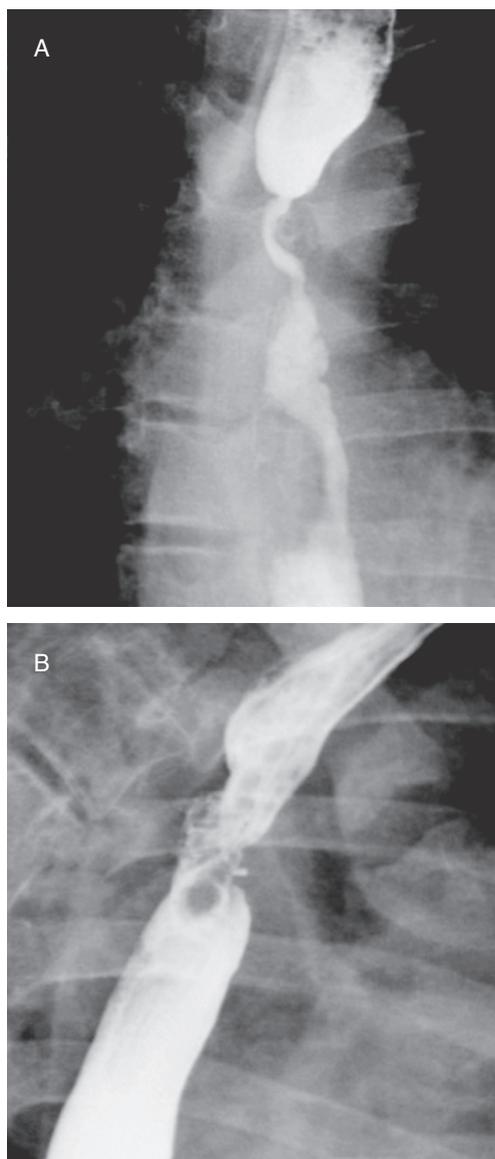


Fig. 2. (A) Peptic stricture caused by severe reflux in patient with cerebral palsy. (B) Esophagogram demonstrates irregular but unimpeded flow following thoracoscopic stricturoplasty of peptic stricture that had been unresponsive to balloon dilatation and fundoplication.

Barrett's Esophagus

Barrett's esophagus is a condition in which the esophageal mucosa undergoes intestinal metaplasia. The condition is sometimes asymptomatic but often occurs in the setting of gastroesophageal reflux disease. Most patients are adults that present with reflux symptoms and are found to have Barrett's esophagus after endoscopic biopsy. Adult patients with Barrett's esophagus have a 50- to 100-fold increased incidence of esophageal adenocarcinoma, which approximates a 1 in 200 annual risk (52). Comparable incidence rates are not well defined in children because of the paucity of reports

about patients with Barrett's esophagus in this age group. Patients who have had a tracheoesophageal fistula repair as an infant appear to be at higher risk for the development of Barrett's esophagus (40,41).

Patients with Barrett's esophagus should be offered treatments including surgery to control symptoms of reflux with the hope that this may also have a favorable effect on the metaplasia. Unfortunately, the relationship of adequate symptom relief to improvement of metaplasia or the prevention of adenocarcinoma is uncertain (53–58). Consequently, the long-term management in adults is controversial. The rare cases in childhood are burdened with the same concerns, and management is further hampered by even less published data (59,60).

Surveillance biopsies of the distal esophagus are performed to evaluate for dysplasia because the diagnosis is made at the histological level and no better screening test is currently available. Evidence of high-grade dysplasia or atypia would suggest the need for mucosal ablation or esophagectomy. Endoscopic luminal ablation of the esophageal mucosa treats the Barrett's metaplasia directly with laser or electrocautery. The procedure is effective in removing metaplastic mucosa but associated with a major complication risk of approximately 10%, including stricture and perforation (61,62). Long-term outcomes are not yet available in adults and thus the value of this modality in pediatric patients is unknown.

Perforation After Esophageal Dilatation

Esophageal dilatation is useful in the treatment of strictures from a variety of causes. Although strictures are amenable to treatment with bougienage or balloon dilatation, balloon dilatation by the interventional radiologist offers the advantage of a postdilatation esophagogram. The response to esophageal dilatation is somewhat dependent on the cause of the stricture: postoperative, corrosive, or peptic. Peptic strictures respond best to this treatment and require fewer repeat dilatations. The corrosive strictures are the most difficult to treat and have the highest rate of recurrence and perforation (63,64).

Fortunately, perforations following dilatation of an esophageal stricture are rare and usually well contained (Fig. 3). Small perforations can be treated with antibiotics, observation, and fasting (65–67). The mortality rate for perforation appears to be significantly less among children than for adults (68,69). For patients who do not improve over 24–48 hours, a repeat swallow study is performed. Continued leakage will require direct repair or diversion.

Achalasia Surgery

The surgery for achalasia has undergone significant change in the past two decades with the advent of botulinum toxin, pneumatic dilatation, and minimally invasive techniques (70). Although botulinum toxin provides relaxation of the lower esophageal sphincter, this is usually temporary and many patients require further therapy. Most patients undergo pneumatic dilatation, but one-half of these will have recurrent symptoms. Esophageal myotomy results in relaxation of the lower esophageal sphincter and can be performed either by a thoracic or abdominal approach (71). Postoperative esophageal reflux has been minimized by the inclusion of an anterior partial fundoplication (72,73). For this reason the laparoscopic approach is favored over thoracoscopy in the treatment of this disease. The thoracoscopic myotomy is reserved for patients



Fig. 3. Esophageal leak (contained) following balloon dilatation.

who have had previous extensive upper abdominal surgery. Indications for reoperations following esophageal myotomy include esophageal leak, the recurrence of achalasia, or the development of esophageal reflux.

A mucosal injury is usually recognizable during the operation and can be repaired primarily using absorbable sutures. The repair is then reinforced with an anterior fundoplication. Rarely an esophageal leak will become apparent in the first few days following surgery. These can be treated with drainage by interventional radiology along with the use of antibiotics, proper nutrition, and bowel rest. Diagnostic evaluation would include an esophageal swallow or upper abdominal CT scan with nonbarium contrast. Operative repair is required for large, poorly contained leaks.

After myotomy, approximately 10% of patients develop recurrence of symptoms. Recurrence can often be treated by pneumatic dilatation, and those who are unresponsive to dilatation can be treated with laparoscopic or open surgery (74). Most patients requiring reoperative surgery have problems related to the performance of the fundoplication; others have either a fibrotic stricture formation or an incomplete myotomy (75). All of the postoperative problems appear to be less common as surgeons gain greater experience with the procedure. Rarely a dilated esophagus does not decrease in size after myotomy and, if symptoms are severe, esophageal replacement is needed.

Esophageal Replacement Surgery

Replacement of the esophagus is challenging because of its unique anatomy and function. A variety of structures have been used for replacement, including skin tubes and enteric interpositions (colon, small intestine, stomach, and stomach tubes). All have complications that are significant for their frequency and severity (76–83). Graft ischemia is perhaps the most dangerous and problematic complication. Ischemia with

intestinal interposition can be mitigated by the use of microvascular anastomoses with cervical vessels to improve perfusion (84). Gastric or jejunal grafts can be used to replace failed colonic interposition (85).

One of the most frequent early complications is anastomotic leakage sometimes caused by graft ischemia. An anastomotic leakage can occur in the first few days following surgery and can be diagnosed clinically by salivary drainage from the wound. Drains are placed at the anastomotic site during surgery to help in the diagnosis and treatment of this situation. Appropriate drainage may allow healing of a leak without further operation.

Partial obstruction of a conduit can occur because of an anastomotic stricture or twisting of the graft (76). Strictures can be treated with balloon dilatation. Twisting of the graft is often a result of redundancy and is especially problematic with colon interposition. Treatment of redundancy is best approached from an abdominal incision which allows dissection into the mediastinum with care to avoid injury to the mesentery. The conduit can then be straightened, and the lower portion of the conduit resected and reanastomosed to the stomach.

Intestinal interposition between the pharynx and stomach can lead to reflux with ulceration of the graft. This can be treated with vagotomy and pharmacological acid suppression. For intestinal interposition, a partial gastric fundoplication can be performed (86). The reflux is thought to be responsible for the development of Barrett's metaplasia in the cervical mucosa, and patients should undergo surveillance endoscopy (87,88).

Esophageal Varices

Over the last 30 years, the treatment of esophageal varices has moved from large surgical operations to create portal systemic shunts to less invasive radiological and endoscopic techniques (89,90). Esophageal varices are caused by portal hypertension, which can be prehepatic, intrahepatic, or posthepatic. In children, the most common cause of portal hypertension is prehepatic caused by portal vein thrombosis; these patients usually have normal liver function. Biliary atresia leading to biliary cirrhosis is the most common cause of intrahepatic obstruction; most of these patients have significant liver disease. Posthepatic obstruction (Budd-Chiari syndrome) is caused by hepatic vein thrombosis, is less common, and presents with ascites and hepatic dysfunction.

The clinical presentation of esophageal varices depends on the etiology but is most often evident after 2 years of age with symptoms of upper GI bleeding. After cardiovascular stabilization and reversal of coagulation abnormalities if present, endoscopy is required to establish the diagnosis and initiate treatment. Endoscopic sclerotherapy typically controls the initial bleeding episode, and repeat sclerotherapy is usually successful at eradication of varices and prevention of recurrent bleeding (91,92). Variceal banding has also been shown to be effective, but is somewhat more difficult because of the size of the pediatric esophagus (93–96). Operative management is considered for failure to control initial bleeding or to prevent further episodes of bleeding (90).

Reoperative options for esophageal varices that persist or recur after sclerotherapy can include a transjugular intrahepatic portal venous shunt. These are placed by interventional radiology via the jugular vein and create a vascular shunt from intrahepatic portal veins into the hepatic veins. This strategy is only useful for intrahepatic portal

hypertension (89). Operative shunting procedures are most helpful for prehepatic portal venous obstruction but also can be used in selected patients with intrahepatic portal venous obstruction who have relatively preserved hepatic function. The variety of procedures includes portal caval, mesocaval, splenorenal, and meso- to left portal vein (Rex) shunts (90,97–99). For patients with poor liver function, the best option to treat portal hypertension is liver transplantation (90). Procedures for operating directly on the esophageal varices, including gastroesophageal devascularization (Sugiura) or esophageal transection, have very high morbidity and recurrence rates and are rarely used but may be helpful in patients whose portal, mesenteric, or splenic veins are not amenable to shunting (100–102).

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11

Reoperation after Failed Fundoplication and Other Gastric Operations

Mary Brindle, MD and Jacob C. Langer, MD

CONTENTS

FUNDOPLICATION
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FUNDOPLICATION

Fundoplication is usually a successful method for dealing with gastroesophageal reflux. The pediatric population requiring fundoplication is heterogeneous, indications for the operation are diverse, techniques of fundoplication vary widely from surgeon to surgeon, and reported success rates are also variable. In the last 15 years, the laparoscopic approach has become very popular, and this may have also changed the indications for and techniques of fundoplication (1).

The goals of a fundoplication are to reconstruct the normal anatomy of the gastroesophageal junction, to recreate the normal physiologic barriers to reflux, and to create a high pressure zone or valve to prevent reflux. Despite the large number of fundoplications that are done each year, complications are seen frequently, and many patients are candidates for reoperation (2,3).

Presentation of the “Failed Fundoplication”

A fundoplication can fail in one of two ways: recurrence of reflux, or complications related to the fundoplication itself. As with children who initially present with reflux, patients who have recurrent reflux can present with either symptoms of reflux (vomiting or pain), or complications of reflux (failure to thrive, esophagitis, or respiratory complications).

Complications of fundoplication include bleeding, infection, and injury to other organs, as with any operation. Longer-term symptoms relating to the fundoplication itself may be caused by a wrap that is too tight (gas bloat syndrome, inability to burp, or dysphagia) or by herniation of the wrap into the chest. Creation of a fundoplication

can also exacerbate an underlying esophageal motility disorder and worsen more subtle symptoms. Pain after fundoplication can occasionally be caused by correctable lesions such as paraesophageal hernia, which can become incarcerated.

Funduplications fail for a number of reasons (2,4). Patient comorbidities such as prematurity, neurological impairment, and other congenital anomalies may predispose children to wrap failure. Inaccurate diagnosis of gastroesophageal reflux in a child with central vomiting will doom the operation to failure. Finally, there are a number of technical pitfalls in performing the operation that may predispose to failure either by leaving a wrap that is too loose or too tight. Many of these remain controversial, and include type of fundoplication (particularly partial versus complete wrap), division of the short gastric vessels, closure of the hiatus, and use of a concomitant gastric emptying procedure.

Incidence and Predisposing Factors for Fundoplication Failure

The risk of fundoplication failure requiring additional surgical intervention is dependent on the population of patients being studied, and ranges from 4 to over 35%. The incidence is highest in patients with neurological impairment (5,6), respiratory disease (7), repaired esophageal atresia (8), and infants who were less than 1 year of age at the time of surgery (9).

In addition to having a high incidence of gastroesophageal reflux, neurologically impaired children often suffer from oropharyngeal dysfunction and develop frequent respiratory infections secondary to this form of aspiration. In patients who have ongoing respiratory issues postoperatively, it is often difficult to determine whether symptoms are related to ongoing reflux or aspiration from above. Gagging and retching also occur frequently in this patient population, and may be one of the reasons for performing a fundoplication in the first place (10). Unfortunately these symptoms commonly persist postoperatively despite an adequate fundoplication. If a wrap is performed in a patient whose primary symptoms are actually related to oromotor dysfunction, symptoms may actually worsen because esophageal clearance is impaired. The incidence of recurrent reflux is also higher in this population, likely because of increased intraabdominal pressure owing to spasticity, and the high incidence of delayed gastric emptying (11).

Children with esophageal atresia also have a higher rate of fundoplication failure, either because of recurrent reflux or, more commonly, dysphagia. This is caused by poor esophageal motility, which is exacerbated by the relative obstruction provided by the fundoplication.

A number of papers have documented a high risk of recurrent reflux in children undergoing fundoplication during infancy, particularly if the child was premature (12). The reasons for this are not well understood.

Nonoperative Approaches to Failed Fundoplication

Not all patients with a failed fundoplication require reoperation. Asymptomatic children, who are noted to have a “slipped” fundoplication (i.e., migrated into the mediastinum) with no demonstrable reflux, do not need intervention. Patients with symptoms of dysphagia that are found to have some degree of obstruction at the site of the wrap can be treated with dilation, although this must be done with caution as it may lead to recurrent reflux or in rare cases to perforation. Mild or moderate recurrent reflux can sometimes be managed pharmacologically with acid reduction and prokinetic agents (13).

Transesophageal endoscopic plication (TEP) is a novel endotherapeutic approach in the management of gastroesophageal reflux disease (14). This procedure involves the endoscopic placement of stitches circumferentially at the base of the esophagus to create a barrier to reflux. Its role in treating recurrent reflux postfundoplication is not established. However, fundoplication after failed TEP has been reported in adults and appears to result in similar outcomes to that achieved after primary fundoplication (15). Other endoscopic techniques that have been reported involve the submucosal implantation of polymer, or the formation of scar with the aid of radiofrequency. The latter procedure has been successfully used in a small group of children with recurrent reflux following fundoplication (16). Further data are necessary before recommending any of these techniques routinely in children who have failed fundoplication, although they may ultimately be very useful particularly in those children who are at high risk for surgery or who have a limited life expectancy.

Another option is the use of distal feeding via a gastrojejunostomy (G-J) tube (17). These tubes are placed through the stomach into the duodenum and jejunum under fluoroscopic guidance. The use of distal feeding tubes has the advantage of avoiding the major morbidity that accompanies an operation. However, there is a high rate of minor complications such as dislodgement, breakage, and clogging for patients with G-J tubes, and ongoing reflux is still an issue in these patients. In addition, the need for continuous rather than bolus feeding can be burdensome for the family.

Preoperative Assessment

Prior to considering surgical management of a failed fundoplication, it is important to develop an understanding of the reasons for the failure. These may include anatomical problems, physiological issues (particularly abnormal esophageal or gastric motility), central neurological problems, or psychological factors. The initial work-up should be targeted at these possibilities, and include a thorough history and physical examination, radiological assessment of the anatomy, evaluation of esophageal motility and gastric emptying, and neurological and psychological consultations if necessary.

An upper gastrointestinal (GI) contrast study can help to determine whether or not the wrap is intact and whether the wrap has slipped down onto the stomach, migrated into the chest, or formed a paraesophageal hernia. A contrast study may also reveal anatomic abnormalities such as malrotation or malformations of the stomach or duodenum that could contribute to the child's presentation. In patients with a history suggestive of reflux, but no reflux seen on the contrast study, 24-hour pH monitoring is a more sensitive technique. Upper endoscopy with esophageal biopsy is also useful in some cases to document the presence of esophagitis and to look for anatomical abnormalities.

Further investigations may include esophageal manometry if there is some question as to whether esophageal dysmotility may be contributing to the child's presentation. In these cases, the wrap itself may significantly worsen symptoms.

Although the need for a gastric emptying scan in patients being evaluated for initial fundoplication is controversial (18), it is probably a good idea in a child with recurrent reflux, because delayed emptying may be part of the reason for the recurrence and a gastric emptying procedure may be beneficial as part of the surgical solution (19).

In some cases there is remaining uncertainty whether reflux is contributing to the child's symptoms. Discriminating between oropharyngeal aspiration and reflux-related aspiration pneumonia can be challenging. The use of bronchoalveolar lavage (BAL)

to assess for lipid-laden macrophages is often used to test for reflux-related aspiration (20). Examination of BAL fluid for pepsin is another recently described strategy.

Surgical Approaches

REPEAT FUNDOPLICATION

Whether the first operation was done laparoscopically or open, repeat fundoplication is associated with a higher risk of injury to the vagus nerves, esophageal perforation, and wrap failure than the initial fundoplication (21). A previous open fundoplication is associated with a higher risk of adhesions, particularly between the liver and the stomach. Patients with a hiatal hernia or paraesophageal hernia may have adhesions that make it difficult to mobilize the stomach from the chest, and the risk of intraoperative pneumothorax is higher. However, previous open surgery is not a contraindication to a laparoscopic approach to repeat fundoplication, and it is usually worth trying a laparoscopic approach first (22).

Technical factors are very important in contributing to the success of a fundoplication, and careful attention to these factors is especially helpful when planning a reoperation (Table 1). Often the details of the operation must be made intraoperatively once the adhesions have been taken down and the anatomy can be identified. The goals of the operation are creation of an acute angle of His, ensuring adequate intraabdominal esophageal length, and creation of a loose but secure wrap. If there is a hiatal hernia, the crus must be closed securely; some surgeons have recommended pledgets for this purpose. If the hiatal hernia is very large, placement of a mesh patch may be necessary (23). In cases where the wrap has come undone, one must suspect that there has been tension on the wrap, and if the short gastric vessels were not taken the first time they should be divided in the reoperation (24). In cases of esophageal shortening, a Collis gastroplasty may be necessary in order to ensure adequate intraabdominal esophageal length. This procedure requires that a stapler be fired down along the lateral aspect of the stomach inline with the lateral border of the esophagus. This angle is difficult to achieve laparoscopically. In this situation, the case can either be converted to an open procedure or a thoracic port can be inserted so that a stapler can be directed down through the hiatus from the chest (25). This is a complicated maneuver. An alternative to this is the wedge gastroplasty, which involves a transverse staple line towards the esophagus from the lateral edge of the fundus (with bougie in place) followed by a

Table 1
Causes of Fundoplication Failure and Surgical Strategies for Revision

<i>Problem</i>	<i>Cause</i>	<i>Strategy for correction</i>
Wrap in chest	Short esophagus	Collis gastroplasty
	Large hiatal hernia	Pledget or patch repair
	Paraesophageal hernia	Take down and repair
Recurrent reflux	Delayed gastric emptying	Pyloroplasty or antromyotomy
	Wrap undone	Divide short gastric vessels
		Convert partial to complete wrap
Dysphagia	Esophageal dysmotility	Convert complete to partial wrap
	Wrap around stomach	Redo wrap around esophagus
	Crural closure too tight	Looser crural repair over bougie
Retching	Delayed gastric emptying	Pyloroplasty or antromyotomy

vertical staple line fired up from below. This alternative has been found to be successful and saves a thoracic approach or conversion to an open technique (26).

If the previous wrap is intact, it should be carefully examined for length, location (whether around the stomach or esophagus), and whether it is loose or tight. The crural defect should also be examined, as an overly tight closure can result in dysphagia. If dysphagia has been an issue, passing the bougie into the stomach under direct vision may be helpful in determining any site of hold-up. Standard use of a bougie to ensure that the wrap is not closed too tightly and that the closure of the crus does not impinge on the esophagus is recommended, especially in reoperation.

Patients with delayed gastric emptying and either recurrent reflux or incapacitating retching may be good candidates for a gastric emptying procedure. This can take the form of a pyloroplasty or an antromyotomy (19). If the repeat fundoplication is being done for dysphagia in a patient who has esophageal dysmotility (i.e., patients who have undergone repair of esophageal atresia), consideration should be given to converting to a partial wrap such as a Toupet or Thal.

SURGICAL JEJUNOSTOMY

Most surgical jejunostomies for feeding are done using a Roux-en-Y configuration (27,28). Some surgeons recommend this approach as the primary method for feeding the neurologically impaired child who has gastroesophageal reflux (29). Others use it for those that have failed fundoplication owing to recurrent reflux. The advantage of this approach is that it is an easier operation than repeat fundoplication, and it is less likely to be associated with repeated tube dislodgement and blockage than placement of a G-J tube (*see* Nonoperative Approaches to Failed Fundoplication). However, the disadvantage of surgical jejunostomy is that the patient continues to have symptoms related to reflux of stomach contents, and most of these children require long-term continuous feeds.

ESOPHAGOGASTRIC DISSOCIATION

Esophagogastric dissociation was originally described by Bianchi, and has recently been reported by a number of authors as an effective operation for neurologically impaired patients either as a primary procedure or as an alternative to repeat fundoplication (30,31). The operation consists of division of the esophagus at the gastroesophageal junction and oversewing of the distal esophagus. A 30- to 40-cm roux-limb is brought up in a retrocolic fashion to the esophagus and anastomosed in an end-end fashion. The patient is fed via gastrostomy. Although reflux is essentially eliminated by this operation, retching remains an ongoing issue. Because it is a major operation, complications have included subphrenic collections, esophagojejunal dehiscence, stenosis, bowel obstruction, and herniation of the stomach into the hiatus requiring partial gastric resection. The rate of morbidity appears to be higher in patients undergoing esophagogastric dissociation as a rescue therapy after a previous fundoplication.

GASTROSTOMY

Gastrostomy tubes provide tremendous value by allowing long-term enteral nutrition to be maintained in infants and children who are unable to take calories and fluids by mouth. Indications for gastrostomy insertion include failure to thrive, risk of oropharyngeal aspiration caused by pharyngeal dyscoordination or neurological impairment,

or inability to achieve adequate oral intake owing to chronic disease (cystic fibrosis, cardiac disease, inflammatory bowel disease, and others) or oral aversion. Gastrostomy tubes can be placed in a number of ways, including percutaneous radiologically guided, percutaneous endoscopic, laparoscopic, and traditional open insertion. All of these techniques have advantages and potential complications.

There are a number of situations in which a gastrostomy tube may require reoperative surgery, including leakage, migration, inappropriate position, and persistent gastrocutaneous fistula after tube removal (32). It is also important to recognize that the placement of a G-tube may unmask or exacerbate gastroesophageal reflux (33). There are a number of proposed reasons for this, such as alteration of the angle of His and increasing the volume of feeds that the stomach experiences. This problem is particularly common in the neurologically impaired child, and many of these go on to require fundoplication after insertion of a gastrostomy (34).

Migration of the Gastrostomy Tube

Gastrostomy tubes tend to migrate cranially on the abdominal wall as the child grows. This is a well-recognized problem that has not been well documented in the literature. In many cases, a tube that has been positioned in the left upper quadrant of an infant migrates up over the costal cartilage when the child gets older. In order to prevent this, gastrostomy tubes that are expected to be long-standing should be positioned just above the umbilicus on the left side. In cases where the migrated tube becomes painful or functions poorly, it should be replaced with a new, properly positioned tube.

Leakage Around the Tube

Leakage around a gastrostomy tube can be a debilitating problem causing significant skin break-down, soft tissue infection, and difficulty maintaining enteral nutrition. Most of these problems can be dealt with by an experienced team of nurses and stomal therapists who can provide parental support and advice. Appropriate barrier creams applied around a tube that leaks may prevent wound breakdown and allow for healing. If the tube has a balloon, it should be pulled up to the abdominal wall so that it blocks the tract. In addition, some balloons can migrate to the pylorus and cause gastric outlet obstruction, which exacerbates the leakage by increasing intragastric pressure. However, the balloon should not be pulled up too tight, as this may lead to necrosis and breakdown of the gastric and abdominal wall with worsening of the leakage problem. It is important that lateral tension not be applied to the tube as it passes through the abdominal wall, as this can lead to creation of a larger defect and increased difficulties with leaking. A wide-based phalange, a low-profile tube, or a button can help with this problem. Low-profile tubes and buttons are also less likely to be accidentally pulled out (35).

If these steps don't work, the tube can be removed and replaced by a smaller gauge tube, which allows the site to close down around the new tube. The old tube can also be exchanged for a longer tube that permits postpyloric feeding while the site heals. On rare occasions, feeds must be held entirely and the gastrostomy tube must be removed completely. If a new tube is required, it should be resited and the old site should be surgically closed if it doesn't close spontaneously.

Infection

Cellulitis and minor wound infections can occur around a gastrostomy tube. Most purulent discharge is related to foreign body reaction with the development of granulation tissue, rather than true infection. Any complaints of fever, redness, tenderness, or other skin changes should be evaluated. Necrotizing fasciitis has occurred around gastrostomy tubes, and this rare but devastating condition requires early diagnosis, broad-spectrum antibiotics, and aggressive abdominal wall debridement (36).

Dislodgement and Malposition of the Tube

Early tube dislodgement often requires operative repair. By 2 weeks, the tract between the stomach and the skin has usually formed well enough so that another tube can be introduced back into the stomach without difficulty. Once the tube is out, the tract may close within hours and a Foley catheter or other replacement tube should be inserted as soon as possible. Early dislodgement of a radiologically or endoscopically placed tube is more problematic, because there is nothing tacking the stomach up to the abdominal wall and the stomach is more likely to fall away from the abdominal wall. In these cases, leaking gastric contents may require surgical management. Attempts to thread a catheter into an immature tract should only be undertaken under fluoroscopic guidance. If this is not possible, attempts may be made to gently thread a Foley catheter into the old G-tube site but it is crucial that these attempts be performed carefully and without undue pressure. Tube position should always be confirmed by contrast study.

Gastric perforation and intraabdominal leak after placement of a gastrostomy tube require operative management. There is often free air on plain radiographs after placement of a gastrostomy tube, but the presence of significant pain and tenderness with this finding may indicate leakage. Enteric feeds should immediately be held and a contrast study via the gastrostomy tube should be performed.

In some cases during percutaneous gastrostomy placement, the tube may traverse the colon or small bowel, resulting in a fistula (37). Occasionally this may not be evident until after the first tube change, when the new tube is positioned in the colon or small bowel. These patients require revision of the gastrostomy with appropriate positioning of the tube in the stomach.

Gastrocutaneous Fistula

Most short-term gastrostomy tube sites will close spontaneously. Many surgeons use acid suppression with a histamine blocker or proton pump inhibitor in order to decrease the volume of gastric secretion and to minimize skin injury from leaking gastric acid. Topical application of silver nitrate to minimize granulation tissue may also help accelerate healing of the site. Factors that prevent closure include the presence of foreign body (retained suture or tube remnant), epithelialization, local infection, or granulomatous disease (i.e., Crohn's disease). The technique of tube insertion does not appear to have an effect. Length of time that the tube has been in place appears to be the most important determinant (38).

For those sites that have not closed by 1 month after tube removal, surgical closure is usually the best alternative. This can be done either as an open procedure or a combined open and laparoscopic approach. In most cases, a small incision around the tract, excising it completely down to gastric wall, and primary closure of the stomach and abdominal wall in layers will solve the problem and leave a small incision.

PYLOROMYOTOMY

Pyloric stenosis is one of the most common conditions faced by the pediatric surgeon. Results of surgical correction are generally excellent. The principles of repair as described by Ramstedt involve splitting the hypertrophic pyloric muscle from the antrum to its most distal aspect, where the hypertrophic muscle ends and the duodenum begins. Traditionally, the operation was performed using an open approach through either a right upper quadrant incision or, more recently, through a periumbilical incision. Many surgeons now prefer a laparoscopic approach, although it remains unclear whether there is any difference in outcome between the open and laparoscopic approaches (39).

An adequate pyloromyotomy requires that the pyloric muscle be divided completely, allowing the mucosa to pout into the pyloromyotomy and allow the superior and inferior margins of the pyloric muscle to move freely. Too aggressive a pyloromyotomy will result in perforation of the mucosa, and an incomplete pyloromyotomy will result in ongoing gastric outlet obstruction (40).

Perforation

The incidence of perforation during pyloromyotomy varies in the literature, but in most series it is rare and usually results from extending the pyloromyotomy too far distally. Perforation of the mucosa can and should be recognized by careful inspection of the mucosa after completion of the pyloromyotomy. If the perforation is recognized at the time of the primary operation, the surgeon can either repair the mucosal perforation alone or close the pyloromyotomy site completely and perform a second pyloromyotomy in a new position.

An unrecognized perforation is associated with the greatest morbidity and potential mortality in patients who have undergone pyloromyotomy. A high index of suspicion must be maintained in infants who have excessive pain, fever or instability postoperatively. In a child who is clearly unwell and has peritoneal signs on physical examination, no further investigation is necessary. If there is any question, a contrast study with water-soluble contrast material should be done. The surgical approach is the same as for the initial pyloromyotomy and the previous incision should be adequate. As with a perforation discovered at the time of initial pyloromyotomy, the options include simple closure of the perforation or complete closure of the pyloromyotomy with a second pyloromyotomy in a different location. Butressing the repair with omentum may also be helpful, although the omentum in an infant is usually quite small and flimsy.

Incomplete Pyloromyotomy

The incidence of incomplete myotomy is rare but should be suspected in patients who have ongoing emesis that is prolonged after surgery, recognizing that most children continue to vomit for 12–48 hours after surgery. Determining when to consider investigating and potentially reoperating on these children is a matter of clinical judgement. Gastroesophageal reflux is a common pediatric condition that must be differentiated from obstructive vomiting. Infants who have ongoing difficulties with spitting up postoperatively but gain weight and maintain their hydration are unlikely to have recurrent pyloric stenosis and appropriate parental counseling as well as medical management for reflux is usually sufficient. For infants who have ongoing pathologic vomiting, a careful review of the child's history, physical, and investigations may

make an alternate diagnosis clear. Radiological studies including ultrasound and upper GI contrast study may be confusing, as they frequently look the same as they did preoperatively long after a successful operation is completed (41). The duration of abnormal length of the pylorus after pyloromyotomy has been found to persist for at least 6 months, although the thickness of the muscle appears to normalize by 3 months. Prior to a second operation, an upper GI contrast study is valuable not so much to demonstrate ongoing pyloric stenosis, but to suggest an alternative diagnosis. It is important to recognize that duodenal duplications and webs may present in a fashion similar to pyloric stenosis, and may occur rarely in a child who also develops pyloric stenosis.

An incomplete myotomy can be approached through the same incision. Most surgeons prefer to do a new myotomy in a different site around the circumference of the pylorus, rather than attempt to extend the previous myotomy.

There are rare reports of repeat pyloromyotomy necessary in infants who have already had successful pyloromyotomy in the first few weeks of life (42). These infants may initially have resolution of symptoms but develop recurrence of projectile vomiting within the weeks following. It is felt that this may be progression of the initial condition with ongoing muscular hypertrophy.

In rare cases when a patient is not able to tolerate a second anesthetic, nonoperative management with atropine can be tried for a child with persistent or recurrent obstruction after a pyloromyotomy. This is also a recognized alternative to pyloromyotomy in some countries, and has been particularly popular in Japan (43). The majority of the time, this nonoperative approach would be considered only in patients who would not tolerate operative intervention, including patients with severe, uncorrected cardiac anomalies. The atropine is initially administered intravenously, and is gradually transitioned to the oral route. Even if this technique does not work, most children with pyloric stenosis will resolve over time, as long as hydration and nutrition are maintained.

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12

Reoperation after Duodenal Atresia Repair and Management of Duodenal Fistulas

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Duodenal obstruction, which includes duodenal atresia, stenosis, web, and annular pancreas, is a relatively common cause of neonatal intestinal obstruction, and occurs with a frequency of 1 per 5000 to 10,000 live births. More than 50% of infants with duodenal obstruction will have other congenital anomalies including congenital heart disease, esophageal atresia, and imperforate anus. Trisomy 21 occurs in approximately 30% of newborns with duodenal obstruction and may increase the risk of postoperative feeding problems.

Advances in anesthesiology and neonatal intensive care have improved the survival for newborns with duodenal obstruction, particularly those who are premature. The overall survival rate for these infants is now 90%, and normal growth and development can be expected for the majority of these patients. Mortality is generally caused by associated cardiac anomalies. Nevertheless, with the increased survival of very small and or complex neonates with duodenal obstruction, the rate of complications and reoperation will necessarily increase.

The incidence of complications leading to reoperation following repair of duodenal obstruction is unknown. However, a large, single-institution study recently reported a complication rate of 12% and a late mortality rate of 6%, stressing the need for long-term follow-up in these children. This chapter will focus on the management of the complications following surgical repair of duodenal obstruction.

INCIDENCE

The success rate for operative repair of duodenal obstruction is generally considered to be 95%. Most early deaths are caused by associated cardiac disease. The incidence of complications following duodenal repair is difficult to ascertain because the few studies that are available tend to include not only technical failures (such as strictures and leaks) but also diagnostic failures, which include patients with multiple obstructive lesions or malrotation missed at initial operation.

EARLY COMPLICATIONS AFTER REPAIR OF CONGENITAL DUODENAL OBSTRUCTION

Complications following repair of duodenal obstruction can be arbitrarily divided into early (<1 month following repair), late (>1 month to years following repair), and "very late" (years to decades following repair). Early complications include: missed multiple obstructions, anastomotic failure (leak or stricture), postoperative intestinal obstruction, and missed malrotation. Zhang et al. reported that seven of 12 reoperations among 298 patients with congenital duodenal obstruction were for "other duodenal atresias that were not discovered initially" (1). These authors stressed the importance of examining the distal gastrointestinal tract by injecting saline into the lumen beyond the repair to detect other obstructions.

Although greatly feared, anastomotic leak is seldom reported, and probably occurs only rarely. Zhang et al. reported only one leak (0.3%), and Escobar et al. reported two leaks (1.2%) in their respective series. A single case report described an enterocutaneous fistula secondary to an anastomotic leak in a premature infant. It was successfully treated with a course of octreotide (2). Anastomotic stricture is also either extremely rare or underreported, as it goes unmentioned in several large series. Spigland and Yazbeck reviewed 33 patients with duodenal repairs and placed one patient in the category of "ischemic stricture," but provided no other details except to state that the patient ultimately died of sepsis (3). Another rare early complication of duodenal repair is inadvertent duodenocolostomy, which leads to intractable diarrhea (4).

The management of infants with both duodenal obstruction and esophageal atresia has been the subject of debate. In the early 1980s, Spitz et al. reported a 33% survival for newborns with this combination of anomalies. They recommended that the tracheoesophageal fistula and esophageal atresia be repaired first, along with placement of a gastrostomy. The duodenal obstruction "may safely be postponed for a few days" (5). Thus, staged repair of these lesions became accepted as standard pediatric surgical practice. However, more recently, Dave and Shi have argued that "primary simultaneous repair of both anomalies without a gastrostomy is justified." Nevertheless, analysis of their data revealed that only two of nine patients were treated in this fashion (6). Furthermore, because of the incidence of additional anomalies (particularly cardiac) in these patients, the prolonged operative time required for simultaneous repair of these lesions must be viewed with caution. In fact, analysis of a larger case series by Ein et al. (24 patients) concluded that "staged repair (ideally within 1 week) is a safe and suitable management" (7).

Finally, Bittencourt et al. have argued that prenatal diagnosis of congenital duodenal obstruction not only improves outcome but lowers the early complication rate (8). This conclusion was based on an evaluation of 23 infants in whom 10 had a prenatal

diagnosis of congenital duodenal obstruction. Unfortunately, the small sample size, retrospective nature, and the potential selection bias of this study (mothers who underwent ultrasound evaluations likely had more prenatal care) make these findings preliminary at best.

LATE COMPLICATIONS AFTER REPAIR OF CONGENITAL DUODENAL OBSTRUCTION

Perhaps the largest single-institution experience with duodenal obstruction was reported by Escobar et al. in 2004. This report describes a late complication rate of 12% among 169 patients with duodenal obstruction. Of these 169 patients, 20 required additional abdominal operations, and several patients had more than one procedure performed (9). The common late complications of surgical correction of duodenal obstruction include: gastroesophageal reflux, megaduodenum, gastric ulcer, and motility disorders (10).

One of the most common late complications after duodenal repair is gastroesophageal reflux disease (GERD). It is not clear if GERD is truly a complication of the repair, or part of the pathophysiology of in-utero duodenal obstruction leading to gastric dilation, poor motility, and GERD at birth. Although the initial management of GERD associated with duodenal obstruction is medical, up to 5% of these patients will eventually require surgical correction with a fundoplication. Grosfeld and Rescorla reported that six of 103 patients required fundoplication 8 months to 7 years following duodenal repair (11).

Megaduodenum, dilation of the proximal duodenum resulting in functional obstruction, is the lesion most likely to lead to revision of the original surgical repair. The etiology is unclear, and at least some authors specifically state that it occurs in the absence of stricture. The diagnosis of megaduodenum is made by upper gastrointestinal contrast study (UGI). Surgical management of megaduodenum is accomplished by reducing the volume of the proximal duodenum until it has approximately the same caliber as the distal (postanastomotic) duodenum. There are a variety of surgical techniques that can be used. Revision of the duodenal repair was reported in 16 patients in Escobar's series. The operations included: tapering duodenoplasty or duodenal plication (seven patients), conversion of duodenojejunostomy to duodenoduodenostomy (three patients), redo duodenojejunostomy (three patients), redo duodenoduodenostomy (two patients), and conversion of gastrojejunostomy to duodenoduodenostomy (one patient).

Bleeding gastric ulcer is a rare late complication. Grosfeld and Rescorla reported three patients (3%) who required suture ligation for gastric hemorrhage related to ulcers (11). Escobar also reported two bleeding duodenal ulcers (1%) requiring operation (9). More recent series do not report this complication, suggesting that, with the advances in medical management of gastric acid secretion, these cases have all but disappeared.

The unusual association of duodenal obstruction and subsequent choledochal cyst has been noted by a number of authors. Grosfeld and Rescorla reported a fusiform choledochal cyst that developed 13 years after duodenojejunostomy. It was repaired by cyst resection, conversion to duoduodenostomy, and hepaticojejunostomy (11). Shih et al. attribute three cases to the world's medical literature, and added their own case, a 7-year-old girl with a type IV choledochal cyst with anomalous pancreaticobiliary

junction. She was successfully treated with hepaticoduodenostomy (12). Biliary atresia has also been rarely reported in patients following correction of duodenal obstruction. Spigland reported postoperative jaundice and biliary tract anomalies in four of 33 patients (12%) with duodenal obstruction. Two of these cases were confirmed to have biliary atresia. One was not treated and died, the other underwent a Kasai procedure but succumbed to sepsis (3).

The incidence of duodenal motility disorders following repair of duodenal obstruction is difficult to assess, because these cases are difficult to separate from megaduodenum (*see* previous discussion). Escobar lists seven (4%) late complications with “megaduodenum, motility disorder, \pm duodengastric reflux.” Dalla Vecchia, reviewing 138 patients with duodenal obstruction, similarly reported “late duodenal dysmotility resulting in megaduodenum that required tapering duodenoplasty in five (4%)” (13).

VERY LATE COMPLICATIONS AFTER REPAIR OF CONGENITAL DUODENAL OBSTRUCTION

The incidence of “very late” (years to decades) complications of duodenal repair is unknown, as few surgeons have been brave enough to report these cases. One exception is Ein et al., who reported three patients with “late nonfunctioning duodenal atresia repair” in 1986, and reported two additional cases in 2000 (14,15). The first report described three patients between 6 and 18 months, two of which were treated with multiple procedures including duodenal bypass with poor results; the third was treated with “plication only of the dilated atonic proximal duodenum,” with prompt return of gastrointestinal function. In the second report, two patients with obstructive symptoms 5 and 24 years following repair were described. The 5-year-old was “cured immediately” with plication of the proximal dilated duodenum; the second patient underwent a successful plication after undergoing other failed procedures.

However, duodenoplasty or duodenal plication is not successful in all cases. Spigland reported a 17-year-old with “megaduodenum with biliary reflux” who was initially treated (at birth) with duodenojejunostomy, who underwent partial excision of a missed membrane, takedown of duodenojejunostomy, and creation of a duodenoduodenostomy. A more radical technique for megaduodenum repair involves subtotal duodenal resection with the proximal jejunum used as an onlay patch. Endo et al. described this technique in two children (ages 16 months and 8 years) who had massive duodenal dilation (15 cm), multiple previous operations, and failure to thrive. Both children had excellent outcomes with catch-up growth (16). Compared to duodenal plication or duodenoplasty, the theoretical advantage of this operation is that it replaces the “diseased” duodenal wall (which may have intrinsic dysmotility) with “healthy” jejunum.

CONCLUSIONS

The vast majority of patients who undergo repair of congenital duodenal obstruction have excellent outcomes. Postoperative duodenal repair patients with poor function should be evaluated not only for reflux (with a pH probe and esophagram) but also for recurrence of the duodenal obstruction, which could lead to reflux symptoms or megaduodenum. Early reoperations may be required if a more distal obstruction or

malrotation was overlooked during the initial evaluation. Functional obstruction of the duodenum (megaduodenum) can develop months, years, and sometimes even decades after an initially successful repair. The most common approaches to megaduodenum repair are plication and duodenoplasty. However, more radical approaches, such as the resection with jejunal onlay patch, have been described.

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Reoperation after Surgery of the Small Bowel

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STOMA AND FEEDING ACCESS DEVICES

A series of novel techniques have been developed in pediatric surgery to mitigate stoma as well as feeding access problems for the pediatric patient. The sheer numbers of pediatric patients who might be benefited by such procedures, as well as patient complexities and comorbidities that include malnutrition, tortuous habitus, behavioral challenges, and other congenital and acquired defects, makes the routine management of stomas as well as feeding access devices challenging and fraught with considerable morbidity (1). Enterostomies have been described for a series of generic indications: feeding and/or medication access, access for antegrade irrigation, and the conventional decompression, diversion, and evacuation (1). This section will deal with those complications that either require reoperation or remedial correction following a complication of the enterostomy.

ACCESS FOR FEEDING OR MEDICATION ADMINISTRATION

Gastrostomy

The issues of gastrostomy placement relate predominantly to the nature of the indwelling product and the presence or absence of underlying gastroesophageal reflux. The techniques in most common use today for children include the percutaneous

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endoscopic gastrostomy (PEG) (2), a laparoscopic assisted placement of a gastrostomy, or a conventional open Stamm gastrostomy, either as a part of another procedure or as an isolated event in selected patients. For the PEG, typically a long and somewhat stiff original "kit" tube is replaced to a skin-level device following initial tube placement. The Gauderer "Genie" device can be exchanged to a skin-level button either at the time of initial placement or within a couple of weeks thereafter; the latter is preferred to allow initial postoperative swelling to subside and the appropriate "fit" of the tube length before cutting (3). After either a laparoscopic assisted gastrostomy or an open Stamm gastrostomy, either an externally extending tube (balloon catheter, flange-tipped catheter) or a skin-level device (button device) can be placed at the initial access procedure. Subsequently, first tube exchange can be deferred for an extended time; and when the exchange is done (except for those that require simple tube balloon deflation), local injectable or topically applied anesthetics are used. Tube extraction is done by an outward pull against an inward supporting hand to provide resistance of the abdominal wall to permit easy tube withdrawal. Replacement with an appropriately sized diameter (same size or 1-F size diameter up or down) and measured-for-length skin-level button with an inflatable balloon (MIC-KEY button device) would be the safest replacement. Demonstrating an intact balloon preinsertion, the device is passed through a well-lubricated tract. After balloon inflation, the intragastric position of the device is confirmed by the to-and-fro instillation of saline/aspiration of gastric content. Failure to be secure in this assessment, especially with tube replacement for the first time or with delayed tube replacement after a tube has been dislodged for several hours, should be followed by contrast radiography to assure appropriate position and the absence of contrast leak, which would be seen if the stomach/parietal peritoneal "seal" had been disrupted and the stomach pushed away from the anterior abdominal wall. In the advent of disruption, operative replacement is emergently indicated with control of the peritoneal contamination (4).

Gastrocutaneous fistulas that persist following gastrostomy tube removal might be managed with conservative or operative techniques. Those patients whose tube has been in for more than 6 months, those who have distal bowel dysmotility or delayed gastric emptying, or those with poor wound healing from a variety of causes are prone to persistence of the stomach-skin tract opening after gastrostomy device removal. Trials of topical sclerosing agents (e.g., silver nitrate) to cauterize and therefore "coapt" the mucosa-lined tract most often will not work. Therefore, operative closure of the tract may take the form of simple tract excision, which requires temporary tube reinsertion during the immediate postoperative period (5), to formal takedown of the fistula and gastric wall suture closure.

Tube-site granulomas or local skin irritation are the most common issues associated with enterostomy tube placement, but in the era of skin-level device use, which minimizes tube motion and distraction by attached feeding tubes, this issue is much less prevalent. Topical sclerosing agents (silver nitrate) to cauterize and destroy granulation tissue are often useful, but equally beneficial are topical applications of corticosteroid ointments to minimize the local inflammatory response.

Intractable gastroesophageal reflux after gastrostomy tube placement has a series of long-term management options: distal permanent feeding jejunostomy, antireflux fundoplication, and even gastric disconnect procedures. However, for short-term management, the simplest and most prudent option is initial pharmacologic and nutritional medical management to minimize reflux, maneuvers that would include

proton pump inhibitors, gastric motility enhancing agents, and altered feeding regimens in which smaller volume constant infusion feeds are administered. If feasible, a trans-gastric guided transpyloric feeding tube may be inserted under fluoroscopic guidance, with the gastrostomy device being concomitantly replaced to facilitate gastric decompression. The same objective, though a less reliable and long-lasting access route, can be achieved by placement of a nasojejunal tube.

Jejunostomy

The major advantage of a jejunal access route is the bypass, by virtue of its postpyloric location, of the stomach, a strategy that should minimize the issue of gastroesophageal reflux. Nasojejunal tubes are at best a short-term solution associated with patient discomfort because of tube nasal location, the facilitation of gastroesophageal reflux because of its trans-lower esophageal sphincter placement, and because of frequent occlusion or dislodgement, which requires repeated replacement, often necessitating repeat fluoroscopy (6). Therefore, alternative operative techniques include a roux-Y configured access device or a direct jejunostomy feeding tube or button placement. The technical challenges of jejunal feeding tube placement are several. First, when working through a limited incision, the proximal jejunum just beyond the Ligament of Treitz must be identified with certainty and chosen for the access site. Second, when creating a roux-Y dissection for a jejunal feeding stoma, bowel ischemia must be avoided and adequate length of the roux-Y limb must be fashioned to mitigate retrograde regurgitation of feeds, which could initiate reflux. Third, for adolescents, it is often preferable to directly place the feeding device into the jejunum as a tube or skin-level "button;" however, if a balloon device is inserted in a younger child, the inflated balloon cannot be so large as to compromise the luminal size of the jejunum. In summary, jejunostomies are technically difficult, are frequently associated with more local skin irritation and breakdown, and they also are associated with internal herniation and peristomal bowel obstructions.

ACCESS FOR ANTEGRADE IRRIGATION

Appendicostomy, Tube Cecostomy, Tube Sigmoidostomy

Bowel management protocols have now been refined to offer patients suffering from incontinence or chronic dysmotility syndromes an alternative to a permanent decompressing enterostomy with an external appliance. Instead, a port for irrigation of the colon in an antegrade fashion permits a controlled "on the toilet" evacuation of the colon, which eliminates the need for a formal stoma and appliance. Appendicostomies are most often done, or a comparable conduit may be constructed by tubularizing the ileum in the absence of an appendix (7,8). Retrograde leakage of irrigant or colon content onto the abdominal wall is the most common complication. Operative plication of the cecal wall around the conduit is the most commonly applied remedial technique that minimizes such leakage, whereas creation of an internal intususcepted valve is less desirable and more difficult to "hold." Application of a prosthetic "button" device can also be used, but both skin irritation as well as leakage is not an uncommon consequence (9).

ACCESS FOR PROXIMAL DECOMPRESSION AS WELL AS DISTAL IRRIGATION

Mikulicz, Bishop–Koop, and Santulli Enterostomies

Initially described for application to neonates with distal intraluminal intestinal obstruction such as characterizes meconium ileus, these procedures were applied to minimize the need for subsequent laparotomy and the morbidity of stomal takedown in a patient population that had an extremely high overall morbidity and mortality (10). Today, a functional comparable outcome is most often achieved by the insertion of an enteral “T-tube,” which serves as a route for postoperative intraluminal irrigation while decompressing the proximal bowel (11). Perhaps the major advantage that these procedures offer is the simple provision of an access route to the distal jejunioileum, through which solubilizing agents or antegrade enemas might be administered to assess as well as “open” the obstructed distal intestine. Rarely, such patients will avoid formal reoperation and transperitoneal takedown and anastomosis of proximal and distal intestine at the enterostomy site; however, the procedure can be done at a time when the patient is physiologically and nutritionally optimized.

DECOMPRESSING ENTEROSTOMIES

The creation of an optimal stoma for a child requires a series of appropriate decisions, and because small bowel stomas in particular are typically temporary, the planning must also take into account a stoma closure strategy. The selection of a preferred anterior abdominal wall “stoma site” must assess previous scars and incisions, the belt line, as well as the stoma appliance position when both sitting and upright, an assessment that should be done jointly by patient (or parent) and an experienced care provider. Stoma placement must also consider in a right-to-left orientation the use of the rectus muscle as the preferred location for bowel passage that most appropriately will aid the fixation of the conduit to the abdominal wall. Selection of the correct level of the gastrointestinal tract, small or large intestine, the correct proximal-distal orientation, and the appropriate length from the anorectum if an eventual pull-through is planned are each decisions that are facilitated by careful dissection and exploration at the time of stoma placement. The next critical decision is to assure the vascularity of the exteriorized intestine, a loop stoma being the safest whereas an end stoma must be carefully constructed to prevent ischemia. This decision also requires a plan for fixation of the stoma and its mesentery to the abdominal wall, a maneuver designed to minimize the chance of ischemia caused by peristomal twisting or volvulus as well as to minimize the opportunity for stoma prolapse. Finally, stoma construction must also consider stoma maturation. The construction of a “matured intestinal bud” is preferred in older children as well as in a loop diverting stoma. Stoma “maturation” may require a dissection on the bowel wall for a length of 6–8 cm to achieve an intestinal segment of sufficient length to facilitate turning back on itself, creating a “bud” for appropriate appliance application. This same strategy, however, is unsafe in the small premature baby, especially when stricken with ischemia-producing entities such as necrotizing enterocolitis (NEC). Exteriorizing 2–3 cm of intestine in such infants will result in a “stoma self-maturation,” which will eventually produce an acceptable stoma protrusion suitable for an appliance.

Enterostomy Closure

The timing and preparation for enterostomy closure varies with the level of diversion, as well as the initial indication for the stoma. In general, more proximal small bowel stomas should be closed sooner, typically 4–6 weeks after placement, whereas colonic stomas will provide greater flexibility in the timing of closure because they are less likely to produce fluid, electrolyte, and nutritional consequences. In all ischemic enteropathies, a distal contrast study as well as intraoperative irrigation of distal bowel should be routinely done prior to, as well as during, operation to assure luminal patency. For colostomy closures, a mechanical and antibiotic bowel preparation should be done prior to the procedure, a practice that may be avoided in most small bowel stoma closures, though a preoperative clear liquid diet the day prior to operation may be prudent. For all stoma takedowns, antibiotic prophylaxis is indicated.

Enterostomy Complications (see Table 1)

Prolapse of a small bowel stoma is not uncommon in the pediatric patient, and the extrusion may range from a minor self-reducing prolapse to a major prolapse with the elongated segment at risk for traumatic or ischemic injury. The prolapsing segment may be the proximal functional stoma or the distal nonfunctioning portion of a loop or end enterostomy. Prevention of the latter is typically done by making the stoma fascial and cutaneous opening very small. Reduction of the prolapsed segment may require general anesthesia, manual reduction, or even laparotomy and a combination of a push-pull reduction to salvage the prolapsed intestine, often rendered ischemic because of occlusion of the venous return. After reduction, prevention of subsequent

Table 1
Common Complications of Enterostomas

Prolapse
Stricture
Retraction
Wound separation, dehiscence
Wound infection, postoperative sepsis
Parastomal hernia
Intestinal wall separation or perforation with catheter change
Exteriorization of wrong intestinal segment or end
Intestinal obstruction (adhesion, internal hernia)
Intestinal torsion with ischemia
Fistula formation
Perforation by feeding or irrigating catheter
Poor appliance fitting and leakage
Psychological trauma
Skin excoriation, candidiasis, dermatitis
Mucosal excoriation and bleeding
Granulation tissue of mucosa skin interface
Variceal bleeding with portal hypertension
Electrolyte imbalance
Acidosis (caused by urine absorption in the distal loop of intestine)
Fecal impaction (in the distal loop of intestine)

prolapse, short of stoma revision, can be done by an internal bowel fixation or an external circlage pursestring stitch at the stoma-cutaneous junction or by a suspending U-stitch through the abdominal wall tied over an intraluminal and an external prosthetic pledget (12).

Stricture typically follows stoma ischemia or a technical error of too small a fascial opening. Revision of the stoma, if repeated and graded dilatation proves unsuccessful, is typically accomplished by an extraperitoneal operative approach in which both skin and fascia are incised in a radial fashion to enlarge the stoma orifice. Formal laparotomy and stoma revision is only indicated when a parastomal hernia is associated with the stricture. Retraction with resultant inadequate stoma height is certain to produce appliance-sealing difficulty as well as potential spillover into a distal lumen in the presence of a loop enterostomy. With a loop stoma, such retraction might be prevented with either a skin bridge or a supporting rod/rubber ring technique (1). Stoma revision may be done either by mobilization and advancing more intestinal length through an incision around the stoma or a separate laparotomy may be necessary.

Stoma takedown and bowel reanastomosis have a substantial complication rate in the pediatric population. Complications include wound infection, wound or anastomotic dehiscence, bowel obstruction, or intestinal fistula formation.

COMPROMISE OF INTESTINAL INTEGRITY: PERFORATIONS/LEAKS/FISTULAE

Etiologies

Traumatic intestinal injury following blunt or penetrating abdominal trauma is uncommon; however, with a high degree of suspicion (abdominal seat belt sign, lumbar vertebral fracture), occasional application of peritoneal lavage, and appropriate additional diagnostic testing that may include imaging as well as laparoscopy, a resection of ischemic or perforated bowel can be done and continuity restored with a primary anastomosis (13). Intestinal ischemia that progresses to necrosis in the pediatric patient may occur secondary to adhesive closed-loop obstruction and various low-flow states of shock, but the “deadly bilious vomit” most often will herald a mid-gut volvulus or malrotation of the colon. Reoperative “second-look laparotomy” may be useful in an effort to define extent of irreversible ischemia, and in doing so, limit the extent of resection and the creation of short bowel syndrome (SBS) (*see next section*) (14). Iatrogenic intestinal injury may follow open operative procedures such as adhesiolysis, interventional radiologic procedures such as the drainage of intraabdominal abscesses, endoscopy such as colonoscopy with biopsy, or minimally invasive laparoscopic procedures where port placement, scope advancement, electrocauterization, or other instrument manipulations might cause a bowel perforation (15). Recognized small bowel leakage, in the absence of a more distal intestinal obstruction, is generally managed by either laparotomy or laparoscopy with debridement of the site of perforation followed by a primary single or two-layer closure. It might also be prudent to consider a limited intestinal resection with anastomosis. Both options might be augmented with topical omental patching. Perioperative antibiotic therapy is appropriate. If instead the patient is unstable, if distal obstruction is present, or if there is a need to “protect” a distal anastomosis, then either the creation of a controlled fistula with enterostomy tube insertion (a T-shaped tube is a favorite choice) (16) or enterotomy exteriorization as a stoma may be preferred.

NEC

The controversy surrounding the preferred treatment of NEC has in part been clarified; but the spectrum of intestinal ischemia-focal perforation to NEC-totalis—as well as the spectrum of the afflicted host—micropremature baby to term-birth infant—make appropriate operative intervention a challenge. This procedure selection may also influence outcomes that minimize the complication of SBS, and therefore several options for treatment exist.

DRAINAGE VERSUS LAPAROTOMY

Traditional operation for NEC involves laparotomy for resection of grossly diseased bowel, debridement of peritoneal contamination, and fecal diversion through enterostomy formation. Peritoneal drainage was introduced in the late 1970s for the management of NEC in newborns who were felt to be too small or too ill to undergo laparotomy (17). Initially conceived of as a temporizing measure, peritoneal drainage has increasingly been adopted as a primary surgical modality for many newborns with NEC, especially those with very low birthweight (18–20). Comparisons of peritoneal drainage and laparotomy for NEC have been heavily confounded by a selection bias that favors drainage in the smallest newborns and laparotomy in the larger newborns (21). Outcomes with either peritoneal drainage or laparotomy may be more reflective of comorbid patient factors, rather than a particular surgical approach (22).

Published data regarding SBS in newborns treated with peritoneal drainage versus laparotomy are difficult to interpret. Nonrandomized retrospective series comparing drainage and laparotomy suggest that the incidence of SBS following peritoneal drainage is 4–9%, compared to 10–46% following laparotomy (23,24). One multi-institutional, prospective study of peritoneal drainage of NEC reported SBS in only one patient of 44 treated with primary drainage (2%) (18).

Simple peritoneal drainage is typically performed at the patient's bedside in the neonatal intensive care unit (18,19). Using sedation and local anesthesia, a small incision is made in the lower quadrant of the patient's abdomen, in which the majority of gas or fluid resides (typically the right side). Using careful blunt dissection, the peritoneal space is entered, fluid and gas are evacuated, and a Penrose or small catheter drain is introduced. The drain is sutured and left in place until the drainage abates, systemic illness resolves, and intestinal motility returns. Data reveal that 24–36% of patients initially treated with peritoneal drainage will subsequently require laparotomy for continued clinical deterioration, with subsequent salvage rates dependent on patient size and time between drainage and deterioration (18–20). Despite the presence of a perforation, approximately one-third of patients treated with peritoneal drainage alone will seal the perforation and recover full intestinal continuity without further operation. This remarkable observation, which illustrates the restorative capacity of the neonatal gastrointestinal tract, has led many to advocate for simple drainage as the preferred bowel-preserving operation for NEC. However, in a randomized prospective multiinstitutional study focusing on laparotomy versus drainage only, the type of operative procedure done for perforative NEC did not influence early disease outcomes in the preterm infant (25).

PATCH, DRAIN, AND WAIT

An interesting modification of simple peritoneal drainage—the “patch, drain, and wait” strategy—has been championed by Moore and colleagues (26). This approach is

based on the principle of resecting no bowel and performing no enterostomies. This minimal resection strategy places bowel preservation as the primary goal. It is based on the observations of “spontaneous bowel anastomosis” in isolated cases of meconium peritonitis and drained NEC. In addition, it seeks to mimic the environment of in utero intestinal atresia, where the peritoneum is kept sterile (with antibiotics and drainage) and necrotic areas of bowel are processed by the body with maximal preservation of marginal bowel through the formation of hypoxia-induced angiogenesis. Moore has demonstrated a 24-fold elevation of vascular endothelial growth factor (VEGF) in the peritoneal fluid of these neonates, and he suggests that this angiogenic protein-induced “good-angiogenesis” may in part be responsible for bowel perfusion and preservation (26).

The newborn is taken to the operating room for midline laparotomy. Gross contamination is debrided and irrigated. A Stamm gastrostomy is performed for postoperative gastrointestinal decompression. The small bowel and large bowel are inspected for NEC. Focal areas of necrosis or perforation are “patched” using simple imbrication with interrupted silk sutures or by securing adjacent healthy bowel or omentum to these areas. Long segments of clustered intestinal perforation are treated with resection and primary anastomosis. In cases of extensive, widespread intestinal involvement, no “patching” maneuvers are performed. “Draining” the abdomen involves placing a Penrose drain on each side of the abdomen from the hemidiaphragm down to the pelvis, exiting out both lower quadrants. “Waiting” involves intravenous antibiotics, total parenteral nutrition (TPN), decompression through the gastrostomy, and ongoing Penrose drainage. Fecal fistulas may form along the drains, but second laparotomy is strictly avoided during the first 14 days. If gastrointestinal continuity has not returned by 2 months, a second operation is undertaken at that time. A single center experience of 23 NEC patients over 15 years managed with “patch, drain, and wait” revealed no mortality, no morbidity, no stricture, no progressive sepsis, and no SBS (26).

PROXIMAL DIVERTING ENTEROSTOMY

Another operative strategy that involves no bowel resection is the use of a proximal enterostomy (28). Proximal enterostomy without resection employs the principle that diversion of enteric contents allows downstream NEC to subside in the absence of inflammatory substrate. It also has the putative benefit of decompressing distended bowel where the intraluminal pressure itself could compromise capillary flow, further aggravating transmural ischemia because of an impaired microcirculation. By resecting no bowel, this approach permits the patient’s own abdomen to sort out viability, marginality, and necrosis, and thus maximize bowel length preservation.

At initial laparotomy, an enterostomy is fashioned proximal to the intestine involved with NEC. Peritoneal drainage may or may not be used as an adjunct. A second laparotomy is eventually done after the inflammatory and septic response has subsided, and limited resection with one or more anastomoses restores intestinal continuity. A single-institution series reported overall initial operative survival of 74%. Of the 14 long-term operative survivors, two (14%) had SBS, but both subsequently outgrew their need for TPN (28).

RESECTION AND PRIMARY ANASTOMOSIS

After an initial resection for NEC, the creation of stomas requires reoperation for restoration of intestinal continuity. This “staged approach” to intestinal ischemia and

perforation could result in a degree of bowel length loss because of stoma creation followed by “takedown.” Therefore, resection followed by primary intestinal anastomosis was promoted as an alternate approach if outcomes would only approximate those achieved with the staged technique. Several groups subsequently trialed and reported favorable patient outcomes with this strategy (28). Thereafter, a subset of NEC, the focal neonatal intestinal perforation, began to be recognized with greater frequency; namely, the premature baby with isolated ileal perforation, often following indomethacin therapy (29). It was this group of babies that were particularly amenable to the primary resection and anastomosis strategy. The entire concept was thereafter strengthened by a more recent report of a substantial incidence of both morbidity (68%) and mortality (26%) related to the staged technique of stoma creation and closure in premature babies (30).

The application of resection and primary anastomosis is designed most appropriately for limited intestinal disease as well as in those circumstances where a clear demarcation exists between viable and compromised intestine and where peritoneal cavity contamination is either absent or limited. Ancillary techniques have been applied that facilitate the definition of viable intestine (31), applying intraoperative techniques that define intestinal perfusion and putative viability. A limited resection is done, followed by a careful end-to-end primary anastomosis using precise technique designed to minimize further intestinal injury. Both anastomotic leak as well as narrowing with delayed obstruction has been reported as outcomes.

Small Bowel Obstruction (SBO)

Adhesive SBO secondary to either congenital or postoperative adhesions can produce ischemic injury and perforation of the small bowel. Typical early postoperative adhesive SBO is less likely to result in ischemic change; however, late (weeks to months to years) adhesive obstruction always presents with that risk. The postlaparotomy “free-air” is typically gone in 1–2 days after neonatal laparotomy and in 3–5 days following abdominal surgery in older children (32). A useful adage of “never let the sun set on SBO” would most often avoid ischemic injury. It is also useful to bear in mind that an incidence of postoperative obstruction in children following neonatal surgery approximates 5–10%, and is most common in children who had abdominal wall gastroschisis defect repair or those following malrotation surgery (33). Clinical assessments that includes fever, tachycardia, signs of peritoneal irritation, white blood cell count elevation or left shift, or elevated levels of inflammatory mediators (e.g., sedimentation rate, C-reactive protein) are useful, as are radiographic findings of a persistently fixed and dilated intestinal loop, evidence of a closed-loop obstruction, or signs of pneumatosis intestinalis. All of these parameters suggest the potential for irreversible ischemic injury and a need to intervene operatively. Abdominal computed tomography (CT) has become a recent useful adjunct in defining SBO, both as a tool to define proximal/distal intestinal size disparity as well as an adjunct to define mesenteric vessel orientation when diagnosing rotational anomalies (34). The index of suspicion and the need to operate early for SBO should be heightened in the face of a competent antireflux fundoplication, an anatomy that functionally creates a “closed-loop” intestinal obstruction.

The conservative management of adhesive SBO in children has been shown to be safe as well as of substantive success if signs of intestinal ischemia are closely followed and used as a reason to operate. As many as 40% of children managed nonoperatively

have been spared a laparotomy (35,36). However, classic teaching has advocated for the value of early operation for SBO (37); and in the laparoscopic era, early intervention is not only feasible, but it can be done in most patients with a reduced operative morbidity (38).

INTUSSUSCEPTION

Ischemic injury may follow either recurrence or incomplete hydrostatic reduction of an intussusception. In contrast, complete hydrostatic reduction will not typically occur if irreversibly ischemic intestine is left behind. Perforation during attempted hydrostatic reduction for ileocolic intussusception has several unusual features: air as the hydrostatic medium is characterized by smaller perforations; less bowel damage and certainly less peritoneal contamination; the perforation site is typically distal to the intussusceptum and would likely be in the distal colon; and laparotomy with resection and anastomosis (air contrast) or resection and diversion (barium contrast) would be the preferred management.

Postoperative intussusception is typically a small bowel intussusception. The typical presentation is a delayed postoperative SBO 7–10 days after the initial abdominal procedure. Fortunately, such patients will rarely progress to having intestinal ischemia. However, pneumatic reduction is not a treatment option for the persistent obstruction, and laparotomy or laparoscopy is needed to manually deintussuscept the bowel (39,40).

MALROTATION WITH VOLVULUS

A Ladd procedure to treat this entity inherently is a frequent cause of subsequent SBO, likely because it includes a substantial amount of dissection and mobilization of the duodenojejunum, maneuvers that create adhesions that have the benefit of “fixing” the intraabdominal position of the bowel designed to prevent recurrent volvulus (41).

Interestingly, more recent reports of laparoscopic Ladd procedures seem to likewise prevent recurrent volvulus while producing a lower frequency of postoperative adhesive obstruction (42).

ISCHEMIC ENTEROPATHY

This entity rarely occurs after a low perfusion state that might characterize a postoperative patient undergoing correction, complete or incomplete, of cyanotic congenital heart disease. However, this entity is difficult to diagnose before irreversible ischemic injury has occurred. A high index of suspicion is required, clinical or radiographic findings as noted above for SBO might be present, and a background setting such as a heterotaxia syndrome may herald a risk for this complication (42). In addition, findings compatible with an abdominal compartment syndrome, suggested by bedside clinical exam and confirmed by elevated intravesicle or intragastric pressures, would be a sign compatible with an intraabdominal catastrophe.

SBS

Medical Management

The preferred medical management of SBS or intestinal insufficiency is prevention. Therapy is designed to enhance enteral feeding volume and calories, minimize or even eliminate TPN support, and in doing so, restore normal nutritional physiology, growth and development, and protect the patient from exogenous nutrient toxicity

or endogenous nutrient deficiency. Any such maneuver that protects the baby from intestinal failure-induced progressive liver dysfunction will assure normal growth and development and obviate consideration for liver or liver/bowel transplantation. Certainly, aggressive nutritional support coupled with medical management should be the initial strategy for SBS. A series of pharmacologic maneuvers might also be considered. H-2 blocking agents or proton pump inhibitors are designed to mitigate or at least minimize the hypergastrinemic state, which characterizes massive small bowel resection. On occasion it is also useful to administer motility slowing agents such as loperamide, which by slowing transit are designed to enhance mucosal exposure to intraluminal nutrient as well as decrease either stoma output or anorectal output and secondary perianal skin excoriation. The greatest but not proven opportunity in medically treating SBS would be the administration of gut trophic agents. Either parenteral or enteral glutamine has not been unequivocally proven in children to be efficacious, and the other experimental trophic agents, amongst which are epidermal as well as hepatotrophic growth factor, have only had efficacy in experimental animal model-created SBS. This leaves nutritional manipulation as the most efficacious medical therapy for SBS. Utilized diets are typically elemental in nature, and what is varied is administration technique (bolus versus continuous feeding), site of delivery (stomach versus small bowel), caloric density, and adjunctive nutrient agents. The current agents added include vitamins, trace metals, and probiotics. The slow but steady progression of enteral feeds and concomitant tapering of parenteral support is the combination therapy designed to prevent the development of liver dysfunction that typically characterizes SBS, TPN, and repeated episodes of bacterial overgrowth and systemic sepsis. Therefore, the prevention of such bacterial overgrowth would be efficacious; but to date, mostly empiric cyclic enteral antibiotic administration has been used.

Operative Management

Typically any consideration for an operative intervention should be delayed in our estimation for an interval as long as 1 year. It should be recognized that such strategies are applicable to severe SBS independent of its etiology, but typically an operative strategy should be applied to intractable SBS before severe or irreversible liver dysfunction occurs as a complication of either SBS or its management. Table 2 summarizes the approaches that are available.

Table 2
Operative Management Options for Established SBS

<i>Slow Transit</i>	<i>Improve Peristaltic Function</i>	<i>Enhance Mucosal Absorption</i>
Close Enterostomies	Strictureplasty	Bianchi procedure
Intestinal valves	Tapering Enteroplasty	Iowa procedure
Reverse bowel segments	Intestinal plication	STEP procedure
Colon interposition		Intestinal transplantation

Options include those designed to slow intestinal transit, those whose outcome may improve effective peristaltic activity, and those that will either increase absolute mucosal absorptive area or absorptive function.

SBS, short bowel syndrome; STEP, serial transverse enteroplasty.

SLOWING INTESTINAL INTRALUMINAL NUTRIENT TRANSIT

Enterostomy or stomal closure with restoration of oral to anal continuity that includes interposition of the anal sphincter is perhaps the single most important first step in intestinal rehabilitation where a stoma has been created. Unfortunately, this maneuver has not been studied in a prospective fashion; however, there are multiple anecdotal examples that emphasize the importance of this step. This is particularly important if most if not all of the colon is intact, and certainly may be particularly important if distal ileum and even ileocecal valve can be interposed back into the fecal stream. Though there is little data to suggest that the ileocecal valve impacts patient survival, there is a suggestion that the course of parenteral nutrition support may be prolonged in the absence of this valve (43). However, even in the absence of a large portion of colon, there is real value in continuity restoration if the main feature is interposing the anal sphincter mechanism alone.

The technique of stoma takedown and closure follows standard operative techniques typically using single-layer interrupted suture technique. Timing of such an intervention is difficult to gauge. It is best to preoperatively exclude distal bowel obstruction with a contrast enema, and intraoperatively it is best to also assure a patent downstream intestine by injection of intraluminal saline through the anastomotic site. Finally, meticulous attention to perianal skin care can prevent breakdown and discomfort related to a higher frequency and volume of anticipated stool output.

Intestinal valves have been anecdotally trialed in SBS. Their putative value may be several-fold: increase intestinal transit time; minimize or prevent colon to small bowel regurgitation of bacteria and other colonic content; and to produce dilatation of the more proximal bowel, augmenting its potential for an eventual lengthening procedure. These typically have been constructed by intussuscepting proximal into distal small bowel, typically for a 2-cm segment. There are several adjunctive techniques designed to help hold the valve and prevent its deintussusception: the serosa may be denuded to create an inflammatory bond, which may help “secure” the valve; or a prosthetic patch may be used to encircle the bowel prior to the intussusception. The overlaid bowel intussusceptum is then sutured to this patch to facilitate holding the valve in place. Such valves were first trialed in 1970 (44), but their more recent application has been as a part of a sequential treatment strategy applied as a bowel-dilating mechanism (45) to prepare the patient’s bowel for an eventual lengthening procedure.

The application of antiperistaltic or reversed small bowel segments to the management of SBS is also based on the premise of slowing intestinal transit or prolonging nutrient exposure to the small bowel absorptive surface. The technique of this procedure is poorly described: a mid to distal small bowel segment, 3–5 cm in length is divided from its proximal and distal *in situ* connection, carrying the dissection carefully to the root of the small bowel mesentery to facilitate a gentle torque or twist of the segment on its mesentery. The “reversed” bowel is then connected by two end-to-end anastomoses such that the segment is now interposed in an antiperistaltic direction. The danger of this procedure is using a segment length that is too long, which might convert the patient from too fast a transit to complete SBO. First described in 1959 (46), there have been several small pediatric experiences reported (47,48), the largest being a five patient series where original bowel length varied from 20–94 cm and the reversed segment was 3 cm in length. Four of five of these patients survived TPN-free.

Small bowel colon interposition was first described in 1971 (49,50). The technique is especially applicable to the child with short small bowel who has an intact colon. Whether the colon segment is inserted prejejunal or preileal or whether it is inserted in an isoperistaltic or antiperistaltic direction does not seem to influence its salutary benefit, which is to slow transit time, enhance vitamin B12, carbohydrate, and fat absorption and augment weight gain presumably secondary to a more prolonged exposure of nutrients to the absorbing small bowel mucosa. This operation is done by isolating an 8- to 24-cm segment of either the ascending, transverse, or descending colon on its respective blood supply and interposing the colon between two ends of small bowel with end-to-end anastomoses. One recognized outcome of this procedure likely secondary to bacterial overgrowth is the development of D-lactic acidosis, a unique complication that requires aggressive treatment. In a series of patients reported from two institutions where patients had residual small bowel lengths of 15–63 cm, four of seven patients were long-term survivors (51,52).

IMPROVING PERISTALTIC FUNCTION

The singular pathophysiologic mechanism that is most challenging to manage in SBS is bacterial overgrowth and its associated malabsorption and low-grade sepsis. Mechanisms that contribute to this sequence of events include intraluminal nutrients and bacterial stasis secondary to partial obstruction, disparate anastomotic size, or ectatic and dysmotile segments of intestine that preclude apposition of the intestinal wall during peristalsis. In the face of intestinal narrowing and secondary partial obstruction, stricturoplasty may be used to relieve the obstruction yet preserve intestinal length. Disparate sized bowel secondary to partial obstruction, an eccentric anastomosis, or bowel ectasia can be managed by several techniques. Where small bowel length is not an issue, resection of the dilated segment followed by a new end-to-end anastomosis is the best solution. An alternate technique would be a tapering enteroplasty, in which the dilated segment is narrowed or tapered along its antemesenteric border, most effectively done by using an automated stapling device (53). Where length is an issue, either imbrication or plication of the ante-mesenteric border accomplishes the needed narrowing without sacrificing mucosal surface area (54). The main issue with the latter technique is that the plicated bowel may eventually become “undone.” In a series of 11 children subjected to tapering enteroplasty segments that varied in site and length, nine patients were completely weaned from TPN and the other two had considerable reduction in their parenteral support (55).

INCREASE MUCOSAL SURFACE AREA

The tools in use today are the intestinal lengthening procedures that likely constrain absolute surface area. Instead, these techniques do lengthen bowel, make luminal size smaller such that the walls coapt during peristalsis, and the altered intestinal motility may enhance the exposure of intraluminal nutrient to functional mucosa.

The Bianchi procedure (also called the Autologous Gastrointestinal Reconstruction [AGIR]) is a technique that divides the intestine in a longitudinal plane separating the leaves of mesentery into two equal sides (56). The two parallel tubes of neointestine, representing half of the original diameter must then be connected in a properistaltic fashion by end-to-end anastomoses in a gentle “S” configuration. Kimura described a modification of a bowel lengthening procedure, the Iowa procedure, in which shortened intestine was sutured to either the undersurface of the liver or the undersurface of the

rectus abdominus muscle, from which a parasitized blood supply was generated (57,58). Once neovascularity was established, the bowel could be divided in a horizontal plane to again double its length. If the Iowa procedure followed the Bianchi procedure, then there could be a potential bowel length increase by a factor of four. Georgeson added to this procedure the sequential lengthening process, including the use of an interposed valve, to produce bowel dilatation that would facilitate a lengthening procedure (59). The most recent contribution in this field is the Serial Transverse Enteroplasty (STEP) procedure, described by Kim (60). This technically simpler and safer procedure is done typically by the serial, alternated, and parallel application of a gastrointestinal stapling device between mesenteric vessels spaced to produce a zig-zag intestinal configuration in which the neointestine is of common but considerably smaller size. The lengthening procedures are especially useful techniques in the face of focal or elongated intestinal dilatation. Data from the Iowa procedure is represented by isolated case reports. The AGIR and STEP procedure has received more emphasis and study. Bianchi has described his personal series of 20 patients, nine of whom survived long-term (61). Lengthening in the nine survivors occurred at a mean age of 23.5 months, and the achieved length increase went from a mean of 62 to 97 cm. Interestingly, in the nonsurvivors, the absolute length was less (43 to 68 cm), but strikingly, the mean age of the procedure was 2.75 months, suggesting that they represented a population of babies in trouble with their SBS, likely with developing liver dysfunction. The STEP procedure has been shown in both patients as well as in experimental animals to improve nutritional status, intestinal absorption, and to have a salutary benefit on bacterial overgrowth in SBS (62).

Intestinal transplantation would be the prototype of a SBS treatment to enhance bowel surface area (63). The spectrum of the transplant could be an isolated small bowel graft from a cadaver or related living donor, a combined cadaveric graft of liver and small bowel used to treat those children who have concomitant liver failure as a complication of their SBS, or a multivisceral graft to treat the same intestinal failure, liver failure complex. Further discussion of bowel transplantation is beyond the scope of this chapter.

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14

Interventions for Appendiceal Complications

Renata Fabia, MD and Steven Teich, MD

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INTRODUCTION

Appendicitis remains the most common cause of acute abdominal surgical disease in children (1–3). The treatment of acute appendicitis engenders very little controversy; appendectomy has been the accepted treatment of choice for over 100 years, with excellent results. However, once the appendix has perforated, the potential morbidity increases, along with controversy regarding its treatment. Children, especially those under 8 years of age are more likely to present with perforated appendicitis than adults (4). Despite the commonality of ruptured appendicitis in children, there is no treatment consensus.

This chapter discusses the current diagnostic tools and therapeutic approaches to complications of appendicitis in children, as well as controversies in management.

RISK OF PERFORATION AND ITS MORBIDITY

Perforation of the inflamed appendix is rare within the first 24 hours of untreated symptoms but climbs to 6 % among patients not yet treated by 36 hours after symptom onset (5). After 36 hours of symptoms, the risk of rupture remains steady at approximately 5% for each ensuing 12-hour period (6). Perforation of an inflamed appendix occurs in 15–57 % of all patients treated surgically for suspected acute appendicitis

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(7,8). The highest rates are encountered in children, with some series reporting up to 65% perforation rate in their pediatric population, with the perforation rate approaching 100% in children under the age of two years (4,9–12). Major complications from ruptured appendicitis occur in 6.4–19.8% of cases (4,13–15).

DRAINS, LAVAGE, AND WOUND CLOSURE

Many surgical techniques have been used in an attempt to minimize complications following appendectomy. These included drainage and/or lavage of the peritoneal cavity and different methods of skin closure.

Recently, it has been established that drains do not reduce complications after appendectomy for any stage of appendicitis (16). Drains can potentially be harmful if they become a conduit for invasion of the wound by skin pathogens. In the overwhelming majority of cases, a drain is not necessary. However, if the appendiceal stump or cecum is of suboptimal quality, placement of a closed drain may be prudent (17). It should be brought out through a separate small incision in the right lower quadrant and laid along the cecum into the pelvis to drain those dependent areas. Considering that drains pose a risk and accomplish little, they should rarely be used. When used they should be removed as soon as possible. Prophylactic antibiotics should not be prolonged to “cover” indwelling drains (18).

Peritoneal lavage versus drainage for perforated appendicitis in children has been investigated. Peritoneal lavage appears to be superior to intraperitoneal tube drainage for the management of perforated appendicitis in children (19,20). The purpose of irrigation is to remove all debris, purulent fluid collections, and blood from the surgical field. In early phlegmonous appendicitis without any pus, there is no advantage to irrigation but rather risks spreading contaminated fluid throughout the abdomen. Otherwise, in case of advanced appendicitis, with the appendix removed, a thorough lavage of the area should be performed. In particular, the pelvis has to be well exposed and any residual contaminated fluid should be aspirated and irrigated by retracting the sigmoid colon and exposing the pouch of Douglas.

A variety of skin closure techniques have been utilized for appendectomies in order to prevent the most common complication, wound infection. These include: leaving the skin open with packing, delayed primary closure using skin tapes, interrupted approximation of the skin with or without various aseptic wound wicks, and subcuticular wound closure with various sutures (21–24). The use of wound wicks is not associated with decreased wound infection rates following appendectomy (21). Furthermore, a large study demonstrated that primary closure of the skin and subcutaneous tissue after appendectomy for gangrenous or perforated appendicitis, combined with the use of antibiotic therapy in the perioperative period, is not associated with an increased risk of wound infection when compared with delayed skin closure (25). Subcuticular closure is therefore deemed appropriate in view of its greater convenience and safety. In addition, syringe pressure irrigation of the subcutaneous tissue after appendectomy was found to significantly decrease the incidence of postoperative wound infection in complicated cases (26).

ROLE OF ANTIBIOTICS

Empiric antibiotic therapy has been shown to significantly decrease complication rates in patients with acute as well as complicated appendicitis (27–29). Current controversies involve choice, duration, and route of antibiotic treatment. It is generally

believed that it is critical to administer antibiotics before making the surgical incision (30). However, a recent study suggested that antibiotic prophylaxis is effective in the prevention of postoperative complications in appendicitis patients, whether given pre-, peri-, or postoperatively (31). Children with perforated appendicitis can be managed effectively with multiple antibiotics or a single broad-spectrum antibiotic, such as piperacillin/tazobactam (30–34). The appropriate duration of antibiotic use following appendectomy for advanced appendicitis in children is still being debated. There is an ongoing effort to simplify the antibiotic regimen and reduce length of therapy for advanced appendicitis (35,36).

MANAGEMENT OF THE DIFFICULT APPENDICEAL STUMP

Open Appendectomy

The appendix can usually be removed by the standard technique of stump ligation after transection, with or without inversion of the stump into the base of the cecum. The inversion of the stump into the cecum may however be associated with a greater risk of bowel obstruction (37). When inflammation or necrosis extends into the cecum and the appendix cannot be ligated at its base, there are several options that can be employed. These include appendicocostomy, partial cecal excision with a stapling instrument, and ileocectomy or right colon resection (38,39). Appendicocostomy involves placing a catheter into the cecum through the appendiceal orifice at the convergence of the taeniae, and leaving it in place for 2–3 weeks. After that time the catheter can be removed; resulting in a controlled fistula that usually closes within one week if there is no residual disease in the colon (38). If inflammatory or necrotic changes of appendicitis involve the base of appendix, but there is a sufficient amount of healthy tissue between the cecal base and the ileocecal valve, a partial cecal resection with appendectomy using a stapling instrument can be performed. When faced with a difficult appendiceal stump, these techniques help minimize the risk of appendiceal stump breakdown with intraperitoneal soilage or cecal fistula (38).

When a patient presents with perforated appendicitis with a large abscess or phlegmon involving the cecum and adjacent structures, a routine appendectomy is hazardous and may lead to serious complications including fecal fistula, intestinal obstruction, and intraabdominal abscess (39). Ileocectomy, with limited resection of the terminal 5–7 cm of ileum and primary anastomosis, is a safe and effective therapy in selected patients (39). Right hemicolectomy is an alternative procedure that can be utilized in patients with excessive inflammatory involvement in the area.

Laparoscopic Appendectomy

Various techniques of appendiceal stump closure and control of the meso-appendix have been described (40–42). These include: catgut ligature (40), the Endo-loop tying technique (41), use of a linear stapler (40), use of an Endo-GIA automatic stapler (41), and an electro-coagulator technique (42). The Endo-loop technique is cheaper and requires only a 5-mm port; however, it requires more skill and may initially take more time. The stapling technique needs less skill, is safe, and initially saves time, but it is more expensive and requires a 12-mm port (41). The length of surgery and postoperative complication rate (intraoperative fecal soilage, and postoperative ileus) may be reduced by using a stapler versus the ligature technique (40).

INTRAOPERATIVE CULTURES

Intraoperative cultures obtained at the initial operation for ruptured appendicitis, once believed to be important for guiding postoperative antibiotic therapy, has been shown to be of no clinical value (43–45). Antibiotic changes based on intraoperative cultures do not affect the clinical outcome or prevent postoperative complications (43,46,47). Lack of clinical improvement does not indicate a failure of an appropriately selected initial antibiotic regimen and does not support simply changing the antibiotics (45). It highly suggests the probability of intraabdominal abscess or wound infection, which is responsive to drainage rather than antibiotic treatment (45). However, all postappendectomy abscess cavities should be cultured when they are drained, either surgically or percutaneously. In this instance, antibiotic therapy should be individualized based upon abscess culture results, because the abscess will usually grow only one to three organisms (48).

INTRA-ABDOMINAL ABSCESS

An intraabdominal abscess can occur during two phases of appendicitis: either during initial patient presentation as an aspect of ruptured appendicitis or postoperatively as a complication of advanced appendicitis. Between 2–6% of cases of acute appendicitis are complicated by development of a periappendicular mass, ranging from a phlegmon to an abscess (49). The incidence of postappendectomy intra-abdominal abscess is between 5–20% (4,13–15,50,51), but is significantly higher in the subset of patients less than 10 years of age (45).

Diagnostic Evaluation

INITIAL PRESENTATION

Patients usually present with abdominal discomfort, anorexia, change in bowel habits, fever, and other nonspecific symptoms. Typically, there is a long duration of symptoms and patients often present after 5–7 days of symptoms. On physical examination an abdominal mass may be present but abdominal wall guarding may preclude palpation unless performed under general anesthesia. Typically, fever and white cell count (WBC) are higher than in uncomplicated appendicitis (49). Abdominal computed tomography (CT) is useful to determine whether an appendiceal abscess is present, to differentiate the abscess from a phlegmon, and to determine whether the abscess is amenable to percutaneous drainage. High-resolution ultrasonography with graded compression also has very good sensitivity and specificity, but is operator-dependent, and there is a limited field of view (52,53).

FOLLOWING APPENDECTOMY

Typically, these patients do not improve following appendectomy and develop recurrent fevers, abdominal pain, anorexia, and elevated WBC. These symptoms should lead to an evaluation for the presence of a postoperative intra-abdominal abscess (45).

Initial Nonoperative Management—Percutaneous Abscess Drainage

The management of appendiceal mass has been evolving, with significant changes over the last decade. Initial nonoperative treatment of an intra-abdominal abscess by

percutaneous drainage and broad-spectrum intravenous antibiotics has been demonstrated to be safe and effective (49,53–55). The abscess drainage is usually performed by an interventional radiologist either percutaneously or transrectally using ultrasound or CT guidance (53,56) (*see radiology chapter of this book*).

This approach avoids extensive surgery with the risk of intra-operative injuries, postoperative development of fistulae or recurrent abscesses, and prolonged postoperative recovery that is typically associated with immediate surgical treatment of intra-abdominal abscesses (49,55). Approximately 80-90% of patients have been reported to benefit from this approach (49,55). The patients who do not improve within 72 hours after percutaneous drainage will require operative intervention (54–59). When initial nonsurgical treatment is successful, length of hospitalization, cost, and morbidity are decreased compared to initial surgical management (57).

Operative Management

Patients with appendiceal abscess who present with signs of diffuse peritonitis, multiple abscesses or bowel obstruction, and appear toxic, typically are not candidates for percutaneous drainage and require early surgical intervention (60). Advocates of initial surgery as the treatment of choice in all cases of appendicitis complicated by intra-abdominal abscess cite low morbidity, no need for interval appendectomy (61,62), and elimination of the risk of recurrent appendicitis (63). However, the complications of immediate surgery, when encountered, can be extremely serious. Moreover, injudicious surgical treatment of patients with periappendiceal masses has occasionally required emergency right hemicolectomy (61,64).

Early identification of the factors associated with failure of nonoperative initial management is important. It has been reported that patients requiring early appendectomy have a more frequent finding of a small bowel obstruction (SBO) on their initial X-rays and a higher percent band count on their initial differential WBC count than those successfully treated with interval appendectomy (59). An initial band count less than 15% is predictive of an uncomplicated course (59).

Role of Interval Appendectomy

Interval appendectomy (performing an elective appendectomy in the “interval” between bouts of appendicitis) remains controversial (55–62). Interval appendectomy is advocated to avoid a high recurrence rate of appendicitis and to confirm the diagnosis (58). Yet, some authors suggest that interval appendectomy is unnecessary (49,65). They point out that the recurrence rate is low and the removed appendix often shows no evidence of previous inflammation; therefore, the procedure is not justified (58,65). In other reported series, pathologic examination demonstrated that less than 10% of the appendices removed during interval appendectomy demonstrated no abnormalities (61). Furthermore, delayed or interval appendectomy specimens often have a characteristic inflammatory pattern that includes granulomas, xanthogranulomatous inflammation, mural fibrosis/thickening, and transmural chronic granulomatous inflammation (66). It would be desirable to perform interval appendectomy only for those patients in whom recurrent appendicitis is very likely. However, because a reliable and noninvasive test to accurately determine the risk does not exist, the reasonable approach is to advise interval appendectomy for all patients (67). A recent study supports the need for interval appendectomy by showing pathological findings in the majority of

excised appendices, and suggests that laparoscopy is a safe alternative to open surgery for interval appendectomy with a low complication rate (68,69).

SHORT-TERM POSTOPERATIVE COMPLICATIONS

Wound Infection

Wound infection is the most common source of morbidity following open appendectomy for appendicitis, but there has been a steady decline in its occurrence in recent years (15,70). It generally occurs 5–7 days after surgery. The incidence of wound infection is more frequent in patients with advanced appendicitis (9), and it may contribute to a prolonged hospital stay, increased cost of treatment, a longer period of discomfort and pain, and delayed return to normal activities (71). One advantage of the laparoscopic approach over open appendectomy for patients with early appendiceal perforation is reduction of the incidence of wound infections (69,72).

Incidence

With the current practice of antibiotic use, pediatric patients with acute, nonperforated appendicitis have a wound infection rate of less than 1% (70,73). Recent series of complicated appendicitis in children report wound infection rates less than 5% (15,25,70,73), yet some institutions still report infectious complication rates that are significantly higher, some over 20% (60,71,74,75). Laparoscopic surgery has been demonstrated to decrease wound complication rates even for advanced appendicitis to approximately 1% (69,72), provided operative time is less than one hour, which is accomplished in the majority of cases (18).

Diagnostic Evaluation and Treatment

Postoperative wound infections present as erythema, tenderness, edema, and occasionally purulent drainage. The wound is often soft or fluctuant at the site of the infection. The patient may have a leukocytosis and a low-grade fever. Management of this complication depends on the depth of the infection. For a superficial wound infection that involves the skin and subcutaneous tissue, the skin staples or sutures are removed over the area of the infection and a cotton-tipped applicator is passed into the wound, typically with efflux of purulent material. The wound should be gently explored and loculations broken up. Debridement of any nonviable tissue is also important. An assessment of the integrity of the fascia should be made, because wound dehiscence may occur. If the fascia is intact, wound packing with wet to dry dressing changes or wound vacuum-assisted closure is commonly instituted. However, if the fascia has separated there is obvious concern about an intra-abdominal abscess, which may require either CT-guided drainage or possible surgical intervention (9). Incision and drainage of a superficial wound infection can usually be accomplished in the outpatient setting under conscious sedation (69).

Antibiotics are generally not required for a wound infection, unless there is evidence of cellulitis, deep tissue penetration, systemic toxicity, an immune compromised patient or an indwelling prosthetic device (74). Necrotizing fasciitis is an extremely rare complication of appendicitis, and there are only a few cases documented in the literature. Once necrotizing fasciitis occurs, the mortality rate increases dramatically, so that correct diagnosis and prompt debridement are mandatory (76).

Pathogenic Organisms

Although the most common microorganisms in acute appendicitis are *Escherichia coli* and *Bacteroides fragilis*, the organisms most frequently found in postappendectomy wound infection are *Bacterioides* and *Clostridium welchii* (15,43). Necrotizing fasciitis, along with myositis of the abdominal wall is a very rare but devastating complication. Polymicrobial infection is responsible for this entity: group A *Streptococcus*, *Staphylococcus aureus*, *E. coli*, and anaerobes like *Clostridia* and *Bacteroides* (76).

Prevention

There have been multiple techniques proposed for wound prophylaxis in pediatric appendicitis (71). These include: draining and/or packing the wound, wound irrigation, antibiotic powder into the wound, and preoperative intravenous antibiotics alone. A clinical study demonstrated that patients that received preoperative (or intraoperative) intravenous antibiotic (cefoxitin) plus wound antibiotic powder (cefoxitin) had a lower infection rate than those receiving either topical or intravenous antibiotic alone (73). The benefit was significant compared to the group receiving no treatment (73). Antibiotic prophylaxis has been determined to be effective in the prevention of postoperative complications in appendectomy patients (30,31). However, adding a course of outpatient oral antibiotics, after completing a course of IV antibiotics, does not decrease postoperative infectious complications in appendicitis patients (77). Mechanical removal of infectious material, whether by peritoneal lavage or wound pressure irrigation has been shown to decrease the wound infection rate (19,20,26). Diligence with aseptic technique and avoidance of a hematoma at the incision site comprise additional factors that aid in wound infection prevention. Most recent pediatric series dealing with complicated appendicitis use protocols of preoperative antibiotics with aerobic and anaerobic coverage, intraoperative lavage, no peritoneal or wound drains, and continuation of antibiotics postoperatively (69).

Postappendectomy Abscess

Lack of clinical improvement after appendectomy with continuing fevers, leukocytosis, ileus, or signs of sepsis indicate a high probability of an intra-abdominal abscess (45). Fevers in combination with abdominal pain are the most common sign associated with this finding (45,78). The only independent risk factors for development of postappendectomy intra-abdominal abscess are appendiceal perforation and age less than 10 years (78). Drain placement during appendectomy was not found to make a significant difference in intra-abdominal abscess formation (78). Studies comparing intra-abdominal abscess rates following laparoscopic and open appendectomy have shown conflicting results. Early reports demonstrated an increased rate of abscess formation after laparoscopic appendectomy (79,80). However, more recent studies demonstrate no statistically significant difference in the rate of postappendectomy abscess among children following laparoscopic and open procedures (81).

Diagnostic Evaluation and Treatment

The diagnosis of postappendectomy abscess can be made by clinical findings alone, such as in the case of spontaneous drainage of a subfascial purulent collection. It can also be made by abdominal CT scan or ultrasonography. Abdominal CT diagnosis

is made by demonstrating a nonenhancing intra-abdominal fluid collection in patients with clinical signs of abdominal sepsis (78).

Drainage of the purulent material is usually required in order to treat this complication. Cultures of the drained abscesses should be obtained, as the pathogens are usually different from those found during appendectomy and may be resistant to the current antibiotic regimen (45). Drainage procedures may be performed by a percutaneous method using CT or ultrasound guidance or by an open surgical approach. The success rate of both techniques is similar and the hospital stay is the same, regardless of which drainage method is utilized (78).

Suppurative Pylephlebitis

Pylephlebitis or portal pyemia, also known as septic thrombophlebitis of the portal vein, or occasionally the superior mesenteric vein (SMV), was common in the preantibiotic era. Today, it is encountered extremely rarely in association with intra-abdominal infections in the region drained by the portal venous system, including diverticular disease and complicated appendicitis (82). Although extremely rare, it is associated with a high mortality, even in the modern era (83). It is characterized by jaundice, chills, and high fever. This serious illness frequently leads to multiple liver abscesses and the infecting organism is usually *E. coli*. Prompt diagnosis and treatment are necessary to prevent hepatic abscess, septicemia, and complete thrombosis of the portal circulation (82,84).

Diagnostic Evaluation and Operative Management

Diagnosis of pylephlebitis is accomplished by analysis of symptoms, clinical examination, and imaging studies including abdominal CT. Surgical therapy consists of appendectomy but occasionally a right hemicolectomy is required (83,85). Liver abscesses must be drained and removal of the thrombus can be performed by using a Fogarty catheter. Postoperative histopathological examination is performed to confirm the diagnoses of appendicitis and septic thrombophlebitis of the portal vein and/or SMV. The patient is then treated with long-term antibiotics and anticoagulation. Complete recovery with appropriate medical and surgical treatment can be achieved (83,85).

Postoperative SBO

Intestinal obstruction is considered early when it develops within three weeks after the operation. Intestinal dysfunction is the second most common short-term complication after appendectomy. In the first few weeks postoperatively, it is usually caused by a combination of paralytic ileus secondary to peritonitis and mechanical obstruction from fibrinous adhesions. A small bowel intussusception may also cause an early intestinal obstruction postoperatively, with an incidence of 0.06–0.8% (86). The clinical picture is one of a patient who initially manifests a return of gut function and advances to a diet, but then has loss of bowel function along with abdominal distension and pain.

DIAGNOSTIC EVALUATION

Plain abdominal films, when performed serially, are sometimes useful in the assessment of the postoperative patient with a suspected SBO. Abdominal CT is the best initial imaging technique in a patient with a history of a laparotomy who complains

of mild intermittent abdominal pain but has few physical findings and nonspecific plain radiographs. CT enteroclysis should be used after the conventional abdominal CT study if additional management questions are left unanswered (87). Small bowel intussusception can be identified by abdominal CT or ultrasound, but is rarely diagnosed before surgery (86).

OPERATIVE VERSUS NONOPERATIVE MANAGEMENT

Postsurgical patients presenting early after operation with abdominal distension and no signs of bowel compromise (tachycardia, leukocytosis, localized tenderness, or fever) can be treated nonoperatively for several days, with subsequent abdominal CT if the clinical findings and abdominal plain films do not improve. Most early “obstructions” resolve with bowel rest, nasogastric suctioning, and intravenous fluids. If the patient is unable to eat for more than 5–7 days, parenteral nutrition is indicated (9,86). Any postoperative patient at high risk (i.e., very young age or comorbidities present) that demonstrates a clinical picture of secondary loss of bowel function, distension, and abdominal pain should be presumed to have a mechanical SBO, and early operation should be considered (87,88). Patients with fever, tachycardia, leukocytosis, or other signs of bowel compromise should undergo immediate surgical exploration.

Stump Appendicitis

When performing an appendectomy, once the appendix has been completely skeletonized, it is amputated at the base. In general, there is no need for inversion of the appendiceal stump, but it is important to divide the appendix at the level of healthy tissue, even if necessary at the level of the cecum, to avoid breakdown of the suture or staple line. It has been shown that stump inversion at the time of open surgery has no effect on reducing the rate of intraabdominal abscess formation or length of hospital stay compared with simple stump ligation. Conflicting evidence exists, however, whether there is a difference in the rate of wound infection. One randomized clinical trial found no difference whereas another study found a higher rate of infection following stump inversion (89,90).

Residual appendiceal tissue left after an initial appendectomy risks the development of stump appendicitis, which is a real, yet underreported entity. The latest comprehensive review of the English-language literature revealed 36 reported cases of stump appendicitis (91). Typically, patients present with signs and symptoms similar to acute appendicitis; however, because of prior surgery, the diagnosis is difficult and the rate of appendiceal stump perforation is extremely high (91). There is no correlation between simple ligation or inversion of the stump and stump appendicitis (92).

Stump appendicitis can occur after either laparoscopic or open appendectomy. Rare cases of inflammation of the appendiceal stump after open appendectomy have been reported (93,94). However, it has been suggested that stump appendicitis may increase in prevalence with more frequent performance of laparoscopic appendectomy (92). A small field of view, lack of three-dimensional perspective, and absence of tactile feedback are factors that may lead to an increased likelihood of a significant appendiceal stump left after this approach (95). Its incidence may be minimized with accurate visualization of the appendiceal base and creation of an appendiceal stump less than 3 mm in length. It has been postulated that if laparoscopic appendectomy is performed properly, there should not be a sudden increase in the incidence of this entity (92,95).

Colonoscopy, ultrasonography, and abdominal CT have been shown to be useful in the diagnosis of stump appendicitis. Abdominal CT can assist in making an accurate preoperative diagnosis. It may show a characteristic “arrowhead sign” and a retained appendicolith, both individual specific CT signs of appendicitis that, when combined, are pathognomonic for stump appendicitis (96). Although rare, stump appendicitis should be considered in the differential diagnosis of right lower quadrant abdominal pain postappendectomy. Minimizing the length of the appendiceal stump is the only means by which to avoid its occurrence.

LONG-TERM COMPLICATION

SBO

There is a 5% incidence of postoperative adhesive obstruction in children after laparotomy for any reason (15,97). Of these, 80% have been reported to occur within 2 years of the operation (97). Intestinal obstruction can occur after laparotomy for appendicitis; the long-term incidence is likely similar to the risk of patients undergoing laparotomy for other reasons and its frequency varies from center to other (9,98). One study analyzing the risk of surgically treated SBO after open appendectomy showed the accumulated risk is 0.41% after 4 weeks, 0.63% after 1 year, and 1.3% after 30 years of follow-up; compared with 0.003% at 1 year and 0.21% after 30 years of follow-up among nonoperated controls. The highest risk was found after operation for other diagnoses, followed by operation for perforated appendicitis, nonspecific abdominal pain, and mesenteric lymphadenitis compared with operation for nonperforated appendicitis. Women had a slightly lower risk than men (99). The incidence in another two series was low, approximately 1–1.3 %, with most patients presenting in the first 6 months after operation (9,98). In children that were followed 4–6 years after appendectomy, 1.3% developed clinical symptoms consistent with mechanical SBO resulting in relaparotomy and confirmation of the diagnosis. Regarding the relationship between severity of the disease and development of mechanical bowel obstruction, the incidence was 1.8% in the group with a normal appendix, 0% after simple appendicitis, 0.4% after gangrenous appendicitis, and 3.4% after perforated appendicitis (100). On the other hand, the magnitude of readmission for abdominal complaints following appendectomy was found to be more pronounced, and occurred in 2.94% of patients during a 10-year follow-up after appendectomy (101). Nonspecific abdominal pain with no sign of SBO caused almost one-half of the readmissions (45%), mostly in females (76%). Patients with a history of complicated appendicitis or a “normal” appendix were most frequently readmitted (101). Approximately one-half of the patients with symptoms of bowel obstruction required surgical intervention (101). The main cause of bowel obstruction after open appendectomy is adhesions. Incisional hernias also occur in 0.4% of all appendectomies (101).

Intestinal obstruction can occur following all types of laparoscopic abdominal surgery; in the case of appendectomy, the prevalence is as high as that seen in open procedures (102). Specific reasons include retained loose linear cutter staples (103,104), residual appendiceal tissue (105), and even rarely cecal volvulus (106). Retrieving as many loose staples as possible at the termination of the laparoscopic appendectomy is therefore recommended (104).

Fertility

Although perforation of the appendix is considered a risk factor for female tubal infertility, the epidemiologic evidence supporting this relationship is inconsistent. The idea dates back to 1932, when a medical report stated: “It is generally agreed, that acute appendicitis may bring about sterility in women” (107). Subsequent studies, however, have generated conflicting results (108–113). It has been commonly thought that a perforated appendix in females may result in tubal dysfunction caused by peritoneal adhesions after inflammation, and a subsequent increased risk for extrauterine pregnancy and infertility. It has been suggested that early diagnosis and treatment of suspected appendicitis in girls and women of reproductive age may reduce the incidence of tubal infertility resulting from the sequelae of a ruptured appendix (109). The liberal use of laparoscopy in women with suspected appendicitis has also been recommended because of a fear of tubal dysfunction secondary to postsurgical peritoneal adhesions after conventional open appendectomy (114). However, some authors believe that it is unlikely that appendicitis with perforation will cause infertility unless there is an abscess in the pouch of Douglas (108). In more recent studies, neither history of acute appendicitis before puberty nor perforation of the appendix was a statistically significant risk factor for tubal infertility (110). Others studies agree that perforated appendicitis has little, if any role in the etiology of tubal infertility (111,112). This may have important implications for the management of young women with suspected appendicitis, as the liberal attitude toward surgical exploration with a subsequently high rate of negative appendectomy is often justified by a perceived increased risk of infertility after perforation. Women whose appendix was found to be normal at appendectomy in childhood seem to belong to a subgroup of patients with a higher fertility rate than the general population (112).

A causal relationship between appendicitis and infertility cannot be supported by the data currently available. Only a well-designed, prospective clinical study with unbiased ascertainment of exposure and adjustment for confounding variables will provide a definitive answer (115).

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15

Reoperation for Inflammatory Bowel Disease

Christopher R. Moir, MD

CONTENTS

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CROHN'S DISEASE—RECURRENT DISEASE

A child diagnosed with Crohn's disease has a 78% probability of surgery within 20 years (1). The risk is higher for ileocolic disease (92%), whereas approximately two-thirds of patients with small bowel involvement and slightly less than 60% of patients with colonic disease will come to operation (2). Pediatric surgeons planning the first operation do so knowing 50% of their patients will return for another procedure (1,3). This section reviews patterns of Crohn's disease recurrence and operative strategy.

Strictures

CLINICAL PRESENTATION

Recurrent Crohn's disease is found most commonly at the surgical anastomosis (4). The degree of obstruction from recurrent inflammation and scarring establishes the intensity of symptoms. Crampy abdominal pain exacerbated by meals is a common clinical presentation. Onset of recurrence varies with site and extent of initial resection. Much attention has been given to the adequacy of the first surgery; however, recurrence at the anastomosis is a feature of the disease process rather than the presence of microscopic residual disease at the margins (5). Grossly negative resection margins are all that is necessary for a good initial procedure (6). Symptom onset can be expected to occur earlier for patients with diffuse intestinal disease, perianal Crohn's disease, and those who had postoperative surgical complications (7). Although some patients have unfortunately experienced recurrence of symptoms within months of surgery, the average time of symptom recurrence is site-dependent and not expected for several years after operation. At least 85% of surgical patients have histologic evidence

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of recurrence by 3 years after surgery. Such biopsy evidence does not necessarily mean children will go on to develop endoscopic findings of ulceration and strictures. Similarly, endoscopic recurrence does not correlate with development of symptoms or risk of subsequent reoperative surgery. The best rule of thumb when discussing disease recurrence with children and their families is to assume reoperation rates of just higher than 50% for ileal colic disease and 45% each for isolated colonic or small bowel Crohn's disease by 10 years (1,3,8–10).

Perforating Crohn's disease represents a high-risk group thought to fare poorly after intestinal resection. This is only true for the mode of recurrence, which tends to favor perforation once again. The absolute risk of recurrence is similar to obstructing Crohn's disease (11,12). Older adolescents who smoke represent a high-risk group and should be encouraged to quit.

The degree of obstruction dictates the patient's symptoms and further treatment. Medical intervention for control of symptoms is based on progression to the more concerning findings of partial bowel obstruction, bacterial overgrowth, malabsorption, and pain. Such symptoms can make meal time an unpleasant experience, producing a "feeding aversion" that occurs when the patient knows oral intake will produce further pain. Obstructive symptoms are particularly distressing for adolescents and young adults who often eat at irregular intervals associated with their lifestyle and travel. It is not possible to have a good quality of life when oral intake must be regulated throughout the day.

Surgical indications for reoperation are based on the intensity of symptoms and interference with quality of life (13). The amount of obstruction and the medical treatment necessary for its control may lead to determination of treatment failure and surgical intervention (1,3). Perforating Crohn's disease represents a fortunately rare but dramatic subcategory for reoperative indications (11).

DIAGNOSTIC EVALUATION

Patients with Crohn's disease and recurrent symptoms can make the diagnosis themselves. The role of the physician and surgeon is to confirm disease recurrence and assess severity utilizing disease activity indices, radiologic signs and endoscopic findings (9,10). Physical examination may identify abdominal distention or a mass, but most often diagnostic imaging is necessary to quantify the extent of disease and its effects. Although recurrence is usually limited to the previous site of surgery, it is essential to identify other areas of disease that may have occurred postoperatively (14). Standard imaging includes contrast studies and computed tomography (CT) enterography for visualization of luminal disease and quantification of the extraluminal effects (15–17). Despite radiologic advances, endoscopy remains the final arbiter of disease recurrence (13,18). Occasionally, magnetic resonance imaging (MRI) is used to identify disease recurrence, penetration or fistulization to bowel and retroperitoneal structures (19). Similarly, capsule endoscopy improves sensitivity in patients with equivocal contrast studies. Research has identified patient benefit with earlier detection of disease by capsule endoscopy (20–22).

The essential but invasive nature of endoscopy has led to renewed calls for laboratory markers of disease. Measurement of C-reactive protein may lead to earlier discovery of disease and help predict response to relapse therapy. It is more useful for Crohn's disease than ulcerative colitis and superior to erythrocyte sedimentation rate (ESR),

albumin, and complete blood count (CBC) (23). New more specific biologic tests will soon be available (24).

Laboratory investigations are always helpful to establish patient readiness for reoperation. Routine precautions include an evaluation of liver and pancreatic function in addition to routine hematology and electrolyte evaluation.

OPERATIVE TECHNIQUES

Conduct at the second operation is highly dependent on the first. Incision placement, extent of resection, and type of anastomosis all depend on the first surgeon's decisions. In general, it is easier to reopen a midline incision than to continue operating through a transverse scar. Pediatric surgeons favor a transverse approach in smaller children because of the wide access to intraperitoneal structures. As a child grows, the abdomen elongates and narrows, making a midline incision preferable for reoperation. This dilemma is particularly acute when considering a child with a transverse scar that crosses the midline. A new incision that bisects the old scar may increase the risk of delayed wound healing, incisional hernia, and unsatisfactory cosmesis. When possible, it is safe to reoperate through a transverse incision at least one more time. Access to the intraperitoneal structures is usually quite adequate and disease recurrence is generally close by. There may be an opportunity to repair any incisional herniation or subclinical dehiscence of the rectus musculature following the first operation. If necessary for adequate exposure in the older child, a new midline incision is generally well tolerated.

Laparoscopic-assisted surgery obviates issues of incision placement (25,26). Repeat laparoscopy is also feasible. The degree of recurrence, presence of intraperitoneal adhesions, and persistence of the surgeon will determine the success of the second laparoscopic dissection. Laparoscopic-assisted procedures allow for extraction of the bowel through an incision for anastomosis or stricturoplasties. In one small series, laparoscopy produced longer recurrence-free intervals (27). Although such data is an interesting side light to the laparoscopic story, it is also possible that concomitant advances in medical management have decreased recurrence. Additionally, laparoscopy is associated with a higher use of stapled anastomosis, which also reduces the frequency of reoperation.

An important principle of Crohn's disease surgery is to create a widely patent anastomosis. Patients with ileocolic resections are natural candidates for a stapled side-to-side anastomosis that can be twice as large as an end-to-end procedure. Studies of stapled versus handsewn anastomoses have fairly consistently shown improved recurrence-free intervals (28,29). With this information, a similar procedure is now recommended for small bowel surgery. Although it is particularly important to create a large anastomosis at the time of reoperation, the same technique is advocated for initial surgery. Such widely patent anastomoses may be expected to reflux colonic contents into the small bowel. This is of uncertain influence in developing children. Caution should be exercised in smaller children where issues of growth and development are paramount.

Stricturoplasties for active disease as well as burned-out fibrotic strictures remain an excellent option for patients with multifocal disease (30). Such length-sparing procedures are particularly attractive for developing children with extensive disease and skip areas. Limited resection or bypass with stricturoplasty has been employed with excellent success (31,32). There are no data as yet to support the relative contraindication of leaving persistent disease behind in developing children (33,34). Adverse effects on

growth and the risk of malignancy have not been quantified. Fortunately, disease in the area of strictureplasties has been ameliorated by the surgical technique (35,36).

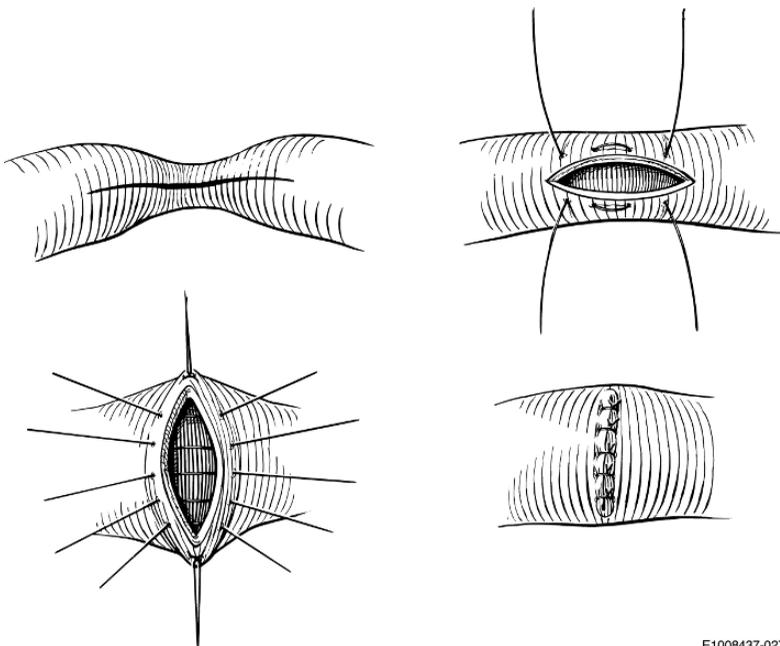
The indications for operation are no longer restricted to the multiply recurrent patient with widespread disease. Any focal lesion without diffuse extraintestinal induration is a candidate for limited intervention. Length of stricture and the noninflammatory nature of the obstruction are the two strongest factors determining operability (30).

The techniques of stricturereplasty vary according to the lesion. The standard technique known to all surgeons resembles a Heineke-Mikulicz pyloroplasty (Fig.1). The procedure is best suited for short fibrotic strictures less than 10 cm without surrounding inflammation adjacent to pliable bowel (37).

The side-to-side Finney stricturereplasty may treat up to 25 cm of diseased bowel (38) (Fig. 2). Other variations include a Jaboulay side-to-side anastomosis that bypasses the disease. Such procedures mimic old-time surgeries for Crohn’s disease, which have again come into fashion (39). Overall, 85% of strictureplasties resemble a standard pyloroplasty; however, a metaanalysis suggests the Finney procedure has a lower recurrence rate (40). In general, the reoperative rate for stricturereplasty approaches 25 and 30% at 5 and 10 years, respectively (41).

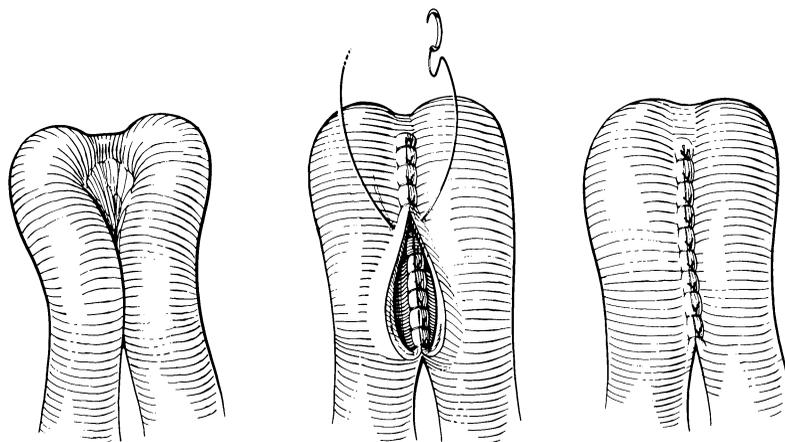
The long-term concerns of persistent disease must be balanced by the tendency for Crohn’s disease to regress over time. Disease recurrence or persistence does not necessarily correlate with symptoms, and therefore, such procedures can be advocated for children as well as adults. Its effectiveness is well established, but increased morbidity and recurrence rates suggest that selective use of the procedure is the best approach (30). Isolated short resections remain the best option for most children with Crohn’s disease.

Options for recurrence of colonic disease are more limited. Patients with obstructing disease at the ileorectal anastomosis may be amenable to local resection or



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Fig. 1. The Heineke-Mikulicz stricturereplasty. A longitudinal incision is closed transversely.



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Fig. 2. The Finney strictureplasty. The bowel is laid side-to-side and anastomosed with handsewn or stapled techniques.

strictureplasty. If this is not possible, the complications of anorectal stricturing, perianal disease, and fistulization to surrounding organs necessitate diversion. Such diversion may be permanent or at least long-term until rectal disease activity abates. Simple diversion with a loop ileostomy is a common approach to children, with the expectation that reanastomosis is performed within 1 year. Unfortunately, many children have chronic persistent disease that does not respond well to therapy. Although it is difficult to permanently remove the rectum in a child or young adult, especially when newer therapies are on the horizon, significant ongoing perianal sepsis mandates such a procedure (42).

Removal of the residual rectal stump via an abdominoperineal approach is reserved for patients with the most resistant disease (43). Local wound care following rectal stump removal continues to be problematic (44). With this in mind and the presence of newer biologic therapy, most surgeons prefer to preserve at least a portion of the rectum for later surgical reconstruction. In an attempt to obviate the complications of reoperative surgery, all efforts should be made to avoid dissection below the peritoneal reflection at the time of disconnection of ileorectal anastomoses or removal of fistulizing disease (45).

Construction of ileostomies in patients expected to keep their stomas for many years requires a more conservative surgical approach. An end ileostomy is less likely to prolapse and should be positioned in an area of the body where it is least expected to interfere with daily function. Loop ileostomies may also be used for long-term diversion; however, it is preferable to divide the bowel completely, leaving the distal limb close by. Placement of an ileostomy through the rectus sheath is mandatory for long-term control of prolapse and parastomal herniation.

NONOPERATIVE MANAGEMENT

Although 80% of patients with Crohn's disease recur following surgery, less than 50% come to reoperation (1). Most children are managed nonoperatively with the reintroduction of immunosuppression (46). The degree and severity of symptoms and intensity of therapy will determine the decision to reoperate. Fortunately, use of infliximab (Remicade) combined with immunosuppression has helped control

symptoms without operation (47). Milder recurrence has a site-dependent response to sulfasalazine or budesonide. Oral steroids are reserved for treatment failures (48). Alterations in dietary management, control of bacterial overgrowth, and nutritional supplementation may be necessary for long-term symptom control.

LONG-TERM ISSUES

Many of the ongoing problems with recurrent Crohn's disease have been addressed in the previous sections. These include control of symptoms by medication once recurrence has occurred and careful operative evaluation when surgery is necessary. Incision placement, reduction of postoperative adhesions, anastomotic technique, and stoma creation are all done with reoperation in mind. Laparoscopy remains possible with multiply recurrent disease. When an open procedure is planned, a midline incision is preferred. Gentle handling of tissues, peritoneal lavage, and placement of an anti-adhesion barrier are effective ways to decrease postoperative scarring. Evidence suggests a wide stapled anastomosis has the best outcome. Long-term stomas work best placed through the rectus musculature in a site chosen on consultation with stoma therapy.

Delayed onset of puberty is particularly problematic in children with Crohn's disease (49). Patients with unremitting chronic disease or those with frequent relapses are at highest risk (50). Causative factors include the effects of circulating inflammatory mediators, medications used, and undernutrition (51). The relative contribution of these factors affects surgical planning. For example, strictureplasty may not be the best option in a patient with very active disease. On the other hand, poor nutrition from multiple resections will continue the delay in puberty.

Subfertility was thought to be a complication of underlying disease. Recently, studies have shown a stronger correlation to patient behavior and surgical sequelae rather than the disease process itself (52). Perianal disease, dyspareunia, and the risk of incontinence may reduce the desire for intercourse (53). Tubal factors are also extremely important (54). There is evidence that fertility may be drastically reduced by 80–90% after ileoanal procedures (55,56). The most important factors for the risk of tubal infertility are location of surgery and pelvic sepsis. Patients with polyposis or those undergoing appendectomy for uncomplicated appendicitis do not share the same tubal infertility risks as patients undergoing deep pelvic procedures for Crohn's disease or ulcerative colitis (57,58). Although the ileoanal procedure and ileorectal anastomosis for Crohn's disease remain excellent operations, care should be taken to reperitonealize the pelvis and protect the tubes from damage.

Anal Fistulas

CLINICAL PRESENTATION

Approximately two-thirds of patients with Crohn's disease will have perianal symptoms at some point during their illness (59). Three-quarters of these patients will develop disease within 10 years of intestinal symptom onset. In approximately one-quarter of patients, perianal disease occurs prior to onset of small or large bowel disease (60). Symptoms are most pronounced in children whose disease is concentrated in the anorectum. These children form a distinct genetic subtype identified on chromosome 5 (61,62).

Curative surgery for perianal disease usually fails or heals poorly. Some patients go on to fecal diversion in an effort to speed healing. The pattern of poor response

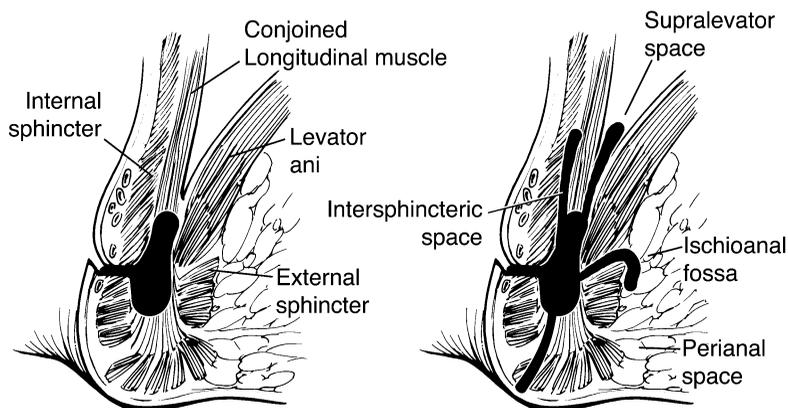
to surgery alerts the surgical team to the possibility of Crohn's disease. Fortunately, treatment with infliximab reduces local induration enough to allow for more definitive surgical treatment (63). Symptoms may be cured with medication alone or allow the patient to carry on without resorting to an ileostomy (46). It is still dangerous to perform local procedures for perianal disease, but if the child has an established response to therapy, more curative surgery can be considered.

The most common conditions the surgeon encounters are fistula-in-ano, anal fissures, and hypertrophy of the perianal skin and anoderm (64). Often seen together, they combine to produce exceptionally painful lesions that drain, bleed, and recurrently abscess. The degree of pain can be almost unbearable, particularly when anal sphincter spasm complicates the presentation. Defecation is an excruciating experience. The intensity of the local symptoms leads to stool holding and "feeding aversion," where patients voluntarily limit oral intake to avoid stooling. Older children are unable to sit in class or participate in sports. School absenteeism is high. This is a very embarrassing complication of Crohn's disease that is difficult to discuss with friends or family. Ongoing surveillance of symptoms is necessary for the early identification of surgical conditions. Perianal sepsis may still progress to severe regional infection that could produce progressive gangrene. Such complications are rare, as most emergency interventions are done in a timely fashion to drain the sepsis (46,65,66).

DIAGNOSTIC EVALUATION

The harbinger of most fistulas is an abscess. Nearly 70% of patients with ischioanal abscesses will go on to develop a fistula (67). In general, the more diseased the rectum, the more likely a fistula will develop and respond poorly to attempts at cure. Fortunately, these difficult situations are uncommon in children with Crohn's disease. Although one-third of patients will develop fistulas, most are intraabdominal. In a population-based study of patients with Crohn's disease, of the 35% found to have fistulas, only 20% had perianal disease (65).

Several types of perianal Crohn's disease may coexist at one time. Physical examination confirms the findings of fistulization, fissuring, or hypertrophy. Crohn's fistulas tend to be complex (Fig. 3). Examination under anesthesia is a kinder option for the patient and the examining physician. The opportunity for endoscopy of the distal



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Fig. 3. Possible location of Crohn's disease fistulas and abscesses.

rectum, biopsy, and identification of the internal openings of the fistula are enhanced with the patient under sedation or anesthesia. The evaluation may also allow for redrainage of small abscesses or surgical debridement of chronic cavities.

Should there be a question of more extensive fistulization, MRI has been helpful. It is a highly sensitive (97%) and specific (100%) tool for detection of fistulas and quantification of surgical success (68). MRI fistulography is superior to endoanal ultrasound but is more expensive and lacks the advantage of portability. The ultrasound probe may be utilized in the examination room, but the procedure can be quite painful. It is most useful as part of the examination under anesthesia (37,69).

Because perianal fistulas may develop in advance of intestinal disease, a thorough history and physical as well as evaluation of the upper and lower gastrointestinal tracts are suggested for all patients with persistent perianal abscesses and fistulas. Concomitant vaginal disease is often missed by regular evaluation. Examination under anesthesia, particularly in a child, is the most sensitive evaluation. In teenage females with Crohn's disease, "Bartholin's cysts" should be diagnosed cautiously. Most vaginal or vulvar masses are the result of perianal fistulization rather than a separate disease process.

OPERATIVE TECHNIQUES

Incision and drainage of perianal abscesses may be life-saving and certainly provides symptom relief. Debridement and unroofing of chronic abscess cavities filled with granulation tissue improves patient well-being and reduces the vexing issue of persistent seropurulent drainage. These two surgical interventions form the mainstay of treatment for perianal Crohn's disease. It is a characteristic of Crohn's disease that definitive curative surgical intervention fails (70). Typically, wounds close slowly or not at all while repaired fistulas recrudescence months to years after surgery. For this reason, the operative interventions for Crohn's may be divided into three categories; the first is described previously as emergency surgical treatment for septic conditions. The second represents ongoing control of symptoms through surgical debridement or placement of a drainage seton suture. These two categories of surgery are sufficient to provide control and healing for most perianal conditions. If not, fecal diversion is reserved for severe unremitting or life-threatening symptoms. The third and more definitive category of surgery involves curative repair. Several procedures, including the traditional fistulotomy and advancement flaps have been tried with success.

Fistulotomy is the definitive cure for persistent anal fistula. Healing occurs in 73–81% of patients with low fistulas laid open in the traditional fashion (64). The risk of incontinence and poor wound healing is high in children with a transsphincteric or complex fistula. These children and those with active rectal disease should not undergo fistulotomy (71). Alternatives to definitive surgical therapy include infliximab and fecal diversion (46). Studies of patients with complex fistulas, including rectovaginal and pouch fistulas, have shown infliximab to produce healing in at least one-third of patients without any further intervention. Other patients had partial healing and control of symptoms using nonoperative methods (72–74).

In those children who have responded to medical therapy but have persistent intersphincteric or complex fistulas, advancement flaps may be considered. Rectal mucosal flaps preserve continence and provide healing in 64–71% of patients with transsphincteric abscesses (64,75). The recurrence rate may be up to 50% (76). Often advancement flaps are done under cover of ileostomy.

Rectovaginal fistulas may also be cured with advancement flaps. In one study, healing was initially successful in 54% of patients, and after reoperation, increased to 60% during follow-up of 5 years (77). A transvaginal approach is recommended for those with persistent rectal disease. High success rates of 90% may be achieved with this approach in patients with controlled disease (64). Rectal dissection is also successful when the disease has settled down after fecal diversion. Cutaneous advancement flaps for persistent mucosal disease have also been advocated (78). A 70% healing rate may be expected under the best of circumstances (79).

The ultimate definitive surgical management is proctocolectomy. Despite removal of all disease, persistent draining sinuses may be expected. It is for this reason such procedures are avoided in children, particularly as most families would like to wait for advances in medical therapy.

NONOPERATIVE MANAGEMENT

Routine administration of infliximab with immunosuppression is the mainstay for perianal Crohn's disease (46). It is essential that sepsis be controlled prior to initiation of therapy. The pediatric surgeon should be consulted and care for the patient as part of the initial evaluation. After the patient has been established on therapy, local wound care remains an essential part of treatment and symptom relief. Perianal hygiene with daily sitz baths, debridement, and application of barrier creams and ointments can be exceptionally effective. Patients may purchase or create specialized cushions for sitting. Bowel management is also helpful in those rare patients with constipation or very loose stools that exacerbate the issue. A draining seton suture is effective for prevention of abscess and recurrent pain (64). The goal of therapy is to produce a well-established draining tract without obstruction or sepsis. The seton is therefore placed in a noncutting fashion using a soft silicon vessel loop or monofilament suture. With the addition of immune and biologic therapy, seton suture placement provides excellent control of disease and will lead to healing in most circumstances (74).

Injection of fibrin glue into the remaining fistula tract has also been moderately successful (80). Once again, it is important to establish that the disease is well controlled and that the internal fistula opening may be sutured closed at the time of fibrin glue injection. There should be minimal to no granulation tissue left within the tract that would prevent adhesion of the glue to the walls. Should a cavity still be present, the seton suture is best left in place for further healing.

LONG-TERM ISSUES

Active severe perianal disease may progress to anorectal stricturing that responds to dilations under anesthesia. Again, dilation is preferred as noninvasive management in combination with infliximab or other immune modulation. Surgical anoplasty is effective for division of the stricture, but is contraindicated in patients with active mucosal disease (81).

Well-established complex perianal fistulization may never heal. Fistulotomy can be curative but may result in a nonhealing open wound or incontinence. Advancement flaps are preferred over fistulotomy in select cases after investigation with MRI and treatment with immune modulators (47). Even so, symptom control is the goal of therapy rather than cure of the disease. Massive hypertrophy of anoderm may be excised, but again, there is a distinct risk of nonhealing. Chronic anal sphincter spasms respond to botulinum toxin injection, which is preferred to sphincterotomy in younger

patients. Botulinum toxin injection requires repetition, and patients usually recover more slowly than if a surgical sphincterotomy was performed, but there is less risk involved.

ULCERATIVE COLITIS

Proctocolectomy cures the intestinal manifestations of ulcerative colitis. Reoperative surgery is most common for the restorative portion of the procedure; creation of an ileal reservoir. This section reviews the complications and repair of ileal pouch anal anastomotic (IPAA) surgery.

Failed Procedure

CLINICAL PRESENTATION

Pouch failure is the most common reason for a poor outcome, and the ileoanal anastomosis is the most common source of the complication (82). The risk of pouch-related failure increases with time, 10% at 10 years is a good estimate (83). Early pelvic or anastomotic sepsis increases long-term risk of failure by five times (84,85). Patients present with anastomotic separation, dense stricturing, abscess formation, and subsequent fistulization. Local symptoms include anal pain, sphincter spasm, and fluctuant sepsis, leading to fistulization. Secondary symptoms from obstruction of the pouch often overshadow the primary manifestations of the complication. Children suffer from urgency, frequent bowel movements, inability to fully evacuate, nighttime incontinence, and poor sleep. Patients may decrease oral intake to reduce the problems with pouch function. These secondary symptoms, especially nighttime incontinence and frequent stooling, are directly correlated to poor quality of life (86,87).

Regional inflammation may also disrupt pelvic floor function, interfering with bladder and urethra control. Inflammation extending through the rectovaginal septum may produce abscesses, fistulization, and dyspareunia. These young patients are miserable, sleep poorly, and must reorder their day to deal with pouch complications. The intensive medical and surgical management contributes to the unacceptable quality of life.

Pouchitis without other technical complications may also lead to pull-through failure. This condition alone is responsible for approximately 10% of all pouch failures (88). Patients have reverted to ileostomy to avoid the ongoing symptoms of stool frequency, bleeding, mucus discharge, urgency, nighttime continence, and extraintestinal manifestations such as arthralgias and erythema nodosum. Prompt treatment will usually ameliorate symptoms. Treatment resistance usually leads to a diagnostic work-up for Crohn's disease (85).

Intraabdominal conditions above the level of the pouch are a rare cause of procedural failure. Patients may have kinking at the bowel pouch interface or present with intermittent small bowel obstruction secondary to adhesions. Both of these conditions will produce abdominal distention, cramping, and poor pouch evacuation. There are not the usual pouch-associated symptoms such as urgency and incontinence, which helps the surgeon differentiate these diagnoses. Patients with familial adenomatous polyposis who develop desmoid tumors have had pouch-related procedural failure secondary to obstructing tumor masses (89). Chemotherapy or radiotherapy may induce tumor necrosis. Pouch salvage is dependent on regression of the desmoid tumor (90,91).

Primary inability to complete the IPAA procedure may ultimately lead to surgical failure. Certain children have insufficient mesenteric length to create a standard J-pouch. Body habitus and the influence of corticosteroids predispose patients to mesenteric infiltration with fat and edema. Uncontrolled disease activity temporarily shortens the mesentery by reducing elasticity while increasing fragility. In these circumstances, intraoperative decision-making will usually prevent procedural failure by delaying reconstruction until all reversible factors have been treated. Failed procedures are converted to delayed procedures for reoperation several months later.

DIAGNOSTIC EVALUATION

Local examination and MRI imaging are the two best modalities for investigation of pouch failure (85). For many children and young adolescents, examination under anesthesia provides the best setting to diagnose complications of stricture, abscess formation, fistulization, and anastomotic separation. A concurrent pouch endoscopy helps exclude severe pouchitis. Care should be taken to biopsy the prepouch ileum as well as the pouch to rule out Crohn's disease. Endoscopic endoanal ultrasound has been reported as a helpful adjunct in adult patients with anastomotic or pouch-related complications. This procedure has not been reported in children. Its effectiveness for local evaluation has been shown to be similar to MRI scanning (69).

MRI is the state-of-the-art for regional identification of disease, including fistulization, cuff abscess, wall thickening, and pelvic sepsis. CT scanning is a reasonable substitute and may be more practical for younger children. If an MRI is indicated, the scan is often combined with the examination under anesthesia.

Because Crohn's disease is such an important factor in pouch failure, a general intestinal evaluation is indicated. Studies include serology, small bowel contrast follow-through and upper and lower endoscopy. Acute phase reactants are usually of less help because most patients will have an inflammatory response to the complication.

Plain radiographs of the abdomen are always helpful to exclude prepouch small bowel complications such as intermittent obstruction secondary to adhesions or kinking at the bowel pouch interface. Contrast through the pouch determines the level and degree of obstruction.

Physiologic function studies of the pouch and the anal sphincter may identify functional causes of IPAA failure. Occasionally, patients will have a large amount of retrograde defecation or incomplete evacuation secondary to pelvic floor dysfunction. Positive studies indicate a bowel training program rather than further surgical intervention.

The 5-fold increased risk of late pouch failure secondary to early anastomotic sepsis mandates careful evaluation of the pouch and the ileoanal anastomosis prior to initial closure of the ileostomy (82,85,92). A pouchogram is performed as a routine investigation. Use of water soluble contrast through a small catheter identifies complications at the anastomosis or within the pouch itself. Should a fistula be identified, further time is allowed for healing prior to stoma closure. For major separations, reevaluation under anesthesia will help direct future management including laparotomy. Children and parents are interested in ileostomy closure as soon as possible; however, surgery should not be considered prior to 6 weeks after colectomy. It is safer to wait 3–6 months after initial operation and mandatory to delay this long if abnormalities are identified on pouchogram. Rarely, leaks are noted in the staple line or at the top

of the pouch (84). Surgical judgment is required to determine whether reoperation or further observation under cover of an ileostomy is indicated.

OPERATIVE TECHNIQUES

A temporary diverting ileostomy is recommended for any major revisional surgery of the pouch. When perianal sepsis is involved, there is a chance ileostomy prior to reoperation will allow for the process to heal without further surgery. If not, and reoperative procedures are necessary, fecal diversion will reduce the amount of induration and provide a more elastic compliant field for operation. Ileostomy is not required for local revisions of more minor anastomotic problems without sepsis.

Reoperative surgery for ulcerative colitis is so strongly influenced by the presence of sepsis that the procedures may be categorized by its presence or absence (82,84,93). Acute and chronic sepsis produces pelvic abscess cavities and perineal fistulas to the skin, vagina, or surrounding viscera. Anastomotic separation and dense stricturing coexist with these complications.

The risk of early sepsis after initial ileoanal procedures decreases with increased surgeon experience (84,85). Identification of the complication prior to ileostomy closure allows for antibiotic management and percutaneous drainage of abscesses. Local transanal drainage of sepsis or curettage of cavities speeds healing. Occasionally, multiple local procedures are necessary prior to stoma closure. Once eradicated, long-term pouch success rates are similar to those without sepsis (92%) (94). Patients with more severe infection requiring abdominal reoperation for control have a more dismal outlook. More than one-half lose their pouch, and less than one-third have good long-term function. Similarly, 30% of patients with extensive separation of the anastomosis have long-term failure despite reoperative attempts (85). Fortunately, most of the disruptions are localized and minor revisions may be considered.

Such revisions may also be considered without fecal diversion. The pouch is dissected free from the cuff for 2–3 cm and advanced over the separation. A two-layer hand-sewn anastomosis is done with absorbable sutures. Success depends on the degree of sepsis and the tension on the suture line. There is a strong preference for pediatric surgeons to revise the pouch using a transanal approach. Salvage is possible in 70–94% of patients when the complication is localized and sepsis controlled (92,93,95–97). Temporary diversion is advocated for more extensive disruptions or ongoing sepsis not amenable to local treatment.

Redo pull-through procedures may be attempted 6–12 months after diverting ileostomy. The procedure is best approached through a combined transanal and abdominal route. Although pediatric surgeons are highly adept at performing muscle cuff dissections from below, the pouch remains tethered to the pelvic floor and is not amenable to full pull-through without mobilization of the proximal bowel and mesentery. It is imperative that a tension-free anastomosis be performed at the anus. The procedure is gratifyingly successful with full mobilization of structures. Generally, there has been interval lengthening of the mesentery, which will allow for a straightforward handsewn anastomosis at the dentate line. Redissection along the muscle cuff is problematic at the site of prior sepsis, but otherwise goes well. A plane may be visualized between the serosa of the ileum and the rectal wall and is followed down to the anastomosis without difficulty. Local resection of the muscle cuff is often necessary at the site of previous fistulization or abscess. Conversion to a Swenson-style pull-through is not recommended. It is generally difficult to perform a stapled anastomosis

on a redo procedure, but with the added length from the abdominal portion, handsewn resuturing at the dentate line is not a problem. Four stay sutures from pouch to muscle cuff help anchor the redo pull-through in position for the final anastomosis between full-thickness pouch and dentate line.

The ileostomy remains for at least 6–12 weeks and is closed after a repeat pouchogram confirms full healing. The patient is examined under anesthesia at the time of ileostomy closure to ensure that the anastomosis is intact and the inevitable stricture is easily dilatable. Successful salvage in combined pediatric and adult reviews approaches 90%, but is more realistically estimated at 50–60% (82,92,96). Furthermore, the accumulative failure rate increases with time of follow-up and presence of Crohn's disease (84). Patients at highest risk have undergone transabdominal revisional operations or have anastomotic separation or vaginal fistulas (85).

Repair of a pouch vaginal fistula depends on the presumed etiology. Patients with technical complications related to dissection of the muscle cuff or stapler injury to the posterior vaginal wall may expect excellent results from salvage surgery. Fistulas resulting from anastomotic break down and pelvic sepsis carry a poorer prognosis, but unless Crohn's disease is implicated local repair is successful (98).

Fistulotomy, seton sutures or direct repair are unsatisfactory approaches to pouch fistulas. Transvaginal and endoanal advancement procedures are successful in 50–78% of cases (98,99). Fistula repair proceeds much like an anterior sagittal approach to imperforate anus, with disconnection of the fistula and closure of the vaginal defect. The ileoanal anastomosis is advanced over the fistula and sewn to the dentate line, where distal advancement is possible. Otherwise, direct suture closure of the anastomotic defect is successful.

Transabdominal repair is necessary for higher fistulas or when there is a longer cuff of retained rectum. Extensive proximal mobilization of the pouch is necessary for distal advancement to the dentate line and reduced tension on the anastomosis. Transabdominal salvage rates approach 80% (85).

Apart from sepsis, ulcerative colitis procedures fail for other mechanical reasons. Several options are available for those patients with inadequate mesenteric length for primary pull-through. A straight ileoanal anastomosis has good long-term functional results. It is technically the best option for patients with significant complications from prior surgery or inadequate mesenteric length. Storage capacity is initially limited but will improve over the ensuing years to provide good quality of life and return to normal activities (100).

Creation of an S-pouch is an alternative to a J-pouch in selected patients. Prior knowledge of an S-pouch creation and specific determination of the length of the outflow limb are essential for good function. Most difficulties with S-pouches occur secondary to partial obstruction to emptying (101).

Delayed primary creation of a J-pouch is the best option for patients found to have technical issues with the primary procedure. High-risk patients include those on maximal steroid therapy, fulminant colitis, and major alterations in body habitus. Such children and their families may be informed of the possibility of delayed repair prior to surgery. An end ileostomy is created, preserving the ileocolic arcade. It is essential to perform a "just adequate" Brooke ileostomy to preserve as much length as possible for the future J-pouch. Once the patient recovers from surgery and is weaned off steroids, body habitus and local mesenteric issues improve such that a J-pouch can be created

and the anastomosis completed. It is advisable to leave a small rectal stump above the level of the peritoneal reflection for ease of dissection.

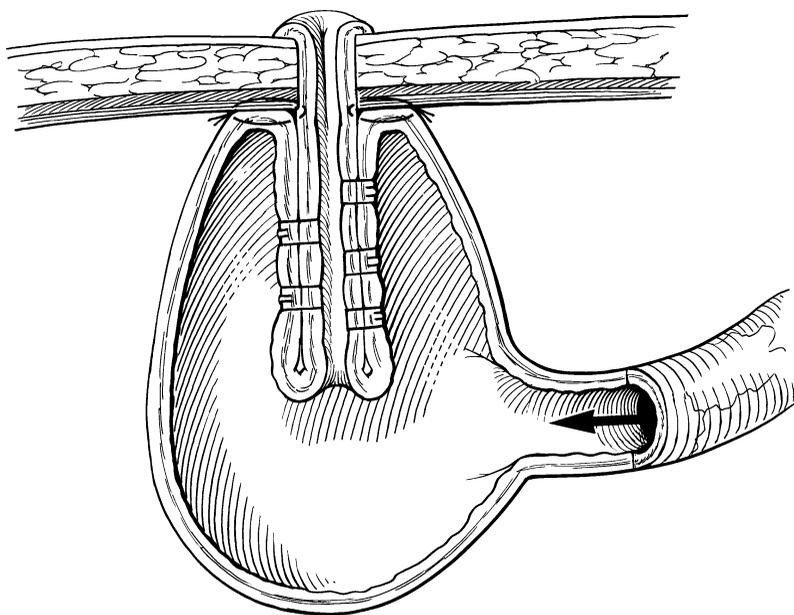
Intraoperatively, a delayed procedure is anticipated at the time of colectomy by a trial prior to creation of the J-pouch. If the bowel does not reach over the pubis, the IPAA will probably not be successful. Mesenteric lengthening is attempted at that time but if the trial again is not successful, mesenteric length will improve over the next 3–6 months with end ileostomy. Reoperation is then identical to a planned staged IPAA.

Occasionally, patients will have already undergone a rectal mucosectomy prior to identification of procedural failure. In these patients, reoperation is made more difficult because of scarring of the muscle cuff; however, completion is still possible. Pelvic dissection is aided by placement of an instrument through the anus to identify the top of the muscle cuff. The cuff is then reopened and expanded to allow passage of a pouch. It may be necessary to split the muscle cuff posteriorly. Again, all attempts should be made to preserve the deep pelvic nerves by staying within the cuff. If the scarring is too intense, resection is certainly indicated and is usually performed at the top of the prior dissection.

For patients with a “frozen pelvis” secondary to multiple episodes of sepsis or repeated surgical attempts, a permanent end ileostomy is the best solution. Creation of a Koch pouch is still possible and performed in highly select cases (102) (Fig. 4). Experience with such procedures is now limited, and consultation with a senior surgeon is recommended.

LONG-TERM ISSUES

Colectomy and end ileostomy cures the intestinal manifestations of ulcerative colitis. Pouch procedures are performed to restore function rather than treat disease. When complications from the restoration become chronic, quality of life is poor. Frequent



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Fig. 4. The Koch pouch is a continent ileostomy used in highly select cases.

defecation and loss of nighttime control seriously impair the abilities of the adolescent or young adult to attend school or embark on a career (86). Multiple procedures and return appointments hamper patient mobility, such that consideration of college and first jobs become problematic. These disadvantages eventually outweigh the stigmata of an ileostomy. When surveyed, patients with permanent ileostomies still prefer a restorative pull-through but have quality of life indicators similar to those without ileostomy (103). It is helpful to set goals for therapy and establish a time line for reevaluation. Although difficult to consider, ileostomy remains an excellent long-term solution for patients with colitis after colectomy.

Pregnancy and vaginal delivery after a pull-through procedure are possible and recommended for those women without serious pouch- or anastomotic-related complications. An updated review shows no difference in birth weight, pregnancy, or delivery complications or vaginal delivery rates before or after IPAA procedures. There was a higher rate of planned Cesarean sections and daytime stool frequency at long-term follow-up; however, vaginal delivery and successful outcomes can be routinely anticipated after surgery for ulcerative colitis. Pouch function and complications of pregnancy were unaffected by surgery (104).

Pouchitis

CLINICAL PRESENTATION

Pouchitis is a nonspecific mucosal inflammation of the ileal reservoir. It occurs in 26–47% of children; the incidence increases with length of follow-up (86,96,97). Children with previously excellent function suddenly experience a three- to fourfold increase in stool frequency. Nighttime incontinence recrudesces and most complain of urgency, mucousy discharge, and occasional bleeding. Extraintestinal symptoms, particularly arthralgias and erythema nodosum, may occur. The symptoms of urgency, frequency, and nighttime stooling are the primary indicators of disease that drive diagnostic investigation and treatment (84,105,106). The complication of pouchitis is not life-threatening nor is it usually life-limiting for most children. The symptoms, however, represent the perception of return to former disease. Patients feel ill, are unable to go to school, and cannot socialize.

DIAGNOSTIC EVALUATION

Pouchitis is a clinical diagnosis based on symptoms. Endoscopic and histologic findings do not necessarily correlate with the severity of the clinical problem (106–108). The creation of an ileal reservoir is for restoration of bowel function. Any malfunction of the pouch defeats that goal and is labeled pouchitis when no other surgical complications are present.

The diagnostic evaluation has two purposes; to identify and treat surgical complications discussed in the previous section, and when possible, discover causative factors for pouchitis. The latter include anal stricture, retained rectum, small volume pouch, medication-induced irritability, and Crohn's disease (105). Such conditions require a different approach to therapy than idiopathic pouchitis (97,109,110). The surgical team is in a unique position to help direct investigation. Their direct knowledge of the patient and the procedure helps prioritize the possible conditions listed previously.

In the simplest of cases, when the likelihood of complications is small, the diagnosis is confirmed by treatment response. Children with sudden onset of classic pouchitis

symptoms without risk factors usually respond to oral antibiotics. A prompt return to baseline pouch function within 12–24 hours makes the diagnosis. This pragmatic approach may include some cases of viral or food-induced illness, but when the goal is rapid return to normal, stool function and quality of life over diagnosis is acceptable.

Children with risk factors or recurrent symptoms require work up. Examination under anesthesia and pouch endoscopy are mainstream investigations (105,109,110). A thorough examination excludes surgical complications and allows for anastomotic dilation. Endoscopy evaluates mucosal disease, notes any progression into the prepouch ileum, and obtains biopsy for histologic confirmation and to exclude Crohn's disease. Typically, diffuse mucosal aphthous ulceration and contact bleeding are identified. The changes revert to normal and the small bowel directly above the pouch. Further investigations depend on findings at examination under anesthesia and prior knowledge of the procedure. A pouchogram, MRI, or CT scan may identify septic complications or mechanical issues of the pouch and its connections.

MEDICAL VERSUS OPERATIVE MANAGEMENT

Pouch dysfunction secondary to complications of stricture, retained rectal cuffitis, Crohn's disease, and local sepsis are treated surgically or with other specific medical intervention (109). For the remaining patients with idiopathic pouchitis, there are no surgical options. Treatment of pouchitis with the oral antibiotics metronidazole and Ciprofloxacin is the most effective strategy. Children promptly respond to initial doses similar to those for treatment of serious infection. Gradual reduction of therapy is possible within days of treatment. Once recurrent, most families learn to manage the symptoms with these antibiotics on a routine basis.

Chronic pouchitis is controlled with daily low-dose metronidazole or Ciprofloxacin, which may be alternated on a weekly or monthly schedule. Amoxicillin and vancomycin have also been successful. Many other therapies have been tried for pouchitis; the most recent has been probiotics (111). Like multiple other agents, its effectiveness remains in question.

Investigation of lifestyle and over-the-counter medication usage are important in the treatment of pouchitis. Smoking, caffeine use, stress, and nonsteroidal antiinflammatory drugs (NSAIDs) all contribute to pouch irritability.

LONG-TERM ISSUES

Approximately 10% of ileoanal procedures fail secondary to severe mucosal pouchitis that is resistant to medical therapy (84). These patients require pouch excision. Of those with chronic pouchitis controlled by medication, most have serious questions about the long-term use of antibiotics. These effects have not been quantified. Dysplasia in chronically inflamed pouches has also been raised as a concern for children facing many years of disease. A relatively short-term follow-up study did not identify abnormalities (112). Finally, in children who are expected to keep their pouch for many decades, aging and decline in function remains a concern. A recent study of 15-year follow-up has shown durable pouch function without decline in continence or quality of life (113).

INDETERMINATE COLITIS—OPERATIVE CONSIDERATION AND MANAGEMENT

Indeterminate colitis is a distinct clinical pathologic entity diagnosed in 5–15% of patients requiring surgery for inflammatory bowel disease. With follow-up, the percentage of patients who continue with this diagnosis decreases, suggesting differentiation into ulcerative colitis or Crohn's categories (114). The major surgical question is the greater morbidity following an IPAA procedure. Adult patients with indeterminate colitis do indeed have a higher rate of serious complications (20%) (115) than those with ulcerative colitis (3.5–10%) (84), but still less than patients with Crohn's disease (35%) (85). The long-term success of IPAA for indeterminate colitis is 73–85%, compared to 89% for ulcerative colitis. Such excellent results make a good case for IPAA in patients with indeterminate colitis (116).

Two precautionary statements are necessary; first, indeterminate colitis patients with features favoring Crohn's disease may benefit from delayed IPAA 6–12 months following colectomy. Second, what can be said for children where no data are available? The answer will depend on age of the child, severity of disease, and the imperative to operate. The younger the child, the more common the presentation of pancolitis in Crohn's disease. Therefore, surgeons who are asked to see children under the age of 8 years for colectomy should proceed cautiously to full reconstruction. In these young patients with severe disease, even though it is technically easier to perform a colectomy with J-pouch reconstruction, it is best to delay reconstructive surgery until pathology confirmation.

The real dilemma occurs in the younger child who is otherwise well but has steroid-resistant disease. These children are usually on low-dose oral corticosteroids and are excellent candidates for immediate reconstruction. Many have decreased quality of lives and are unwilling to consider delayed reconstruction and long-term ileostomy. The options are to go ahead with a J-pouch, knowing a higher percentage may have Crohn's disease, versus a three-stage procedure. Data from adult studies suggest a primary IPAA may proceed with excellent results; however, in patients with features favoring Crohn's disease, the better alternative may be to proceed to ileostomy, which can be converted an ileal J-pouch after eliminating the diagnostic possibility of Crohn's disease during follow-up.

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16

Reoperation for Stoma Complications

Benedict C. Nwomeh, MD

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OVERVIEW OF COMPLICATIONS

Major complications could occur in up to 75% of children following colostomy or ileostomy, with an overall revision rate of approximately 15% (Table 1). Because of the higher rate of complications associated with loop ostomies, most pediatric surgeons prefer divided stomas. A good ostomy is best obtained by careful preoperative planning, meticulous surgical technique, and detailed attention to skin care. Skin complications such as dermatitis, granuloma, and ulceration are the most frequent problems with ostomies. Stomas made in the small intestine (enterostomas) are associated with more complications than colostomies. In addition, transverse colostomies cause more problems than sigmoid colostomies. The expertise of an enterostomal therapist should be sought in the care of ostomies, and in the prevention and management of skin problems. With temporary stomas, the occurrence of complications should prompt consideration for closure of the ostomy, rather than revision. The following discussion is limited to those complications that often require reoperative surgery. Other complications usually requiring the care of a good enterostomal therapist are listed in Table 2.

SPECIFIC COMPLICATIONS

Ischemia/Necrosis

Significant ischemia or frank necrosis is apparent early in the postoperative period. The cause is either arterial insufficiency from excessive stripping of the mesentery, or venous congestion caused by tension on the bowel, or a tight fascial opening. Obese patients are particularly at risk of developing stomal ischemia. Some degree of edema and venous congestion normally occurs with new ostomies, especially if previously distended bowel is used, but usually resolves within 48 hours. If mucosal necrosis is limited to the portion superficial to the fascia, an expectant approach may be employed but the stoma should be monitored closely for progressive necrosis or subsequent development of stenosis, stricture, or retraction.

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Table 1
Stoma Complications in Children

<i>Complication</i>	<i>Incidence (%)</i>
Ischemia/Necrosis	<1
Stenosis/stricture	3–6
Retraction	2–4
Prolapse	12–24
Parastomal hernia	1
Skin excoriation	20–30
Bleeding	1–10
Obstruction	1–6

OPERATIVE TECHNIQUE

Sufficient mobilization of bowel and mesentery will be required to allow resection of the ischemic portion and creation of a new stoma with well-perfused bowel without tension. It is usually necessary to reopen the main incision to facilitate this revision. Mesenteric stripping of the stoma should be limited to the 1–2 cm necessary to evert the bowel for maturation.

Stenosis/Stricture

Stenosis or stricture of the stoma may occur at the skin or fascial level and is clinically apparent as reduced stoma output or frank bowel obstruction. Potential causes include a tight skin or fascial opening, ischemia at the mucocutaneous junction, and prolonged serositis with subsequent fibrosis when the stoma has not been matured. Gentle probing with a soft catheter may help to localize the narrowing. In mild cases, gentle dilatation using serial rubber catheters or the Hegar dilator may resolve the issue, but this procedure carries the risk of bowel perforation. Recurrent Crohn’s disease causing a stricture is best detected using contrast studies and endoscopy.

Table 2
Management of Other Complications of Stomas

<i>Complication</i>	<i>Cause</i>	<i>Treatment</i>
Wound infection	Intraabdominal infection	Antibiotics
	Stoma located in main incision	Drainage
Skin excoriation	Poorly fitting appliance	Enterostomal care
	Contact dermatitis	Stoma revision
	Retraction, prolapse	Stoma relocation
Bleeding	Trauma	Enterostomal care
	Mucosal lesion (e.g., polyps)	Endoscopy through stoma
Fistula	Full thickness placement of suture	Enterostomal care
	Pressure necrosis from appliance	Stoma revision
	Recurrent Crohn’s disease	

OPERATIVE TECHNIQUE

For skin-level stenosis, the scar ring should be removed by circumferential excision. The fascial opening can be enlarged if necessary. The bowel is then mobilized sufficiently to allow revision of the stoma at the same site. For longer or more proximal strictures, and in recurrent Crohn's disease, a laparotomy may be required for more extensive mobilization of bowel and resection of the affected segment.

Retraction

It is important to recognize the cause of stomal retraction. The most common problem is undue tension on the stoma caused by inadequate mobilization of the proximal bowel and its mesentery. Anchoring the bowel to the fascia with sutures does not mitigate the tension in the bowel and cannot be relied upon to prevent retraction of the stoma. Secondly, retraction may follow stomal necrosis. Thirdly, retraction may be caused by prolonged serositis with subsequent shortening of bowel when the stoma has not been matured. Finally, recurrent Crohn's disease may manifest as stomal retraction and should be investigated with contrast studies and endoscopy. The retracted stoma causes leakage of intestinal contents under the stoma pouch, creating difficulty with maintaining the appliance.

OPERATIVE TECHNIQUE

Revision of the stoma is usually required. If the retracted stoma is sufficiently mobile to allow the bowel to be everted, the bowel walls can be fixed together by inserting several interrupted absorbable sutures with full thickness bites. Alternatively, several firings of a noncutting linear stapler can be used to fix the bowel (Fig. 1).

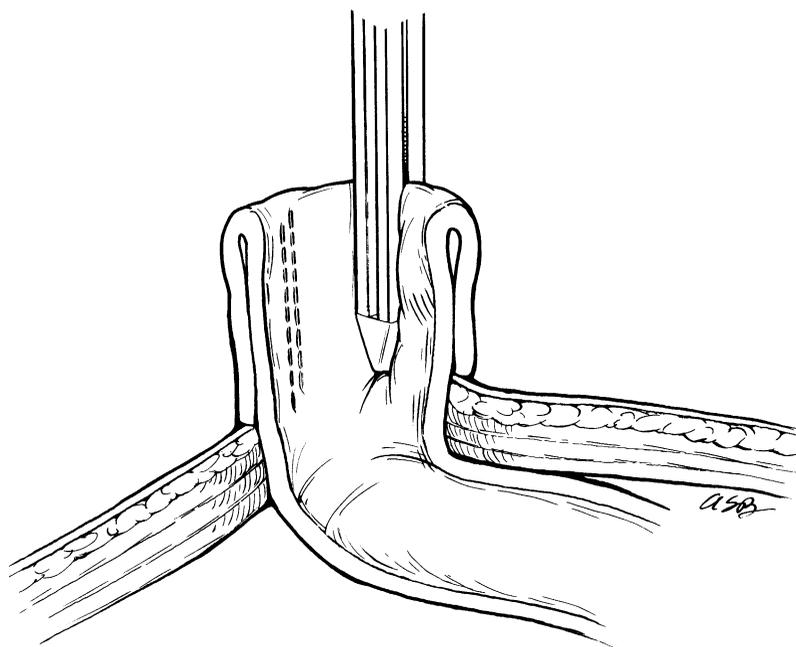


Fig. 1. The stapling technique for fixation of a retracted stoma. The stoma is retracted to its full extent by placing three pairs of Babcock's forceps (not shown). A noncutting linear stapler (such as the GIA stapler without the blade) is placed with the blade toward the mucocutaneous junction between the Babcock forceps. Care is taken to avoid the mesentery before firing the stapler. Three parallel rows of staples fix the two walls of the ileum together.

Adhesion between the serosal surfaces of the everted stoma will usually have occurred before the sutures are absorbed or the staples separate. More commonly, the retracted stoma is fixed in position and the goal is to mobilize sufficient length of bowel and mesentery so that maturation of the stoma can be accomplished without tension. Additional mobilization of bowel often requires a laparotomy. Limited bowel resection is performed to remove ischemic or stenotic segment, or area of recurrent Crohn's disease. The new stoma should be matured at the same site except when retraction has resulted in significant skin excoriation or abdominal wall sepsis; in these cases, a new ostomy site should be established.

Prolapse/Intussusception

A prolapsed ostomy alarms and frightens both child and parents. Prolapse usually occurs when a skin opening is made to accommodate dilated bowel, which upon shrinking leaves a loose stoma. Inadequate fixation of the mesentery to the parietal peritoneum may also lead to prolapse. Prolapse may manifest as protrusion of the terminal portion of the bowel, or as intussusception of the intraabdominal portion through the opening. Loop stomas are more likely to prolapse than end stomas, and the distal segment of the loop ostomy is most frequently affected. A coexistent parastomal hernia may predispose to prolapse. Prolapsed stomas interfere with the application of the stoma appliance, causing leakage. Trauma to the exposed bowel, caused by desiccation or by an ill-fitting appliance, may cause mucosal ulceration and bleeding. Spontaneous or manual reduction is usually possible in the early stage, but surgical intervention is eventually required for persistent prolapse, intestinal obstruction, or strangulated bowel.

OPERATIVE TECHNIQUE

For temporary relief, a simple purse-string suture with a nonabsorbable monofilament material may be used, similar to the Thiersch technique for rectal prolapse (Fig. 2). This is ideally suited for prolapse of a temporary ostomy because the fixed ring would produce stenosis as the child grows. A simple technique described by Gauderer uses a "U" stitch from the lumen of the reduced bowel through the abdominal wall with a double-armed needle (Fig. 3). Before tying the suture ends, each needle is passed through a pledget, thus creating an internal and external bolster that attaches the bowel to the body wall and prevents the suture from cutting through. Amputation of the prolapse is a tempting option that alleviates the problem, but early recurrence is likely. A more definitive revision including resection of prolapsed bowel and fixation of the mesentery may bring permanent relief to troublesome ostomy prolapse. This procedure usually requires reopening of the main abdominal incision. In the case of a prolapsed loop ostomy, the loop may be divided, with the closed distal end returned into the abdomen. This converts the loop to an end ostomy, which is less likely to prolapse. Repair of a coexisting parastomal hernia should be considered in order to reduce the risk of further recurrence. When appropriate, closure of the ostomy is the best option.

Parastomal Hernia

Parastomal hernia appears to occur less frequently in children than adult patients, with incidence less than 1%. The most likely cause is the creation of a fascial aperture relatively larger than the bowel used for the ostomy. Although parastomal hernia

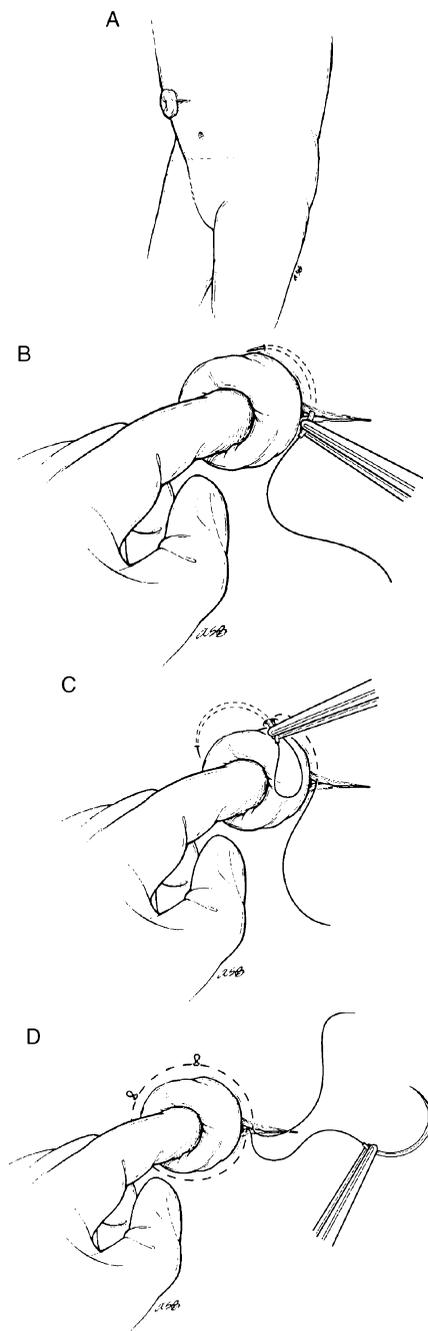


Fig. 2. A purse-string suture technique for correcting stoma prolapse. **(A)** A 1-cm skin incision is made at the medial angle of the stoma down to the subcutaneous tissue. **(B)** A finger is inserted into the stoma as a guide and a 1-0 monofilament nonabsorbable suture with a round, cutting needle is passed around the colostomy staying within the subcutaneous layer. The needle is placed as far as can comfortably go, usually approximately one-quarter of the circumference, then brought out through the skin. **(C)** The needle is passed again through the same skin exit site toward the lateral corner. **(D)** One or two more passes of the needle are made as it is marched circumferentially around the stoma, until it is brought out through the medial incision. With a finger remaining in the lumen, the suture is tied, causing puckering of the stoma without completely occluding the lumen.

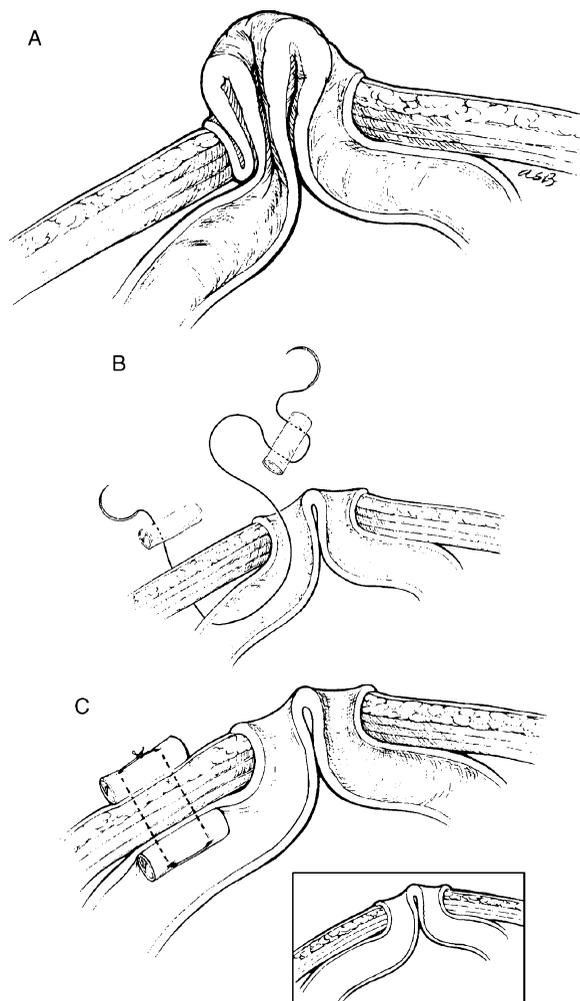


Fig. 3. An alternative technique for temporary control of stoma prolapse. (A) Prolapse of one limb of a looped ostomy. First, the prolapse is reduced with a gentle inward pressure. (B) A double-armed 3-0 nonabsorbable monofilament suture is used. One needle is placed through a latex bolster (or pledget) and the second needle passed 2–3 cm into the reduced limb of the stoma, then through the bowel wall and out the abdominal wall an equal distance from the stoma. The needle is then placed through a separate bolster. (C) The suture is tied without undue tension, sandwiching the bowel and abdominal wall between the bolsters. The insert shows the bowel adherent to the abdominal wall following removal of the bolsters 2 weeks later.

has been related to the creation of the stoma at one end of the working incision, a frequent event in infants especially during procedures for necrotizing enterocolitis, it remains a rare phenomenon in this group of patients. Despite widely accepted dogma, recent evidence disputes the notion that parastomal hernia is more likely to occur when a stoma is brought out lateral to the rectus abdominis muscle. Wound infection, malnutrition, and obesity also predispose to parastomal hernia. Clinically, a bulge is noted in the abdominal wall to the side of the stoma (Fig. 4). This may cause difficulty with retaining the stoma appliance. Fortunately, more serious complications such as intestinal obstruction and strangulation are rare.

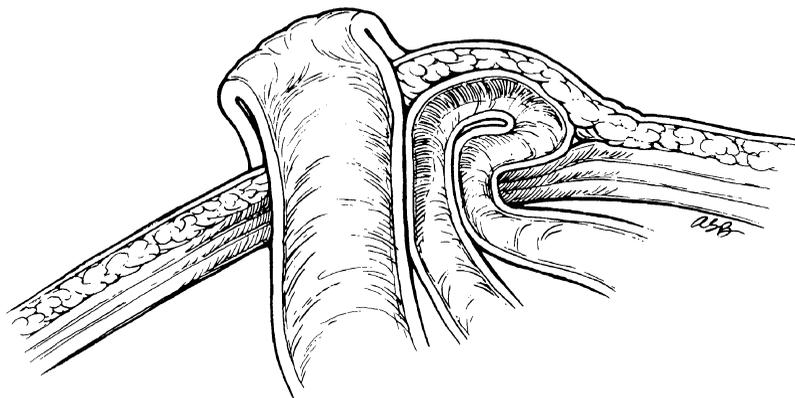


Fig. 4. Parastomal hernia, with a bulge in the abdominal wall to one side of the stoma.

OPERATIVE TECHNIQUE

Operative repair should be undertaken only if the hernia is problematic, for example persistent leakage of the appliance. The simplest procedure is to mobilize the stoma, repair the hernia snugly around the bowel, then mature the ostomy at the same site. Alternatively, the hernia could be repaired and the stoma relocated to another site. In children, the use of prosthetic mesh should be avoided. Because the results of these procedures can be disappointing, the best approach to a parastomal hernia complicating a temporary ostomy is to consider closure of the ostomy.

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Reoperative Surgery for Hirschsprung Disease

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INTRODUCTION

Hirschsprung disease (HD) is the most common cause of neonatal intestinal obstruction. Since Swenson in 1948 described his successful approach, several modifications and new operations had been described in an effort to improve the long-term outcome and quality of life (QOL) for patients with HD. By far the most commonly used approaches nowadays are the transanal approach whether laparoscopically assisted or not, modified Soave's pull-through, and Duhamel's retrorectal pull-through.

Each of these procedures has its own advantages and drawbacks. All of them ultimately have good results concerning the long-term outcome. But at this moment, none of these procedures are perfect and many children suffer from long-term complications. Interestingly, these complications may occur after what may be called a "successful pull-through" without any obvious cause or explanation.

MANIFESTATIONS OF LONG-TERM COMPLICATIONS AFTER A PULL-THROUGH

The most serious long-term complications that strongly impact the QOL of patients with HD after the definitive treatment falls into three groups; soiling/incontinence,

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persistent problems with the passage of stool (e.g., constipation), and recurrent enterocolitis. Not uncommonly, an individual child may have a combination of problems.

Soiling/Incontinence

Soiling/incontinence have been reported in variable percentages and degrees. Rates range from none in several series to as high as 74% in one series (1–3). This problem may be caused by scarring or iatrogenic damage of the anal sphincters. Soiling may also be caused by a loss of rectal sensation (which is critical for differentiating between gas, liquid, and solid stool) owing to destruction of the sensory rectal mucosa from a very low anastomosis. Finally, the most common cause of soiling after a pull-through is chronic constipation with overflow of stool. Incontinence should theoretically be preventable by using adequate surgical techniques; however, the problem persists. The work up for many of these problems is discussed later in this section. The management of incontinence ranges from a simple bowel management program to the placement of a permanent stoma. The use of an antigrade colonic enema may be quite useful in this group of patients. More recently, the use of an artificial sphincter or sphincter reconstruction has been employed. These therapeutic options are not discussed in this chapter.

Persistent Stooling Problems

Persistent stooling problems are a very annoying group of complications. They include recurrent distension, bloating, and constipation. Constipation is probably the most common complaint after a pull-through procedure. Typically, symptoms do not develop until a few weeks to many months after the pull-through (4). However, in the past decade, a number of investigators have recognized that obstructive symptoms are a significant issue, occurring in 11–42% of children after what appears to be a technically excellent operation (5–9). Rescorla et al. found that 88% of patients had difficulty stooling with a follow-up less than 5 years. Although some patients with incontinence were in this group, symptoms improved with greater follow up, with 100% having satisfactory stooling with a 15-year or more follow-up (10). This suggests that many have persistent problems with their internal anal sphincter after removal of all aganglionic tissue, and suggests that we are still missing a complete understanding of the pathophysiology of HD.

Stooling problems may arise from a number of etiologies, and include stricture or retained spur after Duhamel's procedure, retained or acquired aganglionosis, hypoganglionosis or performance of the pull-through at the level of the transition zone, associated motility disorders either focal or general, internal anal sphincter achalasia (IASA), and finally what is called "functional megacolon," or a chronically dilated distal rectum, most likely caused by many years of persistent difficulty in passing bowel movements. In the remaining group of children, there is no identifiable cause for their symptoms, and some of these children fail to respond to surgical or medical relaxation of the sphincter. Most of these children suffer from stool-holding behavior and are best treated by using a bowel management regimen consisting of laxatives, enemas, and behavior modification including support for the child and family (11,12). The approach to address the cause varies and one should be aware of all of these various possibilities to be able to successfully manage a complicated HD case.

HD-Associated Enterocolitis (HAEC)

HAEC remains the most common cause of death in HD. It can occur at any time, including both before as well as after the pull-through procedure. The incidence of enterocolitis in HD varies widely among published series depending on the definition and severity of the enterocolitis. A large multicenter study series showed an incidence ranging from 22–42% (13). The incidence of preoperative HAEC occurs in approximately 15–20% (14–16). Postoperative enterocolitis may affect one-third of patients, with a mortality rate varying from 0–30% (17). Many patients have more than one episode of HAEC (18). Relapses of enterocolitis can occur despite a defunctioning colostomy or a successful definitive procedure (14). Therefore, it is of utmost importance that the treatment of enterocolitis is started early and pursued vigorously to prevent the condition from becoming chronic (19). It is also interesting to know that the frequency of bouts of enterocolitis tends to decrease with time (20).

The exact etiology and pathogenesis is still obscure (21–23). In a multicenter review of primary endorectal pull-throughs, logistic regression analysis of risk factors for the development of enterocolitis showed that stricture development was a significant risk factor (13). Other risk factors include increased length of the aganglionic segment, female sex, Trisomy 21, and the presence of other associated congenital anomalies (24). Currently, no clinical factor or test is available to predict the development of this disorder. Additionally, there is no single strategy for prophylaxis from this major complication.

WHEN TO START WORK-UP FOR COMPLICATIONS

In general, complicated HD cases should be assessed thoroughly by an experienced personnel, and an algorithm should be tailored for each case according to the type and nature of the complication.

A significant number of children who undergo a successful pull-through develop persistent problems or complications. But frequently they outgrow most of these complications by the age of 5 years for unexplained reasons without the need for further operation.

Unfortunately, this is not always the case; some children may develop treatment resistant symptoms and require further intervention. For example, a child with persistent severe constipation leading to progressive abdominal distension may develop failure to thrive in spite of several trials of laxatives, enemas, and dietary control. A second example would be an infant with recurrent severe bouts of enterocolitis requiring multiple admissions irrespective of chronic and aggressive medications and irrigations. Another example would be a patient found to have a stricture or retained spur after Duhamel's procedure during rectal examination. All of these cases deserve further and thorough evaluation and assessment.

WORK-UP OF THE COMPLICATED POSTOPERATIVE PATIENT

Thorough History-Taking and Meticulous Clinical Examination

A detailed history should include the neonatal period, associated anomalies, type of presentation, time and method of diagnosis, preoperative course, type of procedure, detailed postoperative course stating frequency of stooling and number and severity of enterocolitic episodes, and methods of HAEC management.

General examination should be thoroughly done to assess developmental milestones, growth curve, and to allow recognition of associated syndromes and anomalies that may impact the outcome. Abdominal inspection and palpation is necessary to get an appreciation about the magnitude of distension, and to feel any impacted stool.

Rectal examination is essential and can yield very useful information. Patency of the anal canal is readily assessed with palpation of any stricture or narrowing of the anastomosis line, and occasionally retained spur after a Duhamel procedure (although such a spur is often beyond digital palpation). The anastomotic level is palpated, giving an idea about the previous procedure if it is technically done in a correct manner or not. The tone of the anal sphincter is assessed as normal, increased (suggestive of IASA), or low (pointing to incontinence). The rectal vault is also evaluated as either full of stool (supporting encopresis) or empty. A finding of encopresis by history or an abnormally large rectal vault may indicate a stool-holding behavior, and the child may benefit from behavioral/training strategies. A gush of gas and liquid stools with removal of the examining finger typically indicates persistent obstructive symptoms or ongoing HAEC.

Contrast Enema

A radiographic contrast enema can be very useful. It usually can differentiate between a number of problems, including: anastomotic stenosis or stricture, a twist of the pull-through segment, as well as a chronically dilated rectosigmoid colon. Occasionally, the enema can detect such irregularities of the mucosal lining, suggesting chronic inflammation after repeated episodes of HAEC. Such information is useful in understanding the etiology of symptoms as well as deciding the optimum operative approach that effectively addresses these problems.

Anorectal Manometry

Use of manometry is controversial in the postoperative evaluation of patients with HD. Anorectal manometry does have several uses. Manometry can be very helpful in the diagnostic work-up of IASA, which may coexist with HD (25). Manometry may also help identify patients with marked increases or decreases in the basal resting anal tone. One problem is that most children, even after a successful pull-through, do not regain the rectoanal inhibitory reflex. This is well illustrated in a review of children with stooling difficulties after pull-throughs. Moore et al. reported their experience with the use of manometry. In their series of 16 patients with stooling problems, manometric assessment was not significantly different from 28 patients who had normal stooling (26). They noted that of these 44 patients, a normal inhibitory relaxation was seen in only six children, of which three had IASA and three had normal stooling function. Additionally, only two of eight children with a high resting anorectal tone had stooling problems. Another problem is the variability in the normal values for basal resting sphincter tone in published series. This may also be compounded by the numerous artifacts associated with movements or crying. To improve cooperation, it is often helpful to perform manometry with mild sedation or anesthesia in these young infants. The rectoanal inhibitory reflex can be elicited even when manometry is performed using general anesthesia (27). One problem in which anorectal manometry will shed considerable information is in the patient with constant soiling. Clearly, there are two

potential diagnoses in this group of patients, incontinence versus encopresis. A loss of resting anorectal tone will certainly help point to the diagnosis of incontinence.

Colonic Transit Time

Transit time and other more sophisticated colonic motility studies may be helpful in detecting focal or generalized motility disorders. Some investigators report that 25–35% of patient with HD have an associated intestinal neuronal dysplasia (IND), and such motility studies may help to infer this rare diagnosis (28); whereas other investigators rarely encountered IND in association with HD (29). However, medical treatment with laxatives and enemas remains the method of first choice in children without disabling obstruction, and this may be expected to be successful in 75% of the patients (30–32). Anorectal myotomy has been reserved for those who fail these approaches, and is discussed in a later section (33,34). Generally, focal or localized IND will require resection and/or repeat pull-through if it involving the distal colon, and generalized IND requires bowel management regimen or stoma if conservative management failed.

Rectal Biopsy

A rectal biopsy, either mucosal or full thickness, is the most important and precise method for evaluation of chronic problems after a pull-through. For endorectal or Duhamel pull-throughs, a properly placed suction rectal biopsy in the posterior wall should demonstrate ganglion cells. Not uncommonly, attaining this information in an older child is difficult. Should the biopsy show aganglionosis, or be too superficial, the surgeon should then turn to a full-thickness biopsy. The authors would also suggest an intermediate approach. By this, the procedure is performed surgically, but with the biopsy only going through the submucosa. This has the advantage of reducing scarring if additional surgery is needed. As Fig. 1 suggests, a full-thickness biopsy after a pull-through procedure is not straight-forward. First, the distance between the dentate line and anastomosis may not be clear. With growth, the distance from the dentate line to the anastomosis may become greater than expected. Second, a full-thickness biopsy

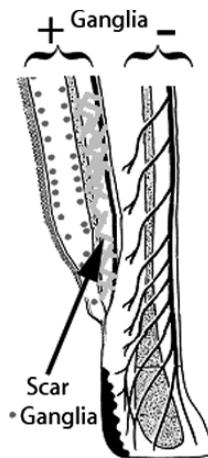


Fig. 1. Cross cut of the anatomy of the anus and rectum after an ERPT. Note the multiple layers of bowel, which might be sent to pathology with a full-thickness biopsy.

will incorporate both the ganglionic pull-through wall as well as the outer aganglionic lining. For all pull-throughs, a large amount of scar tissue may be encountered. Careful labeling of the specimen in terms of distal and proximal orientation is essential for correct pathologic interpretation. It is recommended to take serial biopsies at 1, 2, 4, and 6 cm from the dentate line (35). Evaluation of rectal biopsies may be enhanced by staining for acetylcholinesterase staining (AChE). Newly developed diagnostic kits for enzymatic-histochemistry will likely increase the use of AChE for the diagnosis of HD. The new industrial kits lyophilize the components of the medium that can be sent at room temperature anywhere in the world (Hirschsprung's disease diagnostic kit, BIOOPTICA, Milan, Italy) (25).

The main value of rectal biopsy is to evaluate the neorectum enervation status, whether it is normal or if there is a retained or acquired aganglionosis. An additional point to examine for is the presence of hypertrophied nerves at this level, suggesting the specimen is still in the transition zone and may not function normally. Moreover, IND may also be present at this margin (36,37). As many key decisions regarding a reoperation are dependent on the pathologic interpretation, the authors find reviewing these specimens in person with the pathologist quite useful.

Therefore, the choice of the best approach, either medical or surgical, should rely on the clinical presentation, examination, radiologic findings, and finally the biopsy results.

REOPERATIVE SURGERY FOR HD

Reoperations for HD encompass a broad spectrum of surgeries designed to overcome the spectrum of postoperative complications after a pull-through procedure. In this chapter, we are concerned with reoperations to deal with most common late complications after pull-through procedures.

Repeated Dilatation

Strictures and IASA are the main indications for repeated dilatation. Strictures fall into two basic types. Simple strictures are a moderate narrowing at the anastomotic site. These typically present within a few weeks after the pull-through. In general, these can be readily addressed with serial outpatient dilations, and usually do not require subsequent surgery. More persistent strictures, particularly those associated with a history of an anastomotic leak may be much more difficult to address. Strictures caused by an ischemic segment of bowel tend to be the most persistent, and often will not readily respond to dilations. Both of these latter strictures may require surgical correction. Minor stricturoplasty may address some of these strictures; however, some cases may require a redo-pull-through procedure. In those patients with a stricture located high in the rectal canal, one can generally assume that these are caused by an ischemic segment. These often do not respond well to simple or radiologic dilations. An interesting approach to handle these strictures was described by Langer (38). He used a stapling device to come across the stricture at the same time a redo-Duhamel anastomosis is created (Fig. 2).

Spasm of the anal sphincters after a successful pull-through is quite common, leading to what is sometimes called IASA. The diagnosis is established with anorectal manometry and a rectal biopsy to rule out aganglionosis (11). These children should be initially cared for with bowel management regimens including laxatives, drugs,

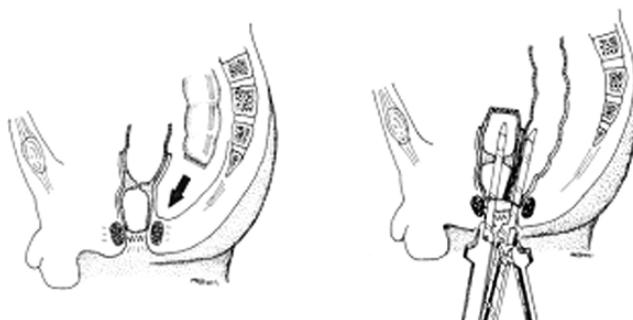


Fig. 2. Technique for using the Duhamel procedure to deal with a high rectal stricture (right panel). Reproduced with permission from (38).

and bio-feedback training. If this is unsuccessful, some advocate anal sphincterectomy (39,40). Others have had success with temporary sphincter-relaxing measures such as botulinum toxin (41,42) or nitroglycerine paste (43–45). The latter choices have some appeal because of the likelihood that the symptoms will improve significantly over time in most of these children (41). Finally, as a last solution some children may eventually need repeat pull-through for persistently severe stooling problems or intractable HAEC.

Myectomy

INDICATIONS

There are basically three indications for a myectomy. The first is the child with persistent enterocolitis who fails to respond to medical treatment. In these cases a stricture has been ruled out, although, the myectomy will often be able to address strictures at the level of the previous anastomosis. Prior to the performance of the myectomy, a biopsy should be done to rule out an extensive aganglionic segment. The second indication for a myectomy is the patient with severe constipative symptoms who has normal ganglion cells on repeat biopsy. Typically, these patients will have a dilated rectum and sigmoid colon. Caution should be taken in these children, as in some older patients this rectal dilation may not be corrected, even with a correctly performed myectomy. Such patients will require a redo-pull-through with resection of this dilated bowel segment. The third group of patients who may benefit from a myectomy after a previous pull-through are those children found to have a retained aganglionic bowel. This is the most problematic group, because if the length of aganglionosis is too long it will not be completely addressed by a simple myectomy. In general, if the length of aganglionosis is more than 5 cm, a redo-pull-through should be performed. Although one may be able to perform a myectomy longer than this, the bowel immediately proximal to this segment (i.e., transition zone) may not function perfectly well. It has been our experience that a redo-pull-through will work better in these patients (52).

Langer has advanced the use of botulinum toxin (Botox) injections into the internal anal sphincter in patients with either severe constipation or recurrent enterocolitis (46). Results have shown that this technique may produce good results in some patients with difficulty stooling or persistent enterocolitis. Ideally, a trial of Botox should be attempted prior to the use of a myectomy. The typical dose is 2–5 U/kg or up to 50 U per treatment in a toddler-sized child. This approach may help the surgeon appreciate

which patient may benefit from a myectomy. Second, there are some patients with long-lasting benefit after the use of Botox. In most patients, however, the effect of Botox is limited and does not last longer than 1–3 months. Some have questioned whether the anal dilation performed at the time of the Botox is actually the important factor, and may be why many of these children get better. Nevertheless, Minkes and Langer noted that more than 50% of patients derived benefit for between 1–6 months, and several benefited for more than 6 months (28%) (47). In general, those patients with rectosigmoid disease derived the greatest benefit.

MYECTOMY TECHNIQUE

There are two basic approaches to a myectomy. The first was described by Hugh Lynn in 1966, and is via a transanal approach (48). The advantage of this procedure is that it is easy to see landmarks, including the dentate line. Additionally, there are no external incisions, thus recovery is quite rapid. A disadvantage is that one often cannot get as high (proximal) as the posterior sagittal approach. The technique begins by creating a curvilinear mucosal/submucosal flap starting just proximal to the dentate line (1 cm or less). Dissection proceeds proximally, and is assisted by extending vertical incisions in the flap to the left and right of the midline. After creation of this flap, a full-thickness incision is made into the retrorectal space. This is then carried upward, removing a strip of muscle approximately 5 mm wide and approximately 5 cm long (Fig. 3). Dissection is carried as far proximal as possible. It is best to keep the specimen intact, as once this strip is transected, the proximal muscle will retract upward and may be quite difficult to reach. The specimen is carefully labeled for the pathologist in terms of orientation. The mucosal/submucosal flap is then closed with interrupted,

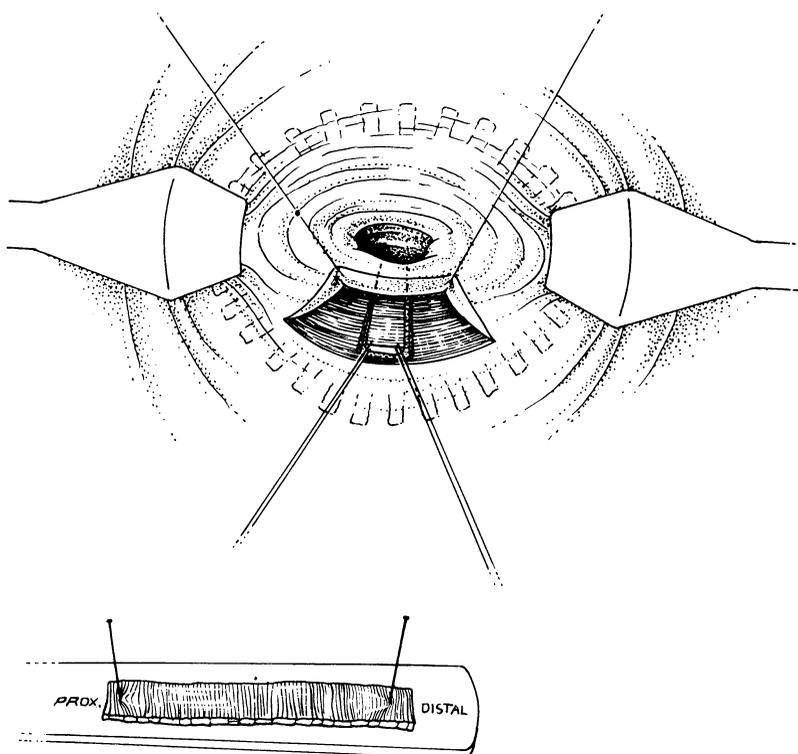


Fig. 3. Transanal approach to an anorectal myectomy (transverse incision).

absorbable sutures. One can easily palpate a “keyhole”-like defect once this flap is closed. Sometimes in postpull-through cases, it is difficult to create this mucosal flap. In this case, one can make a longitudinal elliptical mucosal incision with application of traction sutures as shown in Fig. 4A. One then performs the myectomy similar to the previously mentioned approach. The mucosal defect is then closed longitudinally without incorporating the muscularis propria (Fig. 4B). This dissection can be bloody, and one can often accomplish a fair amount of it with cautery.

The second approach is via a posterior sagittal approach. This technique was first described by Thomas et al. in 1970 to primarily treat HD patients with a short segment of aganglionosis (49). The technique requires the child to be placed in a prone jackknife position, similar to Peña’s posterior sagittal approach. The dissection begins with an incision approximately 1–2 cm behind the anal opening and extends to the coccyx. Dissection then proceeds in the midline, dividing sphincter muscles evenly. The surgeon’s finger, or a Hegar dilator, may be placed in the anal canal to insure that the rectal canal is not entered. One then incises a strip of muscularis propria, starting at the level of the internal anal sphincter and extending the dissection as far proximal as possible (Fig. 5). The strip is sent to pathology with correct orientation, and the external sphincters and levator complex are reapproximated in the midline.

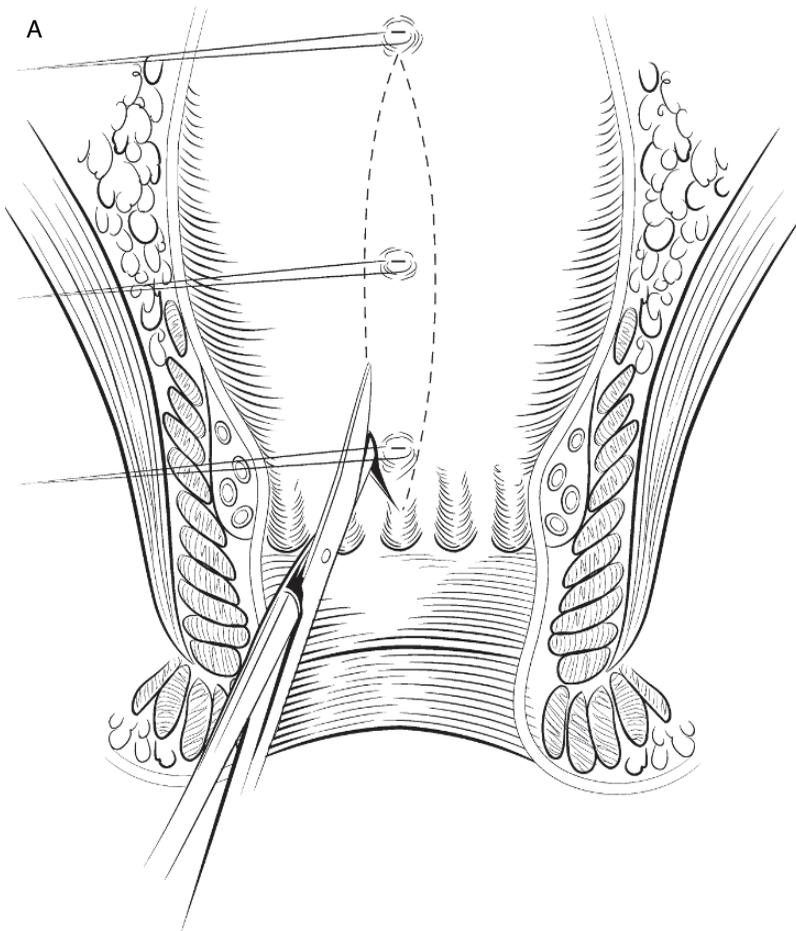


Fig. 4. Transanal approach to an anorectal myectomy (longitudinal incision).

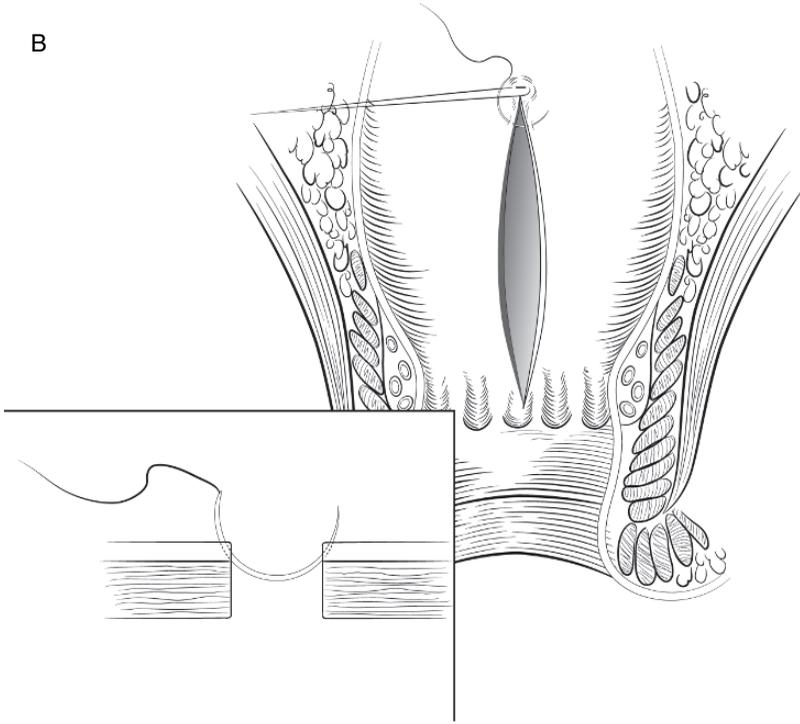


Fig. 4. (Continued).

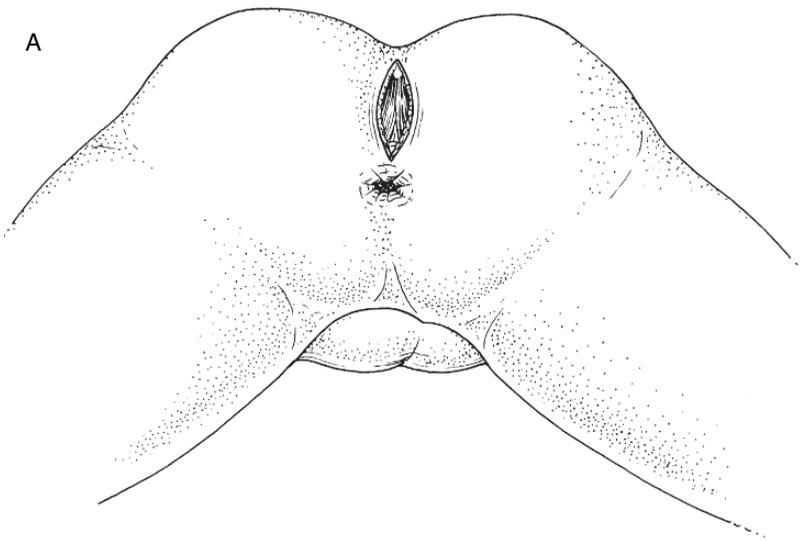


Fig. 5. Posterior sagittal approach to an anorectal myectomy.

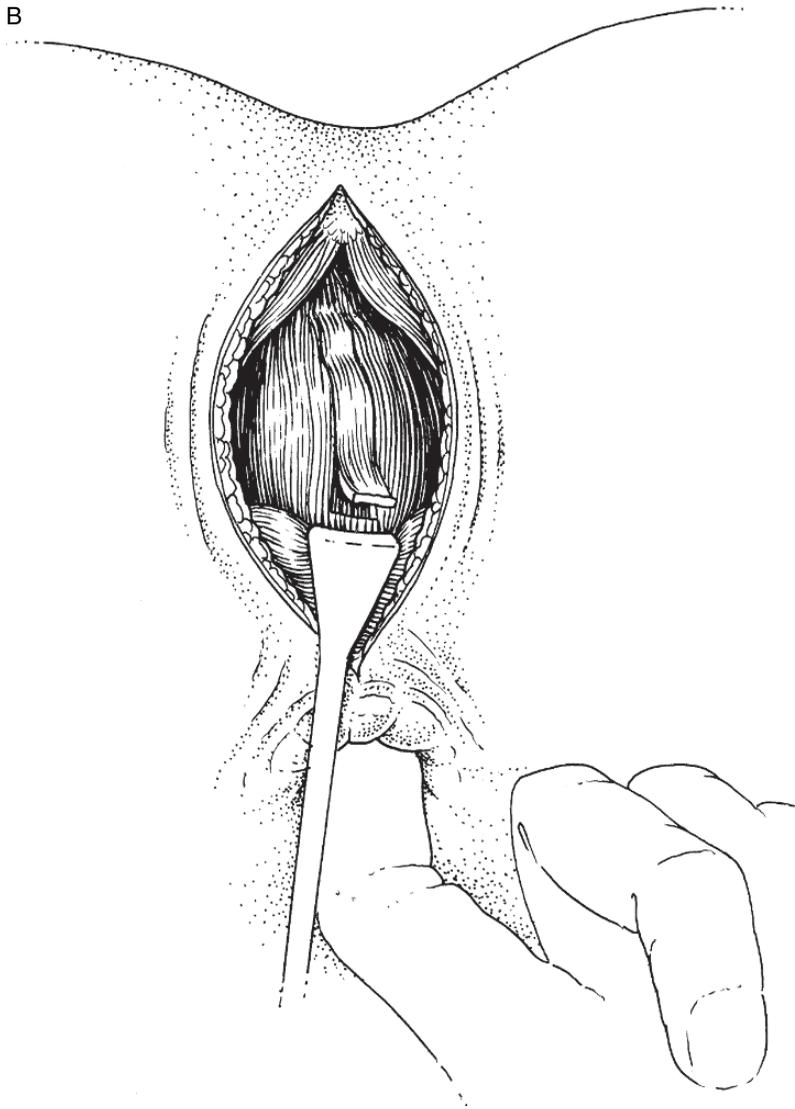


Fig. 5. (Continued).

RESULTS

Several series have described their results with both approaches. Blair et al. described 10 patients, of which nine had good outcomes (50). Kimura presented five cases that underwent a posterior sagittal approach, all of whom had excellent results. In this report, he refers to two previous patients who initially had a transanal approach and had a poor outcome, prompting a switch to this other approach (51). Moore also noted a good response in five out of five patients (26). Over the past 25 years, we have performed

39 myectomies in 34 patients with difficulty stooling or persistent enterocolitis, and 29 of these 37 myectomies were reported before (52). Complete follow-up was obtained in 32 patients (36 myectomies). Of those with pure constipative symptoms (12 with 13 myectomies), seven had aganglionosis postpull-through. Of those with recurrent HAEC (20 cases with 23 myectomies), only three had aganglionosis postpull-through. Myectomy was performed at a mean of 3.1 years postpull-through. Average follow-up was 8.6 years. Of those with chronic constipation, six of 12 (50%) had good results after myectomy with one patient requiring two myectomies. The remainder required a redo-pull-through or colostomy. Interestingly, the majority of patients with retained aganglionosis and chronic constipation did not respond to myectomy six of seven (85.7%). Of those with recurrent HAEC, 16 of 20 (80%) became free of symptoms or achieved a marked decrease in enterocolitic episodes. Four patients (seven myectomies) had a failed myectomy. In three of these cases, a repeat myectomy was performed, again without improvement in symptoms and a repeat pull-through is done. In the fourth patient, a repeat pull-through is now being attempted 1 year after the myectomy. In all patients who responded successfully to the myectomy, no incontinence has been noted.

Repeat Pull-Through Procedures

The decision to perform a repeat pull-through represents a real challenge for pediatric surgeons. It carries risks far greater than the original surgery. It should only be done by a highly experienced surgeon after following an appropriate algorithm of management. However, one should never hesitate to redo the pull-through once it is indicated after careful planning by detailed preoperative biopsies, contrast enemas, and possibly anal manometry, as detailed previously (Fig. 6).

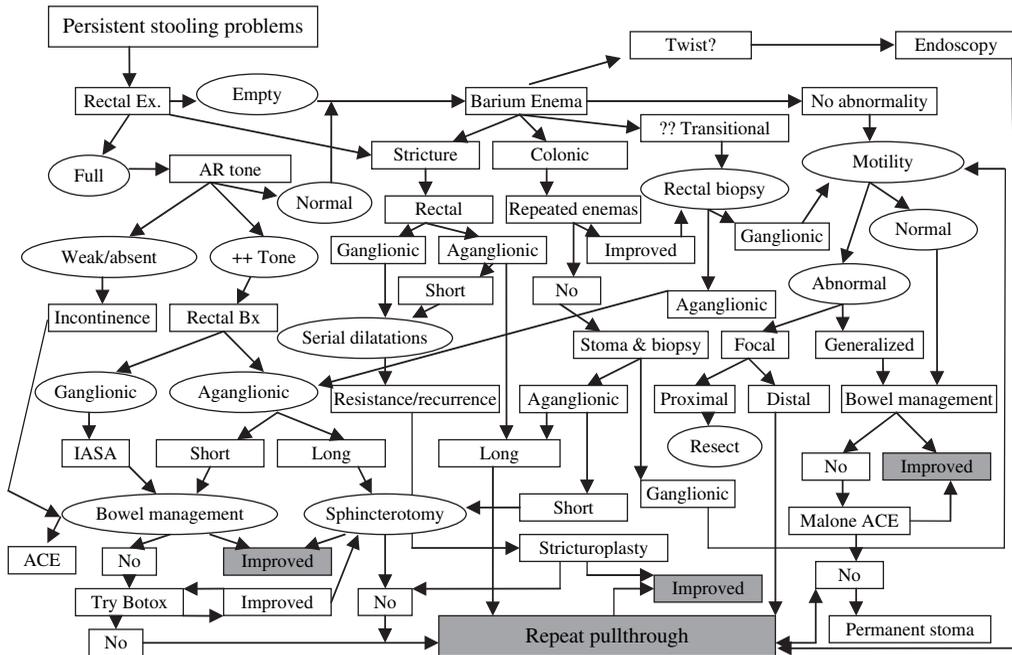


Fig. 6. Algorithm tailored for different types of long-term complications after pull-through. Botox, botulinum toxin; IASA, internal anal sphincter achaliasia; ACE, antegrade continent enema; AR, anorectal.

INDICATIONS

Patients who require a repeat pull-through include those with a retained or acquired aganglionic segment exceeding 5 cm, a transitional zone pull-through with dilated neorectum and sigmoid, associated focal IND, persistent long and/or high stricture after trial of repeated dilatations, a twisted pull-through, those who have failed to respond to a myectomy and have persistent obstructive symptoms, and children with a megacolon or rectum that is nonresponsive to bowel management regimens and myectomy. This latter group is typically seen in those who have had persistent obstructive symptoms for many years.

TECHNIQUES

The approach to performing a repeat pull-through is variable depending on what procedure(s) have been previously performed and when. In general, an open laparotomy is probably the best approach. Laparoscopic biopsies may be initially performed to gain a better idea of the abnormal pathology, but this may actually be obtained during a separate procedure. A lower midline incision is ideal, although if the child has had a previous left hockey-stick type incision, this can be extended, and is usually adequate. A transverse lower abdominal incision is a more cosmetically accepted incision and usually gives an adequate exposure. A thorough lysis of adhesions should be performed. As establishing the correct level for the pull-through segment is of high priority, a pathologist comfortable with reading frozen sections for the presence of ganglion cells and hypertrophied nerves must be available. Second, a complete mobilization of the distal colon is performed. A critical challenge at this point is to ascertain the vascularity of the distal (remaining) bowel. There is the potential for a devascularization of the distal pull-through segment if a marginal artery is transected. This may happen when the time period between the original pull-through and redo-pull-through is relatively close, thus preventing an adequate secondary revascularization.

One variant of a repeat pull-through is when the surgeon is called very shortly after the pull-through and told that the pull-through was performed at an aganglionic level (i.e., misreading of the frozen sections). In this case, it is advisable to proceed with an immediate repeat pull-through within 1 week of the first pull-through. If this approach is taken, few adhesions will be present, and one can typically revise the pull-through through the same transanal approach. One should not be dissuaded from proceeding to redo the surgery based on an initial "positive response," as most infants will start to stool spontaneously; however, obstructive symptoms will eventually return.

For those patients presenting on a more chronic basis, the decision tree in determining the approach to redo-pull-throughs is shown in Fig. 7. The decision as to which pull-through should be performed is one of judgment, and may not be readily apparent until the actual operation is performed. In general, we have always attempted to perform an endorectal pull-through (ERPT) in those patients who have had either a primary Swenson or ERPT. When this fails, because of an inability to adequately get into a submuscular plan, a Duhamel may be utilized (*see* example in Fig. 8). For those patients who underwent a primary Duhamel, it is occasionally possible to perform an ERPT, as the aganglionic and ganglionic walls tend to fuse over time. If this is not possible, a Swenson pull-through should be performed. A Swenson may be particularly challenging if one attempts to evert out the rectum, as the last 3 to 4 cm of rectum are often densely adherent within the pelvis. It may be necessary to perform the

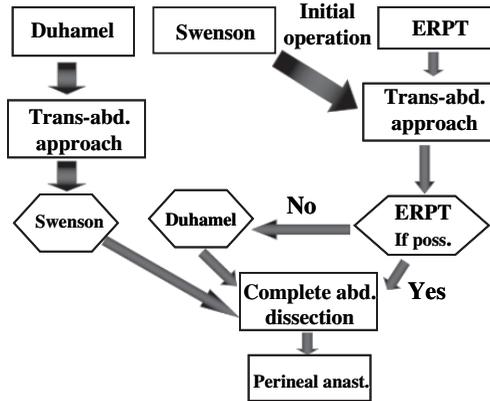


Fig. 7. Surgical approach to the redo-pull-through.

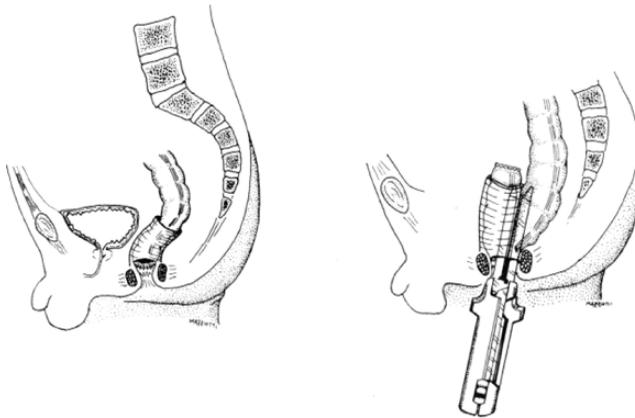


Fig. 8. Technique for a redo-Duhamel procedure. Reproduced with permission from (38).

anastomosis within the anal canal without eversion. An alternative approach is given at the end of the chapter.

At the end of the procedure, one must be sure there are no technical problems with the pull-through including a twisting, kinking, or stretching of the blood supply, or more commonly a kink of the bowel in the pelvis. This may be particularly troublesome as one tends to use more proximal segments of bowel, often proximal to the middle colic vessels. Figure 9A–C show two approaches for pulling through the right colon.

Another technical challenge that may be encountered is difficulty in performing a Duhamel procedure owing to an excess of tissue layers, which must be stapled together. Unlike a primary Duhamel, the surgical stapler is joining another full thickness of colon. In the case of an ERPT as the original procedure, this would include a new segment of ganglionic bowel, the original aganglionic muscularis propria, and the primary pull-through segment, as well as various amounts of scar tissue. One should always use extra long (4.5-mm) staples. We have encountered two cases where the staple line has fallen apart immediately after firing the stapler. In these cases, one must be prepared to perform a hand-sewn anastomosis. Alternatively, one may divert back

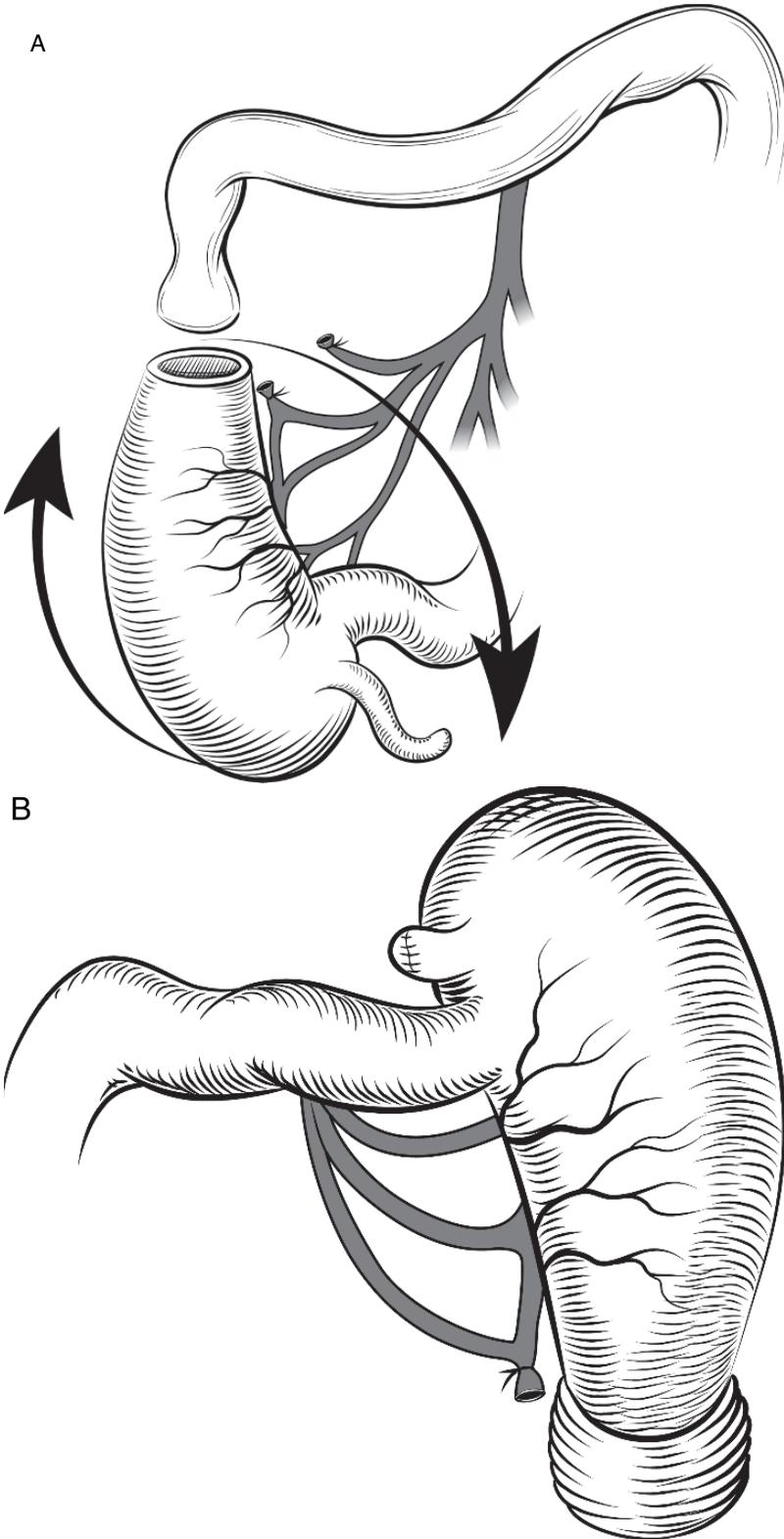


Fig. 9. (Continued).

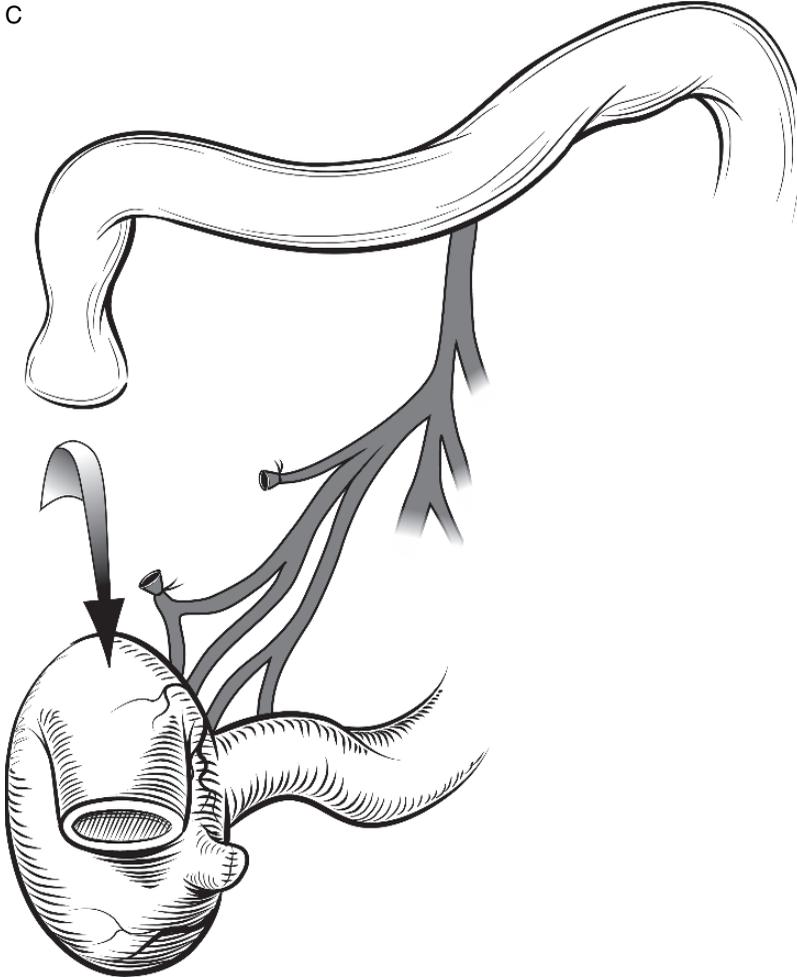


Fig. 9. Two approaches are given for performing a safe pull-through for patients who need the right colon pulled through. **(A)** The first approach is a rotation of the bowel in a clockwise direction. **(B)** This first approach results in the ileum coming into the cecum on the patient's right side, and may require a release of the child's ligament of Treitz. **(C)** An alternative approach may be to rotate the colon anteriorly.

to an original Duhamel procedure, whereby the anastomosis is created by the placement of two extra-long Kocher clamps placed in an inverted 'V' configuration, and kept in place until the crushed bowel forms an anastomosis; generally 7–10 days.

A decision to perform a protective proximal ostomy should be reserved until the end of the redo-pull-through. If the anastomosis is performed with a high degree of tension, or any question remains about the overall success, a proximal colostomy should be placed. Again, great care needs to be made when creating this diverting colostomy, as the distal blood supply in a redo-pull-through could potentially be compromised. The marginal artery could be lost either during the creation of the stoma, or when the ostomy is taken down. In such cases it may actually be safer to perform a temporary loop ileostomy.

RESULTS

Patients requiring a redo-pull-through procedure clearly represent a very diverse population, and are the most challenging group of patients with HD that a pediatric surgeon may encounter. Thus, any estimate of outcome results an individual patient will obtain is difficult. Overall, outcomes reported in the literature are reasonably good; however, it is imperative that parents be aware of the difficulty that their child faces, and realistic expectations should be given to the parents prior to the operation. Below is a brief review of the published literature on this subject as well as our own results in patients undergoing a redo-pull-through.

Wilcox and Kiely in 1998 (53) described 20 cases. The indications for reoperation were anastomotic stenosis (eight), signs of obstruction or constipation (eight), anocutaneous fistula (one), incontinence of feces (six), and enterocolitis (one); some patients had more than one indication. They opted for a modified three-stage Duhamel procedure in all but one of their patients and reported a 70% improvement in symptoms and fecal control. Sarioglu et al. in 1998 (54) reported their experience with five reoperations for similar indications. Prior to the reoperation, their patients had severe strictures in three patients with a total closure of the anastomotic site in one. One child had a rectourethral fistula, and one had an anastomotic disruption. The choice of redo-pull-through was a Duhamel in three patients and a Swenson in the other two patients. Average follow-up was 14 years, during which the patients did well; however, objective data regarding continence was not available. Weber et al. in 1999 (55) presented 38 patients undergoing operative procedures after a primary pull-through. Their data included many patients who required only myectomy (24), septum division (four), colostomy then closure for leak (four), or permanent colostomy (four); and in only five cases was a formal redo-pull-through performed. Indications included retained aganglionic segment (three), enterocolitis after failure of myectomy (one), and necrosis of pull-through (one). Revisions included a Duhamel in three patients and a Soave in two patients. Because of the diverse operations in this report it is difficult to determine exact outcomes of the redo-pull-through patients; however, excellent results were noted in 90% of all patients, though continence in the revisions was not quantified. Langer 1999 (38) reported his series of nine repeat pull-through procedures over a 4-year period for indications that included resistant stricture (three), acquired aganglionosis (five), and aganglionosis plus IND (one). Their choice of reoperation was a Duhamel in all but one child, who underwent a Swenson. Overall results were good in three patients, one had intermittent soiling and the remaining patients had persistent obstructive symptoms, which are being treated in a nonoperative fashion.

From 1974 to the present, over 386 patients with HD have been referred to our surgical group at Mott Children's Hospital. Within this group there are 37 redo-pull-throughs, 19 of which have been previously reported (56). The primary pull-through procedures for these redo cases are ERPT procedure (23 patients), Swenson's procedure (six patients), Duhamel's procedure (six patients), Rehbein's procedure (one case), and transanal ERPT in one cases. The types of cases and indications for a redo-pull-through are quite varied, and most of the children had more than one reason. By far the most common indication was a retained (or acquired) aganglionic segment (16 out of 37 cases). A group of 13 patients required repeat pull-through secondary to a mechanical problem with their primary pull-through including severe and long strictures caused by to ischemia. Another 13 patients had evidence of

residual segments of dilated colon leading to functional failure of their initial operation. Of this latter group there were eight patients with documented aganglionic bowel present at the second pull-through, despite findings of normal ganglion cells during the first biopsy in six of these, and a misreading of the pathology in two patients. The remaining five patients had normal ganglion cells on repeat biopsy. However, despite the normal pathology, all had severe difficulty passing stool, and none had evidence of strictures. Each of these latter patients had an overlying distended rectosigmoid colon and presumed internal sphincter achalasia. Of these only one patient had pathologic evidence of IND. Other indications for repeat pull-through included retained septum (two cases) and fecoloma (two cases) after Duhamel's procedure, 360° twist of the pull-through colon causing obstruction (two cases), retraction of the neorectum (one case), leak (two cases), recurrent abscess and persistent rectocutaneous fistulae (two cases), and transitional zone pull-through (one case) established by documentation of presence of ganglion cells and hypertrophy of the nerve fibers. The presenting symptoms included chronic constipation, repeated and treatment-resistant HAEC episodes, and sometimes the local cause like leak, abscess, or fistulae.

Distinct from other reports, we found that a redo-ERPT could be effectively performed in several of these patients, and yielded excellent results. An ERPT was performed in 21 patients in whom an adequate endorectal dissection could be developed between the native rectal cuff and the previously pulled-through colon. In one of these, the redo-pull-through was performed via a transanal approach. Additional redo procedures included a Duhamel in nine patients and a Swenson in four patients. Our group is also reporting a new technique for the performance of a repeat pull-through using a stapled anastomosis in three cases (*see* next section). One technical challenge is that many redo-pull-throughs require the colon proximal to the middle colic vessel to be pulled through. Twisting of the colon can be encountered more frequently with this type of pull-through. This technical point was initially described by Duhamel; and Fig. 9 illustrates two potential approaches that can be used to perform a pull-through with the right colon.

Follow-up ranges from 6 months to 32 years (mean 14.2 years). There were no deaths in the series and one patient required a third pull-through. An assessment of continence was done in all patients who are not neurologically impaired and were over age 3 years of age. Of these patients all are continent except four children, yielding a continence rate of 89.2%. Two of these patients have minor soiling at night and during activities only, and the other two have daily leakage of stool. One child who is mildly mentally retarded has intermittent accidents. Additionally, one patient developed recurrent and antibiotic-resistant perirectal abscesses, which finally led to chronic sacral osteomyelitis. A permanent stoma was the only solution to resolve this issue. Another patient had leakage from the coloanal anastomosis after closure of the colostomy, and so required a diverting ileostomy. He is now stable and awaiting closure of his ostomy. One patient suffered from constipation and needed Botox injections into the anal sphincters, and subsequently underwent a sphincterotomy after the repeat pull-through. Two patients required repeated dilatations for strictures that developed after the redo procedure; both of these children responded well to the frequent dilatations without further need for another procedure. One final patient developed a cuff abscess that responded to medical treatment and ultrasound-guided drainage. Stool frequency per day range from 1 to 10 (mean 3.2).

REPEAT PULL-THROUGH WITH STAPLED ANASTOMOSIS

We have recently performed a new approach to a repeat pull-through using a stapled anastomosis. The patient is positioned in the lithotomy position. The abdomen is entered via a low transverse incision or left Paramedian incision. A thorough lysis of adhesions and mobilization of the colon is performed, and a series of seromuscular biopsies are sent for frozen section to assess for the correct level of normal ganglionated bowel. The colon proximal to this level is then divided with a gastrointestinal anastomosis (GIA) stapler. One of the greatest difficulties with a repeat pull-through is the last 2–3 cm of rectum. This area is typically quite scarred, and any type of mobilization outside of the rectum is difficult. Therefore, a dissection of the rectum is performed analogous to a Swenson procedure. Dissection is carried down into the pelvis to within 1–1.5 cm above the dentate line. Great care is taken to avoid injury of the pelvic structures by meticulous dissection, perfect hemostasis, and by sticking to the colonic wall. Once the sigmoid colon is adequately mobilized, attention is drawn to the neorectum. A Kelly clamp is then introduced from the anus to the neorectum, which had been mobilized circumferentially, and the colonic stump is everted out of the anus. A TA stapler (or in some cases an EndoGIA stapler) is used to resect the bulk of the neorectum outside the anus in its everted state as close as possible to the dentate line (Fig. 10). The neo-rectum is then reinserted into the pelvis. The end result is approximately a 2.5-cm length of residual rectum. The staple line of the proximal ganglionated colon is then removed, and a purse-string suture is used to secure the anvil of the end-to-end anastomosis (EEA) stapler in place. A 21-mm (or larger) EEA stapler is

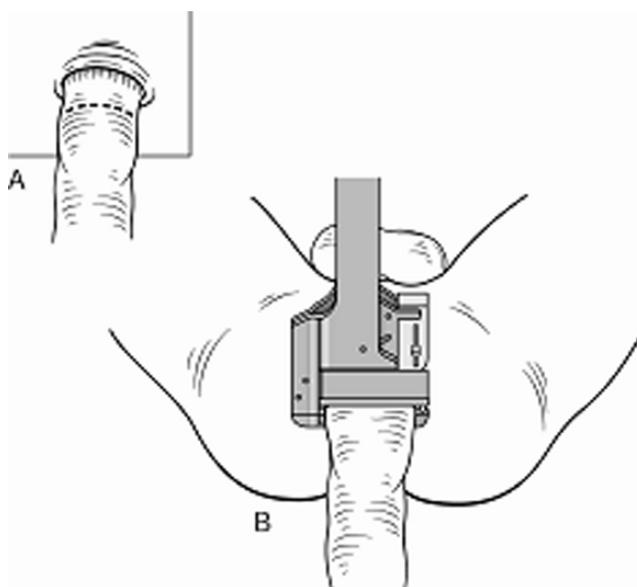


Fig. 10. Approach for performing a repeat pull-through analogous to a Swenson approach, but using a stapled anastomosis. **(A)** Mobilization is done internally until the surgeon encounters significantly adherent bowel in the most distal rectum. **(B)** The rectum is then everted out of the peritoneal cavity, without a complete dissection of the final 3–4 cm, which remain adherent. **(C)** Because the most distal few centimeters of the rectum are tightly adherent and do not allow eversion, a stapled anastomosis is performed using an EEA stapling device within the pelvis. **(D)** Final appearance of the pull-through.

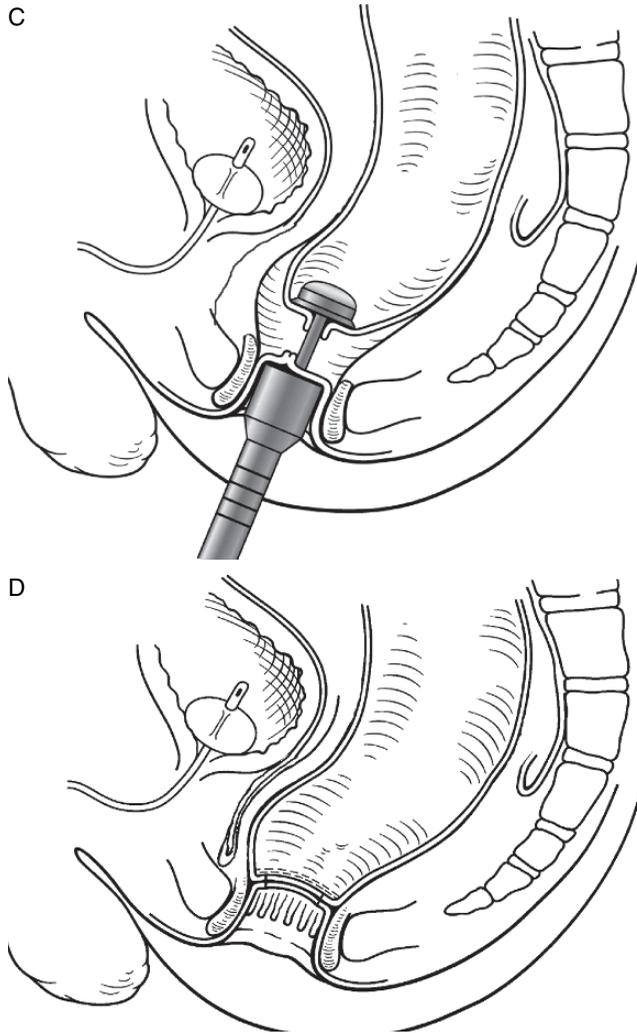


Fig. 10. (Continued).

then inserted into the rectal remnant. An EEA-stapled anastomosis is then performed (Fig. 10C and D). The anastomosis is checked for a leak by verifying complete donuts that contained both mucosa and wall on both sides. In addition, air is injected with high pressure into the rectum, air bubbles should not be noted when the pelvis is filled with saline. The anastomosis should not be under tension. The pelvis is then irrigated with copious amounts of saline and the abdomen is closed in a regular fashion. A protective colostomy or very often an ileostomy should be strongly considered after a repeat pull-through.

Results with the new technique

We have performed this EEA-stapled procedure for three patients. The indications were persistent constipation (two) and intractable HAEC (three). Two of the three patients had a retained aganglionic segment, one of them had a severe stricture that was nonresponsive to serial dilatation and sphincterotomy. The third case had a transitional

zone pull-through with presence of ganglia and hypertrophied nerve fibers in the biopsy. One patient underwent two previous unsuccessful pull-throughs. The ages of the patients were 4 years (two) and 8 years (one). The original operation was Soave's (one), Swenson's (one), and Duhamel's (one). Follow-up period has ranged from 6–9 months. Two out of the three patients are doing excellent with 2–5 daily bowel movements. These two are without soiling and are completely continent. The third, an 8-year-old boy, suffered from anastomotic leak and was diverted by ileostomy. He is now waiting for a reclosure of his colostomy. In general, we are optimistic with the new approach and predict that it will be of great value in redo-pull-through cases where a more simplified endorectal dissection cannot be performed.

Permanent Stoma

There will be a group of patients that may fail a repeat pull-through, or the child's parents and physician may decide that they may not benefit from a repeat pull-through. In these cases, one should consider a permanent stoma. Assistance from an enteral stomal therapist may help ensure that this stoma is placed in the most satisfactory location for the patient's entire life.

CONCLUSIONS

Reoperations for HD clearly require a great deal of planning and a thoughtful work-up. In cases where a reoperation is needed, however, reasonably good results may be found in the majority of cases. An ERPT is a good procedure to select for a repeat pull-through. However, the pediatric surgeon performing such a procedure must be experienced with a number of different approaches, as not uncommonly other techniques must be applied.

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18

Reoperative Surgery for Anorectal Malformations

Marc A. Levitt, MD and Alberto Peña, MD

CONTENTS

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DEFINITIVE REPAIR
OTHER UROLOGIC INJURIES
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Despite significant technical advances in the surgical repair of anorectal malformations, patients sometimes require reoperations when there has been an inadequate preoperative evaluation, in order to repair technical problems that occurred during the definitive repair, and when pitfalls in the postoperative management, both in the short and long term, are encountered. Complications are not isolated to patients with complex malformations, but include the entire spectrum of malformations, and in fact are more common in the but relatively benign malformations.

PREOPERATIVE EVALUATION AND MANAGEMENT

The preoperative evaluation may begin with a prenatal diagnosis, which is an area of rapidly improving technology. Accurate prenatal diagnosis may impact perinatal decisions and location for a baby's delivery. This is particularly important for babies with cloacal malformations and cloacal exstrophy. Most often, an anorectal malformation is not diagnosed prenatally. Once the baby is born, a protocol of evaluation is vital, including both clinical and radiologic assessment (1).

The correct newborn operation, either a newborn pull-through or a colostomy, must be chosen. Future reoperations caused by decisions made in the first 24 hours of life are common. In patients with cloacal malformations, there are unique problems that can occur with improper newborn treatment. In such patients, the clinician must recognize and manage hydrocolpos, perform a colostomy with avoidance of key pitfalls, and make a correct clinical diagnosis (2).

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Fig. 1. Radiograph showing hydrocolpos.

Failure to treat a hydrocolpos (Fig. 1) can result in obstruction of the urinary tract, prompting an unneeded urinary diversion, or lead to pyocolpos. Several techniques to drain a hydrocolpos can lead to difficulties. Catheterization of the common channel is unreliable, as it may inadequately drain the vagina or vaginas if the catheter passes into the bladder. A plasty of the single perineal orifice may provide inadequate drainage, particularly in cases of a long common channel. Dilatation of the common channel likewise may inadequately drain the dilated structures. A tube vaginostomy is the most reliable technique and should be performed at the time of colostomy creation if a hydrocolpos is present. The surgeon must also investigate whether there are two dilated hemivaginas, and drain both of them, either with separate tubes, or with one tube and creation of a window in the septum between the two vaginas.

If an infant with a cloaca is not growing well in the first weeks or months of life after the colostomy, it is most likely because a complicating problem has been missed. If the hydrocolpos is left undrained, patients may suffer from recurrent urinary tract infections, persistent acidosis, and failure to thrive. Occasionally, reoperation is required to properly drain a hydrocolpos, or to revise a colostomy, particularly if the stoma is not completely diverting. Failure to identify associated urologic problems such as vesicoureteral reflux may also be the cause of the baby's problems.

In addition to management of a hydrocolpos, the surgeon must be thinking about the patient's gynecologic structures in order to prevent future problems (3,4). During the colostomy creation, definitive repair if a laparotomy is required, or colostomy closure, identifying and inspecting the gynecologic structures is vital. Patients may have many variations of Mullerian structure atresias or stenoses that can

lead to future obstruction of menstrual flow, amenorrhea, and may affect their ability to conceive and carry a baby to term.

The diagnosis of most anorectal malformations is a clinical one. Once the perineum is inspected and the malformation identified, the correct newborn surgical intervention can be planned. For patients with evidence of rectoperineal fistula, a newborn or delayed pull-through can be performed without a colostomy. For infants with evidence of a rectourethral fistula (the most common anomaly in males) a colostomy is the recommended first step, prior to a future definitive reconstruction.

A single perineal orifice defines a cloaca, and if misdiagnosed, may lead to an unnecessary endocrinologic evaluation for “intersex.” The hypertrophied clitoris, which is sometime visible in cloacas, can be confounding to a clinician not familiar with the external perineal appearance of a cloacal malformation. If the surgeon fails to make the correct diagnosis of a cloaca, they may be prompted to mobilize the rectum, and leave the urogenital sinus intact (Fig. 2). In such a case, a reoperation is required using a posterior sagittal approach, with rectal mobilization, and correction of the persistent urogenital sinus.

The colostomy is an essential component of the newborn management of the majority of patients with anorectal malformations, and is a procedure with many preventable complications (5). A colostomy placed too distally in the sigmoid can interfere with the future pull-through (Fig. 3). Colostomy prolapse is a significant problem, and occurs when the colostomy is created in a mobile portion of the colon (Fig. 4), or if a loop type of colostomy is used. Colostomy prolapse is particularly problematic for patients with anorectal malformation who cannot afford to lose colon, as their capacity to form solid stool impacts their potential for fecal continence. If they are incontinent, they are more manageable with constipation, than with a tendency toward loose stools. Loop colostomies can lead to urinary tract infections from incomplete diversion of the fecal stream (Fig. 5) and fecal impaction in the distal colon. A Hartman procedure is

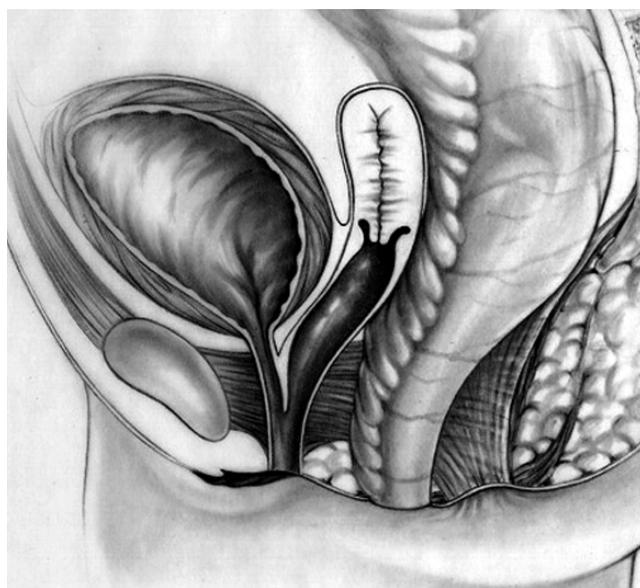


Fig. 2. Pull-through only of rectum in a patient with a cloacal malformation, leaving the urogenital sinus untouched.

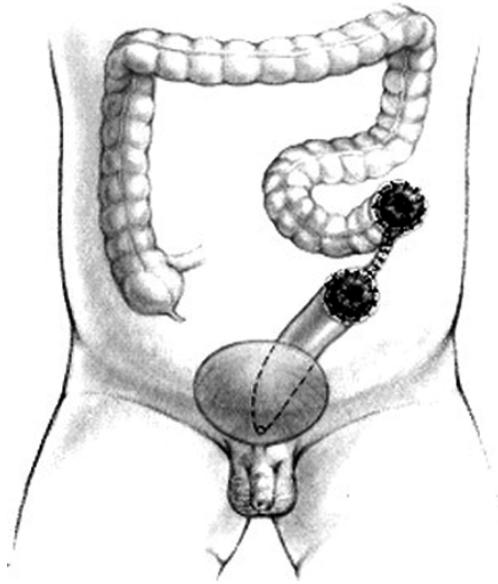


Fig. 3. Colostomy created too distal in the sigmoid, which will interfere with the pull-through.

problematic in patients with anorectal malformations, as there is no access for contrast studies to define the distal rectal anatomy, and these types of colostomies can lead to the development of mucocoeles. Many of these colostomy problems require reoperations (i.e., a colostomy revision).

In a patient with an anorectal malformation, it is recommended that the stomas be separated enough to allow the stoma bag to cover only the proximal stoma, isolating the mucous fistula to prevent contamination. The stoma should not be separated more than necessary, so that a big laparotomy can be avoided at the time of colostomy closure. The opening of a sigmoidostomy in the right upper quadrant can occur if

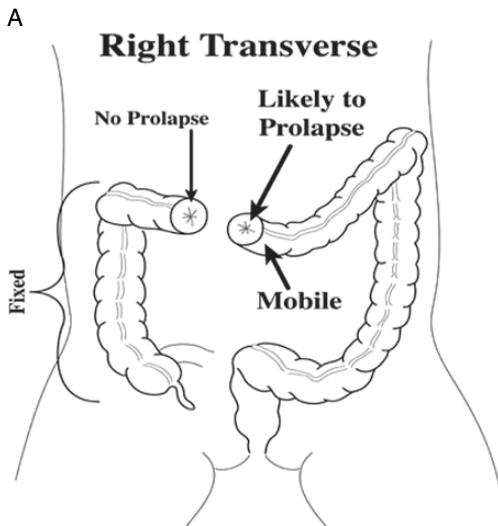


Fig. 4. (Continued).

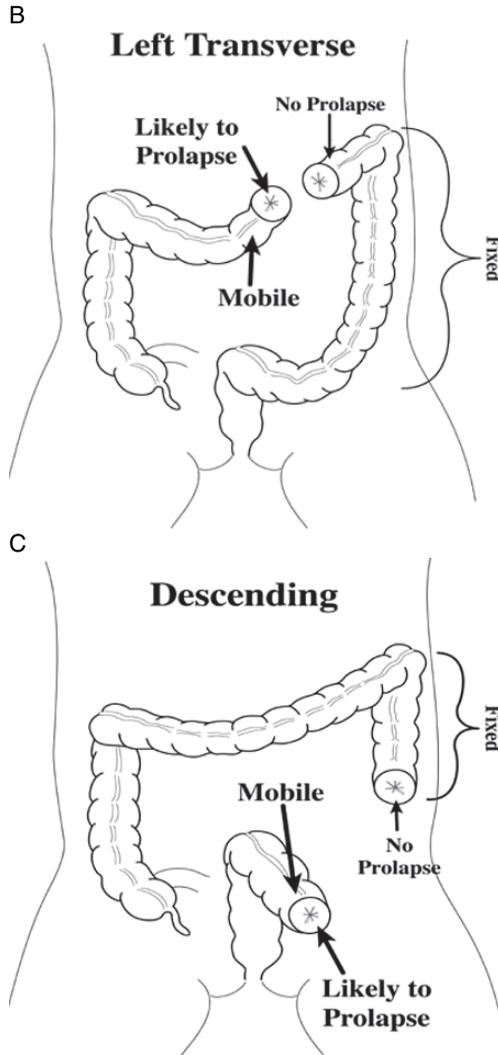


Fig. 4. Colostomy prolapse can occur when the colostomy is placed in a mobile portion of the colon. (A) Right transverse; (B) left transverse; (C) descending.

the surgeon tries to find the transverse colon via a right upper quadrant incision and instead mistakes a dilated rectosigmoid for the transverse colon.

Patients with right transverse colostomies, especially in cases with a long time interval between the opening of the colostomy and the main repair, develop a characteristic microcolon and a more distal megarectosigmoid (Fig. 6). With transverse colostomies, it is also almost impossible to irrigate and clean the distal colon adequately. Also, the distal colostogram is difficult to do, as significantly high hydrostatic pressure is needed to show the precise anatomy. Finally, a longer colonic segment can allow for more resorption of urine and lead to acidosis.

Stricture of the colostomy is caused by a technical problem in which the bowel suffered from ischemia from an inadequate manipulation of the colon’s blood supply. Dehiscences, infections, and sepsis after colostomy closure are also related to technical problems.

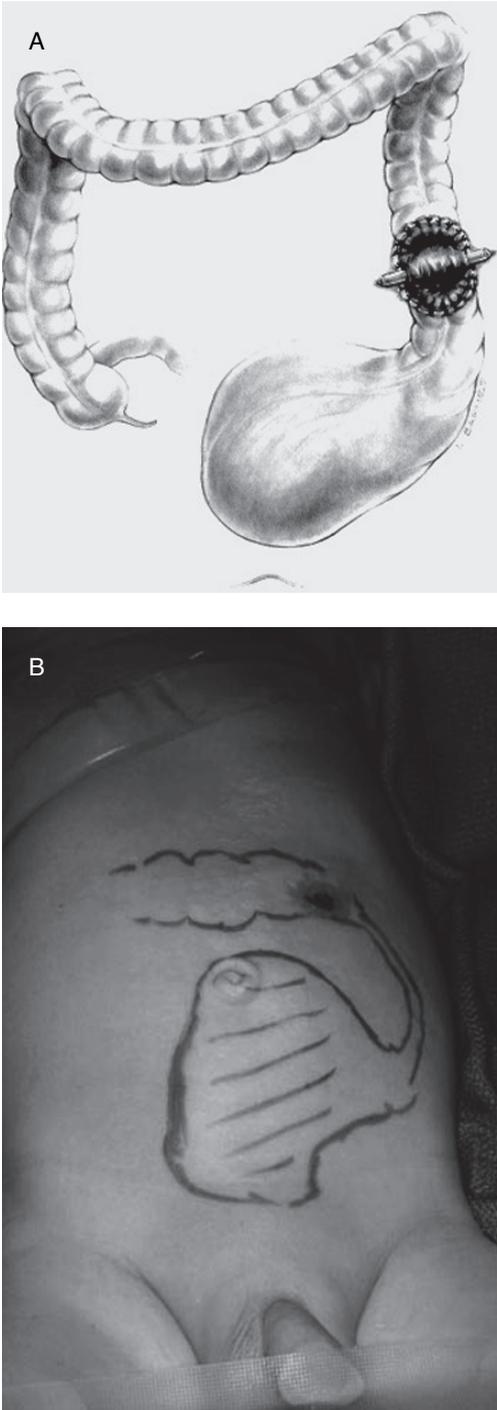


Fig. 5. (A) Diagram of loop colostomy. (B) Picture of loop colostomy.



Fig. 6. Transverse colostomy with distal microcolon and megarectosigmoid.

To avoid complications, the preferred colostomy seems to be one with separated stomas, with the proximal end located just after the sigmoid comes off its left retroperitoneal attachment (5). The stomas should be separated enough to allow for the placement of a colostomy bag on the proximal stoma (Fig. 7). The proximal stoma must be located away from both the umbilicus and from the iliac crest, thus surrounded by a good portion of normal skin, so that the stoma bag can be easily adapted. The mucous fistula should be located medially and lower, and should be fashioned tiny and flat to avoid prolapse, because it is only used for irrigations and diagnostic tests.

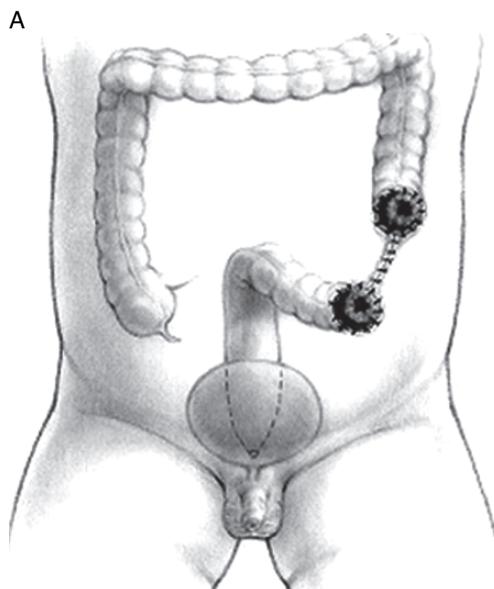


Fig. 7. (Continued).



Fig. 7. (A) Diagram of ideal colostomy. (B) Picture of ideal colostomy.

The purpose of the colostomy, in addition to diverting the fecal stream, is to perform a distal colostogram to define the colorectal anatomy. Understanding the precise location of the distal rectum, and its relationship to the urinary tract, is vital to avoiding complications during the definitive repair (6).

DEFINITIVE REPAIR

The definitive repair has numerous complications associated with it, many of which are preventable with correct preoperative planning and proper technique. Reoperative surgery is necessary for several of these complications, to alleviate pain, discomfort, and other sequelae.

It is clear that a patient's best chance for a good functional result is when the proper operation is performed during the first definitive procedure and complications are avoided (7). This is especially true in those patients born with a good prognosis defect. It is unfortunate when such patients end up with fecal or urinary incontinence resulting from avoidable complications of the surgical repair.

Rectal Problems

Patients can experience dehiscence, retraction, infection, and/or acquired atresia of the rectum related to technical problems during the pull-through. These are usually the result of excessive tension or inadequate blood supply. In addition, anal strictures may result when the prescribed protocol of dilatations is not followed.

Reoperation for these problems can be performed posterior sagittally. In cases of retraction, dehiscence, and acquired atresia, the rectum is usually located somewhere high in the pelvis surrounded by a significant amount of fibrous tissue. Multiple fine silk sutures are placed in the rectal wall in order to exert uniform traction and facilitate a circumferential dissection of the rectum, trying to stay as close as possible to the rectal wall without injuring it. Bands and extrinsic vessels surrounding the rectum are divided and cauterized circumferentially until enough rectal length is gained to place the rectum within the limits of the sphincter mechanism. Short ring-like

rectal strictures can be treated with a Heineke-Mikulicz type of plasty. Strictures that are longer than 1 cm must be resected, with the rectum mobilized until the fibrotic portion can be removed, and a fresh nonscarred portion of rectum pulled down, creating a new anus.

Retraction, dehiscence, and acquired rectal atresias, are most likely caused by inadequate mobilization of the rectum. During a primary procedure, the rectum, when seen posteriorly sagittally, is covered by a very characteristic white fascia that contains vessels to the rectum. The surgeon must dissect this fascia off the rectum, remaining as close as possible to the rectal wall. Uniform traction provided by multiple fine silk sutures is imperative to facilitate this dissection. The intramural blood supply of the rectum is excellent, and the rectum can be dissected to gain significant length provided the rectal wall is not injured. The most likely cause for difficulty in dissection of the rectum is working outside of this fascia. Alternatively, dissection too close to the rectum can injure the rectal wall, interfere with the intramural blood supply, and provoke ischemia. The result of all this is an incomplete mobilization, rectal ischemia, and a rectum to skin anastomosis performed under tension, which may explain many of these complications.

Strictures are also most likely caused by ischemia of the distal rectum. When the rectum is correctly mobilized and the blood supply kept intact, it is extremely unlikely to have an anal stricture. Patients who fail to follow a protocol of dilatations can develop a thin fibrotic ring in the area of the anoplasty, which is easily treated either with an anoplasty or dilatations. A long narrow stricture is most likely caused by rectal ischemia.

Some surgeons do not have their patients follow a protocol of anal dilatations. In order to avoid painful maneuvers to the patient, they follow a specific plan consisting of taking the patient to the operating room every week and under anesthesia performing dilatations. Those forceful dilatations can actually provoke lacerations in the anal verge, which then heal with scarring, only to be reopened during the next forceful dilatation, leading ultimately to an intractable ring of fibrosis.

Rectourinary and Rectovaginal Fistulae

Patients may have various types of complications involving rectogenitourinary tract fistulae. Fistulae can be persistent when the original rectourethral fistula remains untouched during the main repair, even when the rectum was repaired. Recurrent fistulae may occur if the surgeon repaired the fistula but it reopened. Acquired rectourethral fistulae are those that are created during the repair of a benign malformation (6).

Acquired rectovaginal fistula can occur during an attempted failed repair of a rectovestibular fistula. Prior to the introduction of the total urogenital mobilization, urethrovaginal fistula was the most common and feared complication in cloacal malformations (8). In certain circumstances, the vagina can be rotated to try to prevent this complication. But even with that maneuver, these fistulas can occur.

Persistent rectourethral fistulae can occur in patients who were born with a rectourethral bulbar fistula and underwent a repair that did not address the fistula. Surgeons following the old diagnostic approach may have performed an invertogram and found the bubble of rectal air close to the skin. This may have led to an approach through the perineum, with identification of the rectum and subsequent pull-through

and anoplasty. Because the surgeon was completely unaware of a low rectourethral bulbar fistula, it was not repaired.

Recurrent rectourethral fistulae may result if the fistula is closed, but the rectum is not mobilized adequately, leaving the anterior wall under tension. A dehiscence of the anterior rectal wall may explain the recurrence of the fistula. Also, leaving sutures in the rectum adjacent to the sutures in the urethra may lead to formation of a recurrent fistula. Thus, any rectal tapering should always be performed on the posterior wall. Consequently, an injured rectum that requires a repair to the anterior wall may create a situation for the development of a recurrent fistula. The same explanation may apply to rectovaginal fistulae.

Acquired rectourethral fistulae can occur in male patients who were born with rectoperineal fistulae if they undergo their first operation without a Foley catheter in the urethra. During the mobilization of the anterior wall of the rectum, an unrecognized urethral injury can occur and, if the rectum is not adequately mobilized to leave normal rectal wall in front of the urethral injury, an acquired rectourethral fistula will form.

Fistulous complications can be approached posterior sagittally. The posterior rectal wall should be opened and the fistulae identified and closed. The rectum then needs to be separated from the urinary tract or the vagina, and mobilized to ensure that a completely normal anterior rectal wall is left in front of the urethral or vaginal suture line.

Persistent Urogenital Sinus

A persistent urogenital sinus occurs in patients born with a cloacal malformation who underwent an operation in which the rectal component of the malformation was repaired, but the urogenital sinus was ignored (Fig. 2). Their reoperation can also be approached posterior sagittally. The rectum must be completely dissected and reflected out of the way. This allows exposure of the urogenital sinus, which can be repaired using the same technique that is employed during the treatment of a cloaca (9). Many such patients with this problem had an original diagnosis of a "rectovaginal fistula" (10) and the true malformation, a cloaca, was not fully recognized.

Acquired Vaginal Atresia

Complete fibrosis of the vagina caused by excessive dissection can occur when a high vagina is mobilized. This is particularly problematic when the vagina is separated from the urethra during the repair of a cloacal malformation. The dissection of the vagina from the urinary tract is not an easy maneuver, the vagina may become devascularized and as a consequence, patients can develop vaginal ischemia and subsequent atresia. This is why the development of the total urogenital mobilization has dramatically reduced this complication (8).

Acquired Urethral Atresia

Acquired urethral atresia in patients with cloaca can occur from devascularization of the pulled through or reconstructed neourethra. In male patients, it can occur when the urethra is accidentally transected during an attempt to repair an anorectal malformation. This complication most frequently occurs in males in whom the surgeon approaches the reconstruction posterior sagittally without a preoperative high-pressure distal colostogram (6). To repair this problem, the rectum must be mobilized in order to expose the urethral area. Both urethral ends need to be identified, dissected, and mobilized to perform a tension-free end-to-end anastomosis.

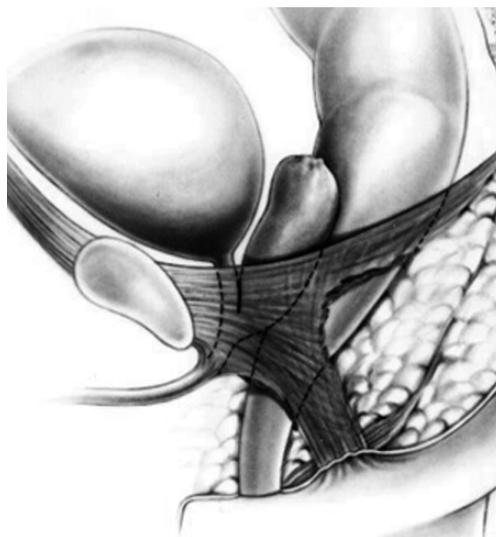


Fig. 8. Posterior urethral diverticulum, representing the undissected former distal rectum.

Posterior Urethral Diverticulum

This complication can occur when a retained portion of the rectum is left attached to the posterior urethra (Fig. 8) and is seen in patients born with a rectourethral bulbar fistula repaired transabdominally. It is easy to understand that the surgeon was unable to reach the fistula site through the abdomen. Consequently, the rectum was amputated, leaving a piece of rectum attached to the urethra. This potential complication is also possible with a laparoscopic approach, particularly if the rectum reaches well below the peritoneal reflection.

These patients are initially asymptomatic, but after several years develop symptoms such as passage of mucous through the urethra, dribbling, orchioepididimitis, urinary tract infections, and urinary incontinence. In addition, we have seen one such patient who, after thirty years, developed an adenocarcinoma in the piece of rectum left attached to the urethra.

A pelvic magnetic resonance imaging (MRI) can diagnose this condition, with visualization of a urine-filled diverticulum behind and connected to the urethra. A posterior sagittal approach can be used to fix this problem, with mobilization of the rectum to expose the posterior aspect of the urinary tract, identification of the diverticulum, and dissection of it down to the urethra. The diverticulum can be separated from the urethra in the same manner as in a primary repair of anorectal anomalies, with the urethra closed and the diverticulum resected.

OTHER UROLOGIC INJURIES

Significant urologic injuries such as transection of the bladder neck, transection of the urethra, injury to vas deferens, seminal vesicles, prostate, and ectopic ureters have occurred, usually when the posterior sagittal approach was performed without a good previous distal colostogram, and thus the precise anatomy was not known prior to the posterior sagittal dissection (6). These injuries often require reoperation.

Neurogenic Bladder

Neurogenic bladder in male patients with anorectal malformations is extremely unusual. In our series, we have only seen it in patients with a very abnormal sacrum or an associated spinal anomaly (11). If it does occur, it may represent a denervation of the bladder and bladder neck during the repair.

Patients with cloacal malformations are very different with regard to bladder function. They often have a deficient emptying mechanism and do not have the typical “christmas tree” type of image of a neurogenic bladder seen in patients with spina bifida, but rather have a flaccid, smooth, large bladder that does not empty completely. Fortunately, most patients with cloacas have a very good bladder neck. The combination of a good bladder neck with a floppy, flaccid bladder make these patients ideal candidates for intermittent catheterization, which keeps them completely dry (9).

Two exceptions to this rule in cloacas exist. One is represented by patients that have a very long common channel, in which the hemivaginas as well as the rectum are attached to the bladder neck. After they are separated, the patients are left with no bladder neck or a very damaged bladder neck. The second group is represented by a small number of patients with cloaca who are born with separated pubic bones, who could be described as having a “covered exstrophy” (12). These patients were born with no bladder neck, and they eventually require a continent diversion type of operation.

Complications of the Laparoscopic Approach to Anorectal Malformations

Long-term results and complications have not yet been described for the laparoscopic approach to imperforate anus, which is a relatively new approach, developed to avoid a laparotomy and to minimize the posterior sagittal incision (13). We have reoperated for several complications from this approach performed at other institutions, and postulate the potential for others.

Avoidance of the perineal exposure afforded by the posterior sagittal approach can lead to inadvertent injuries, such as injury to the bladder neck, urethra, or to an ectopic ureter particularly during passage of the trocar through the perineum. Precise understanding of the anatomic relationships of the pelvis provided by the laparoscopic view is vital to avoid these problems. As with the former transabdominal approach (14), there is potential for leaving behind the distal rectal cuff and thus a posterior urethral diverticulum, particularly for malformations below the peritoneal reflection such as a rectobulbar fistula. Finally, to avoid rectal prolapse a pelvic hitch is employed (13). If this step is omitted, or done incorrectly, the incidence of prolapse may increase. With the avoidance of the posterior sagittal incision, the described laparoscopic operation omits several key steps of the posterior sagittal anorectoplasty (PSARP) that are important to avoid prolapse (15), particularly tacking of the posterior rectal wall to the muscle complex.

Rectal Prolapse

Rectal mucosal prolapse occurs following PSARP, with an incidence of 3% (15). It is more common in patients with higher malformations and with poor sacra and pelvic musculature. Significant prolapse may lead to ulceration, bleeding, and mucous production. It can interfere with anal canal sensation and thus impact a patient’s functional prognosis.

Correction of prolapse can be performed transanally, with mobilization of redundant full-thickness rectum, and redo-anoplasty. This is ideally done prior to colostomy closure. Sometimes, however, prolapse only develops after colostomy closure and in the presence of constipation.

POSTOPERATIVE MANAGEMENT

The long-term concerns following surgery for anorectal malformations mainly involve a child's potential for bowel control. Most children have voluntary bowel movements, but a small number suffer from fecal incontinence despite a technically correct operation (11). Fecal incontinence may occur as a result of a technically incorrect operation. Such a patient must be identified, and may be a candidate for a reoperation particularly for a patient who be expected to have good bowel control. Constipation is a common problem following operations for anorectal malformations, and may lead to significant sequelae if not proactively managed.

Fecal Incontinence

Reoperation to improve a patient's functional prognosis is indicated in several circumstances. Early on reoperative surgery was performed on every patient we evaluated who underwent a repair at another institution and subsequently developed fecal incontinence. During those years, we hoped that the new posterior sagittal approach would give these patients an opportunity to recover bowel control. When the results were evaluated (16,17), only 30% of those patients experienced a significant improvement. Therefore, the indications for reoperative surgery were modified.

Presently, reoperation for fecal incontinence is recommended only for patients with very special criteria. It is for patients born with a malformation associated with a good prognosis, with a mislocated rectum. The rectosigmoid should be present, the sacrum normal, and the sphincter mechanism intact. In such cases, restoration of near normal anatomy may improve the functional outcome.

For these reoperations, the rectum is approached posteriorly. Multiple silk stitches are placed at the mucocutaneous margin in order to apply uniform traction to facilitate the dissection and mobilization of the rectum. A full rectal dissection and mobilization is performed, staying as close as possible to the bowel wall but avoiding injury to it. The limits of the sphincters, including the parasagittal fibers, muscle complex, and levator muscle are determined by electrical stimulation, and the rectum is repositioned within it.

In many cases, the patient is found to have had colon, rather than rectum, pulled down to the anus. The rectum may have been resected. This is identified by the presence of a mesentery attached to the bowel. In such cases, the mesenteric fat should be trimmed from the last few centimeters of the rectum to allow for direct contact between the sphincter mechanism and the colonic wall. An anoplasty, performed within the limits of the sphincter mechanism, completes the reconstruction.

The number of patients that require a reoperation for fecal incontinence has decreased significantly over the years. This is likely due to the increased use of the posterior sagittal approach, which provides superior exposure and prevents the mislocation of the rectum seen with other techniques.

In the past, many patients underwent abdominal perineal pull-throughs with endorectal dissections of the rectosigmoid (14). This procedure essentially resulted in

loss of the rectosigmoid. These patients do not suffer from constipation. Instead they suffer from increased colonic motility and a tendency to diarrhea. Revisional surgery is not offered to them, because they never regain bowel control, owing mainly to the loss of the rectal reservoir. They are provided with bowel management, with the goal of slowing down the colon medically, and emptying it daily with an enema (18). Fortunately, endorectal pull-throughs for anorectal malformations are rarely done and it is rather unusual to see these patients.

Patients born with a poor prognosis defect and fecal incontinence are also considered inappropriate candidates for reoperation. These patients typically have an abnormal sacrum, flat perineum, and poor sphincters. There is usually evidence that they were born with a high rectoprostatic or rectobladder neck fistula, or a cloaca with a common channel longer than 3 cm. Their sacral ratio is almost always low, often less than 0.4. Reoperations on these patients, even if they have a completely mislocated rectum, are not helpful because they do not improve after reoperation. Instead they are offered a bowel management program with a daily enema, (18) in order to prevent soiling and to keep them completely clean.

When revisional surgery for fecal incontinence is offered, the likelihood of the patient regaining bowel control is reviewed with the family. Even with those patients who are expected to improve, the bowel management program is implemented prior to surgery. If it turns out that the patient does not improve enough after reoperation to avoid enemas, the already proven bowel management is reinstated.

SEQUELAE FROM CONSTIPATION

Constipation is the most common functional disorder observed in patients who undergo PSARP (11). Interestingly the incidence of constipation is inversely related to the height of the anorectal malformation. This means that patients with the best prognosis for bowel control have the highest incidence of constipation. Patients with very poor prognosis, such as bladder neck fistula, have a rather low incidence of constipation.

Constipation seems to be related to the degree of preoperative megarectum. Colostomies that do not allow cleaning and irrigation of the distal colon lead to more megarectum. Transverse colostomies lead to a micro left colon with dilatation of the rectosigmoid. Loop colostomies allow for passage of stool and distal fecal impaction. It is clear that keeping the distal rectosigmoid empty and not distended from the time the colostomy is established, and proceeding with pull-through and subsequent colostomy closure as early as possible within several months, reduces the development of megarectosigmoid and results in better ultimate bowel function.

A significant number of patients suffer from sequelae from the mismanagement of their constipation. Many patients are described as having "fecal incontinence" when they actually have untreated severe constipation and chronic impaction. This condition is actually overflow pseudoincontinence. These patients have several common features. All were born with a malformation with good functional prognosis, and all underwent a technically correct, successful operation. Postoperatively, they have severe constipation which is not managed aggressively. They therefore developed megasigmoid and chronic fecal impaction, which initiates a vicious cycle. Adequate treatment of their constipation, with or without a sigmoid resection (19), can render them fecally continent.

All patients in this pseudoincontinent group should undergo a “laxative test” to determine if they are fecally continent. First, large volume enemas are administered until the patient’s colon is clean (disimpacted) as documented on a plain radiograph. Daily laxatives are then administered, increasing the amount each day until the amount necessary to produce colonic evacuation is determined. If the patient demonstrates the capacity to feel the stool in the rectum, reaches the bathroom, has voluntary bowel movements and remains clean every day, the patient is continent. Such a patient can then be offered the option of continuing treatment with large quantities of laxative or undergoing a sigmoid resection (19) in order to make the constipation more manageable and thereby decreasing the laxative requirement.

This group of patients is extremely important to recognize. Some of these patients may be wrongly diagnosed as suffering from true fecal incontinence and some have even undergone reoperations such as gracilis muscle transfers or artificial sphincters, which can actually make the patient worse. This problem should be suspected when one sees a patient who was born with a benign malformation, who underwent a technically correct operation, but was not treated correctly for constipation.

CONCLUSION

Unfortunately, despite great advances in pediatric surgical care, there remains a significant number of patients who undergo attempted anorectal repairs with significant complications requiring reoperations. Many of these problems are preventable. For good results, one must have a thorough understanding of the spectrum of anorectal malformations, employ proper newborn evaluation and management, use meticulous technique, and employ rigorous careful postoperative regimens. These basic fundamentals need to be emphasized in the training of pediatric surgeons, so as to improve the outlook for children born with anorectal malformations.

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19

Reoperation for Recurrent Anal and Perianal Conditions

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INTRODUCTION

Disorders of the anus and rectum present frequently in infants and children, so it is important for the pediatric surgeon to identify conditions that require operative intervention. Fortunately, most minor anorectal disorders respond successfully to initial management and rarely recur. The following diseases will be addressed in this chapter, in relation to recurrence and reoperative surgery: rectal prolapse, fistula-in-ano, hemorrhoids, and anal warts. Management of perianal disease secondary to Crohn's disease and congenital anomalies of the anus and rectum are presented in other sections of this book.

RECURRENT RECTAL PROLAPSE

Rectal prolapse is defined as full-thickness protrusion of the rectal wall out of the anal canal. The typical presentation of rectal prolapse is a 1- to 3-year-old child who develops a bulging mass exiting the anus after defecation (1). The mass will often disappear spontaneously, is usually painless, and may have minor surface bleeding. Rarely, an older child may present with an incarcerated rectal prolapse associated with intense pain and bleeding. The etiology of childhood rectal prolapse is thought to include chronic constipation, lack of fixation of the rectum, prolonged periods of straining during "potty training," and lack of angulation of the pelvis and rectum in the first several years of life (1-3).

Evaluation of a child with rectal prolapse includes a thorough history and physical examination. Stooling patterns, history of constipation and/or diarrhea, evaluation of

muscular or neurological impairment, and dietary habits are all key elements. The physical examination includes growth and development parameters, which may be abnormal in children with previously undiagnosed cystic fibrosis. The physical stigmata of a connective tissue disorder, such as Ehlers-Danlos syndrome, should be assessed during the examination. Visual inspection of the perineum evaluates anal position and external masses, whereas the digital rectal examination assesses anal tone and the presence of internal masses. We have observed that referring physicians often confuse rectal prolapse with external hemorrhoids. In rectal prolapse there are circular concentric folds, whereas in hemorrhoids, the folds are radial, as shown in Fig. 1. If no obvious pathology is visualized on initial examination, it may be helpful to have the child blow up a balloon or to reexamine the child after allowing time to sit on the toilet and attempt defecation. One can also ask the parents to provide a digital photograph of the rectal prolapse when it occurs in the home setting.

A child with documented rectal prolapse should undergo a sweat chloride test, because previously unsuspected cystic fibrosis is found in 6–20% of these patients (3,4). In selected patients, thyroid function studies should be performed to exclude hypothyroidism, and genetic testing should be considered if the physical examination is suggestive of a connective tissue disorder. A contrast enema is helpful in eliminating an anatomic cause for the prolapse.

Because the majority of children with rectal prolapse will have chronic constipation, treatment includes dietary adjustment, high-fiber foods, and a laxative bowel regimen. If the prolapse fails to respond to a carefully followed bowel regimen for at least 6 months, or if the patient has an underlying connective tissue disorder, operative treatment should be considered. The operations designed for the treatment of rectal prolapse are classified into two main approaches, perineal and abdominal (1,3,5–7). Our preferred initial management is placement of a Thiersch wire using an 0-prolene suture as the “wire,” and tightening the suture over a 10-F Hegar dilator (Fig. 2–4). If prolapse recurs after placement of the initial Thiersch wire, we favor a second wire, especially



Fig. 1. Rectal prolapse.

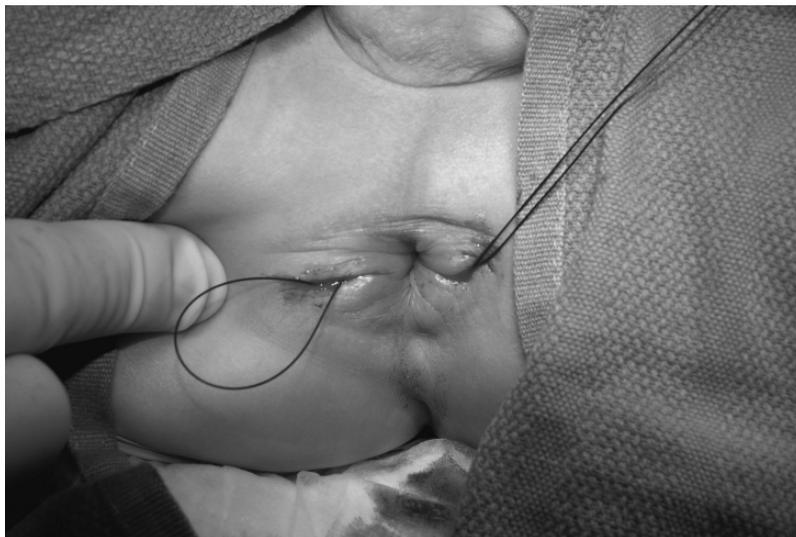


Fig. 2. Thiersch wire placement. Incisions at 3 and 9 o'clock.



Fig. 3. Thiersch wire placement. Tightening of the suture over Hegar dilator.

if it appears that the prolene suture was either too tight or too loose. Recurrence after a second Thiersch wire warrants additional diagnostic evaluation, including anal and colonic manometry. Dysmotility of the colon may be responsive to pharmacologic treatment, although some children may require a colostomy or enterostomy.

If constipation is a major component of the recurrent prolapse and the contrast enema shows a redundant sigmoid colon, the next step is a sigmoidectomy and rectopexy. The rectum is completely mobilized and the rectum is sutured to the presacral fascia with a prolene suture on either side of the rectum. We do not advocate placement of mesh, which has no clear advantage in terms of recurrence, and may be associated with



Fig. 4. Thiersch wire placement. Final appearance.

long-term complications, such as obstruction. If constipation is not a major component, we perform a laparoscopic rectopexy, with a suture securing either side of the fully mobilized rectum to the presacral fascia. Recurrence after rectopexy alone would be followed by a sigmoidectomy and rectopexy. Our recommended treatment algorithm is shown in Fig. 5.

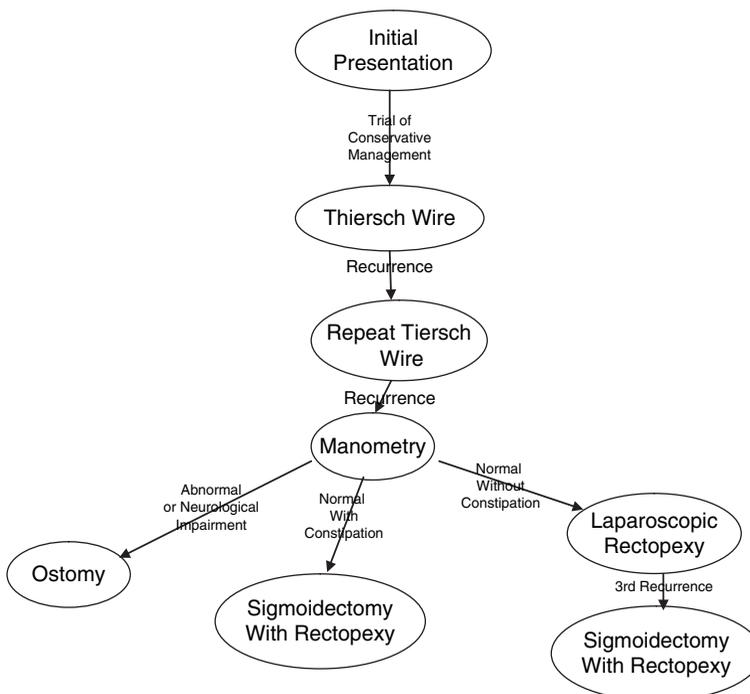


Fig. 5. Treatment algorithm for rectal prolapse.

The treatment of rectal prolapse has not been studied prospectively in clinical trials in either adult or pediatric patients, and a Cochrane review could not delineate a clear advantage of one operative technique (6). The overall recurrence rate for all operations is 10–40%, regardless of the type of operation (1,3,5,7).

RECURRENT PERIRECTAL ABSCESS/FISTULA-IN-ANO

Peri-rectal abscesses develop from an occluded duct of an anal gland with subsequent bacterial overgrowth (8). The typical patient presents in infancy with a tender, erythematous swelling of the soft tissues surrounding the anus. On physical examination the area of involvement is firm, fluctuant, and tender to palpation. Occasionally, there is spontaneous drainage of purulent material. Traditional treatment for recurrent perirectal abscesses has been incision and drainage with packing or removal of a small ellipse of skin (9). We do not recommend probing of the abscess cavity at the time of drainage, because it could promote development of a fistula (9,10). If obvious fluctuance is not appreciated, an ultrasound may be helpful in identifying a fluid collection. In the absence of a clearly fluctuant mass, the infant should be treated with antibiotics to cover Gram-positive and Gram-negative bacteria and symptomatic pain control.

Recurrent perirectal abscesses and fistulae have been reported to occur in 20–50% of patients (9–11). For infants less than 1 year of age, we recommend incision of the recurrent abscess; and if a fistula develops, a period of observation for 3–6 months. Spontaneous closure of perirectal fistulae in infancy has been noted in more than 40% of cases, justifying a period of observation in most patients (11,12). If the fistula persists after this time period, the next step should be fistulotomy, because most of the fistulas will be superficial to the sphincter muscles (10). Recurrence after fistulotomy is in the range of 2–9% (13,14). If the fistula tract involves the sphincter muscle, or if the fistula recurs after a fistulotomy, we proceed with injection of fibrin glue. Although the use of fibrin glue in children has not been reported, extrapolation from adult data suggests that success can be achieved in 14–78% of cases (9,15–19). The instillation of fibrin glue is straightforward, can be repeated on multiple occasions, and carries no risk of incontinence. For those fistulas that persist after fibrin glue instillation, the next step is a mucosal advancement flap (9). Long-term success for this procedure is between 60–90%, although the risk for incontinence approaches 15% (20–22). An interesting new technique for complex fistulas is placement of an anal fistula plug made from Surgisis®. In a clinical trial by Johnson et al., the anal fistula plug had an 87% success rate, in comparison to a 40% success rate with fibrin glue for complex fistulas (23). These results are preliminary and there are no studies of this technique in pediatric patients.

PERSISTENT HEMORRHOIDS

Although a common disorder afflicting approximately 5% of the American adult population, hemorrhoids are a relatively rare disorder in children, and thus recurrent disease is fortunately even rarer (24). The anal cushions, which contain the underlying arterial-venous plexus leading to hemorrhoids, are present in infancy. Although the exact etiology of hemorrhoids is unknown, excessive straining while defecating, spending long amounts of time on the toilet, and chronic constipation are believed to contribute to alterations in the arterial-venous plexus (25,26).

In order to better understand the treatment of hemorrhoids, they are classified as internal when they are located above the dentate line, and external when they are below the dentate line. Internal hemorrhoids are further subcategorized into four types depending upon the amount of mucosa that is protruding (27). First-degree hemorrhoids are those that cause symptoms but do not exit the anal canal, second-degree protrude from the anal canal but reduce on their own, third-degree require manual reduction, and fourth-degree are not able to be manually reduced (usually involving an internal and external component). The typical clinical presentation is a child whose parents are concerned that they have either noticed a bulging mass protruding from the rectum after a bowel movement or the finding of blood in the toilet after stooling. If the hemorrhoid is thrombosed, the major symptom is pain associated with a perianal mass.

When a patient presents with symptoms consistent with hemorrhoids, a dietary and stooling history should be obtained, followed by physical examination. The anus should be inspected with the child relaxed and with the child actively straining. This is best accomplished by having the child sit on the toilet for 5–10 minutes, which helps the pelvic floor and anal muscle complex to relax fully. If no abnormalities are visualized and the symptoms persist after management of any stooling problems, such as constipation, anoscopy should be performed, either under anesthesia for younger children or in the office setting for older adolescents. Anoscopy should reliably confirm the presence of hemorrhoids and differentiate the disorder from rectal prolapse, anal fissures, and skin tags, all of which can be confused with hemorrhoids. If the anoscopic evaluation raises concern for varicosities rather than hemorrhoids, abdominal duplex ultrasonography should be performed to assess for portal hypertension.

Treatment for primary or recurrent hemorrhoidal disease depends on the severity of symptoms. For all patients, initial therapy is diet modification to increase water and fiber intake, lessening the straining with defecation, and decreasing the amount of time on the toilet (28,29). Corticosteroid creams can be used on a short-term basis to alleviate symptoms related to pruritis.

Optimal treatment for an acutely thrombosed hemorrhoid is excision under general anesthesia. In a recent review by Greenspon et al. (30), the time to symptom resolution was quicker (3.9 days versus 24 days) and recurrence was lower (6.3 versus 25.4%) in patients who had immediate operative treatment in comparison to patients in the nonoperative group.

For internal hemorrhoids, treatment is based upon the grade of involvement. Ablative therapy is recommended for patients with persistent internal hemorrhoids and grades 1–3. The three most common ablative methods are sclerotherapy, bipolar diathermy, and rubber-band ligation. Sclerotherapy is a submucosal injection of 3–5 mL of 5% phenol in oil, 5% quinine and urea, or hypertonic saline into the base of the hemorrhoid (26). Bipolar diathermy is the application of heat directly to the hemorrhoidal complex (26,31). Rubber-band ligation involves placing a tight-fitting rubber band around the bulk of the hemorrhoidal tissue, which results in necrosis (26,32–34). Initial success for all three ablative treatments is 70–90% (26,31–34). We have utilized rubber-band ligation in adolescent patients with excellent short- and long-term results. However, recurrence after all ablative modalities ranges from 20–40%, and the patients and parents must be informed that repeated treatment may be required. If the hemorrhoids are too extensive for ablative therapy, or there are concomitant anal conditions (fissure or fistula), a formal hemorrhoidectomy should be considered (26,31). In patients who have had hemorrhoids treated previously, care must be taken to perform

the dissection in the submucosal plane in order to avoid injury to the sphincter muscle complex. Open hemorrhoidectomy is associated with substantial postoperative pain, resulting in prolonged absence from daily activities such as school and sports (35). Recently, the circular stapler has been applied to the excisional treatment of internal hemorrhoids in adult patients, resulting in shorter hospitalization, decreased utilization of analgesics, and earlier return to daily activities (24,26,36,37). In these series, the recurrence rates with the circular stapler are higher than for open hemorrhoidectomy.

RECURRENT/PERSISTENT ANAL WARTS

Anal warts, also known as condylomata acuminata, are caused by one of at least 30 subtypes of human papillomavirus (38). Although most anal warts are related to sexual transmission, it is also possible for an infant to acquire the disease during vaginal delivery from an infected mother (38,39). The lesions rarely produce any symptoms in the patient and the diagnosis can usually be made by physical examination alone. If there is any question about the diagnosis, if the patient is immunocompromised, or the lesions are not responding to therapy, then a biopsy is recommended to make the definitive diagnosis (40).

Untreated anal warts can regress spontaneously, remain stagnant for many years, or progress and grow very rapidly. Treatment is based on the number, location, and size of the lesions, as well as patient and family preference.

There are two basic types of treatment, chemical and ablative. Chemical options include podophyllin (15–20% solution) and trichloroacetic acid (39,40). Podophyllin requires multiple applications and has a success rate of up to 50%, but with the limitation that it cannot be used within the anal canal. Trichloroacetic acid, our first-line therapy, also requires multiple applications and has a success rate up to 75%. It has the advantage of being able to be used in the anal canal. Topical 5-FU, previously utilized for maintenance therapy after operative removal, is no longer recommended (41).

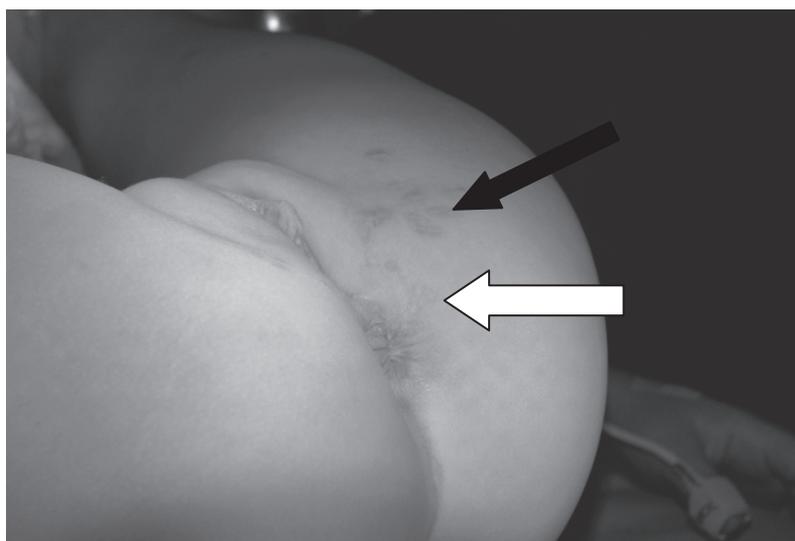


Fig. 6. Recurrent anal warts. Black arrow shows previously treated lesions and white arrow shows new lesions.

Intralesional injection therapies with fluorouracil or interferon are other options usually reserved for refractory cases, and have reported success rates around 70% (41).

Ablative therapies include surgical excision, cryotherapy, cautery, and laser therapy (39,40). These all have excellent success rates approaching 90%, but have limitations of potential scarring, and in children require general anesthesia for each application. Our preferred approach for recurrent and refractory disease is operative resection for larger lesions (>1 centimeter) and laser therapy for smaller lesions. Fig. 6 demonstrates a patient with recurrent disease with scars from previous cautery ablation. Laser therapy appears to be better tolerated with respect to postoperative pain. One must always keep in mind that these lesions have premalignant potential. We recommend biopsy and excision of all anal warts, particularly if they are unresponsive to chemical ablation.

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Reoperation of the Liver and Biliary System

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INTRODUCTION

Reoperative surgery for hepatobiliary disease and its common consequence, portal hypertension, like so many subdisciplines in pediatric surgery, is a study of complication management. Appropriate initial evaluation and operative planning, coupled with defined steps in primary surgical management, will allow many potential problems to be avoided. In this chapter, we attempt to outline not only our approach to the common problems requiring reoperation in hepatobiliary surgery, but also highlight the primary operative steps we feel are important to achieve initial success, avoiding the need for reoperative intervention.

CHOLEDOCHAL CYSTS

Choledochal cysts present a unique form of surgically correctable biliary obstruction. The incidence of one in 13,000 in the United States contrasts sharply with an occurrence rate of one in 1000 admissions to the Kobi Children's Hospital in Japan (1). No clear explanation for this significant difference in incidence in the Orient has emerged. The etiology is unknown, but choledochal cysts are thought to be congenital. The pathologic features frequently include anomalous junction of the pancreatic and common bile ducts (CBDs) (pancreatico-biliary malunion), intrahepatic bile duct dilation and in cases diagnosed later, hepatic fibrosis. It is this pancreatico-biliary malunion, allowing

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the reflux of pancreatic enzymes into the bile duct, which can lead to mucosal inflammation, distal edema and obstruction, and mucosal dysplasia.

Classification

The spectrum of disease that is seen within this unique group of abnormalities was initially analyzed and classified by Alonso Lej and subsequently modified by Todani and his colleagues (2–4). Their classification system divides the spectrum of anomalies into five types. Type I cysts exhibit a segmental or diffuse fusiform dilation of the extrahepatic biliary duct and represent the most common variant. Type II cysts form a diverticulum off of the extrahepatic bile duct, and type III cysts form a choledochoceles of the distal CBD, most often within the duodenal wall. More diffuse and variable forms of bile duct dilations have led to the establishment of types IV and V, which include fusiform dilation of the extrahepatic bile duct (type I anatomy) occurring in conjunction with either intrahepatic bile duct cysts (type IVa), extra hepatic choledochoceles (type IVb), or the presence of single to multiple cystic dilations of the intrahepatic bile duct (type 5) (Fig. 1A). Type I cysts make up the vast majority of cases (>93%), type II make up 6%, and 2% are type III. Type IV and V cysts are very uncommon in all series (5). Miyamao has further modified this classification to subdivide the cysts based on the presence of pancreatobiliary malunion (Fig. 1B) (6,7).

Clinical Presentation

Classification of choledochal cysts can also be based on their clinical presentation rather than their precise ductal anatomy. Patients are generally divided into two categories, infantile and non-infantile. In the infantile form, distal ductal atresia or severe stenosis is frequently seen, caused by either a primary anatomic abnormality or a secondary inflammatory stenosis. In these patients, symptoms occur early in life and are part of the spectrum of prolonged conjugated hyperbilirubinemia in infancy. This presentation must be distinguished from biliary atresia, as up to 80% will have jaundice and acholic stools as well as hepatomegaly. A palpable abdominal mass is present in 60% (8). They can be distinguished from biliary atresia by the presence of distinct intrahepatic biliary ducts. It is very uncommon for these infants to present with the classic triad seen in older children. In the past, their clinical course was often complicated by progressive biliary cirrhosis and portal hypertension. At present, with the almost universal use of prenatal ultrasound, progressive liver damage is thankfully uncommon and most reach operative intervention in infancy (9,10). The noninfantile presentation is more often manifested by the classic clinical triad of abdominal pain, right upper quadrant mass, and variable jaundice with occasional vomiting. Intermittent jaundice and recurrent cholangitis are manifestations of intermittent to complete obstruction (11). Repeated episodes of reflux of pancreatic juice into the extrahepatic bile ducts has been postulated as the etiology inducing progressive injury to the bile duct wall with subsequent ductal dilatation. This progressive ductal injury is associated with a slowly progressive clinical course and incomplete or intermittent obstruction, making cirrhosis and portal hypertension more uncommon complications. Biliary stasis can lead to the formation of both biliary sludge and stones, and pancreatitis is not uncommon.

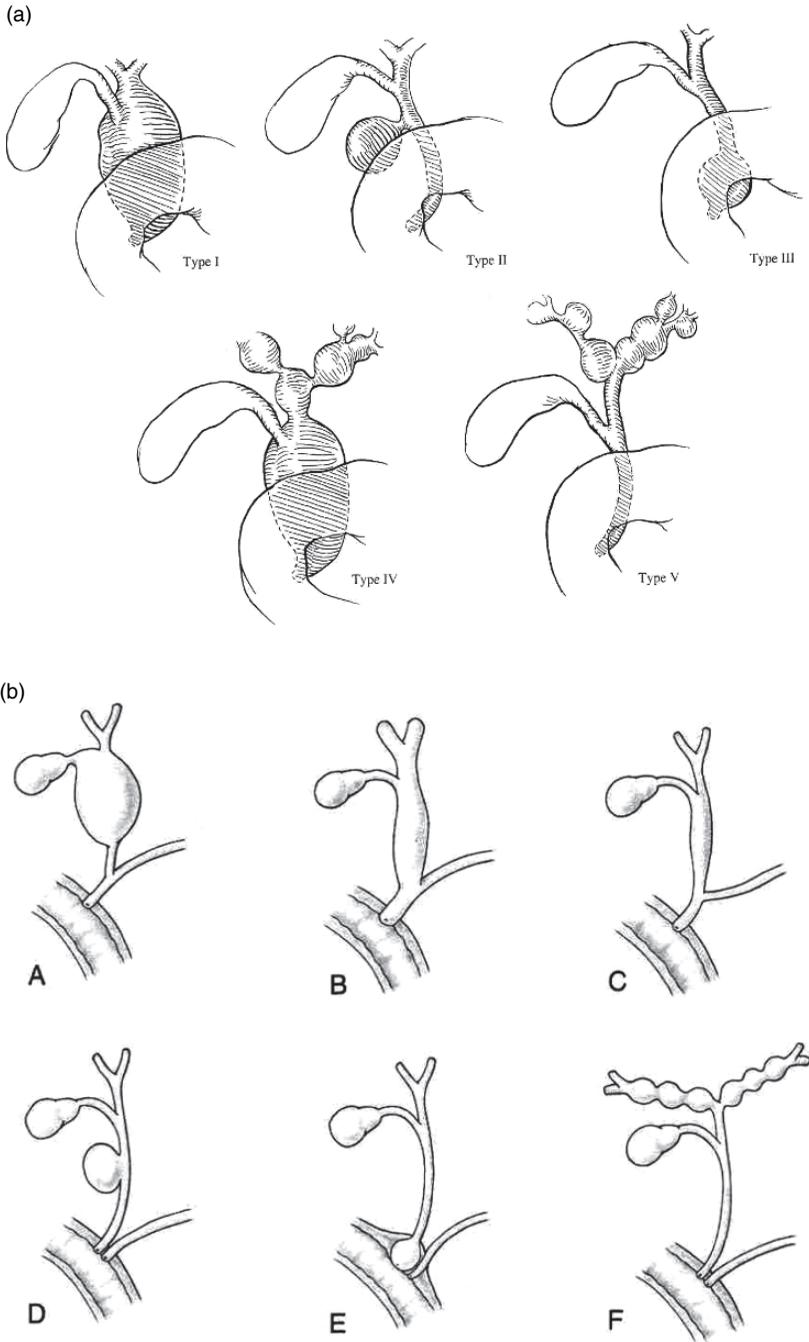


Fig. 1. (A) The Todani modification of the Alonzo-Lej classification of cystic malformations of the biliary ductal system (5). (B) Classification of choledochal cysts with pancreaticobiliary malunion (PBMU). (A) Cystic dilation of the extrahepatic duct; (B) Fusiform dilatation of the extrahepatic bile duct; (C) Forme fruste choledochal cyst without PBMU; (D) Cystic diverticulum of the common bile duct; (E) Choledochocele; (F) Intrahepatic bile duct dilatation alone (Caroli's Disease) (7).

Operative Management

The primary treatment of the common type I choledochal cyst consists of cyst excision followed by choledocho-jejunostomy with Roux en-Y reconstruction. Type II cysts, the diverticular variety, can usually be completely excised with primary closure of the bile duct. Type III cysts, or choledochoceles, are best treated by unroofing the cyst. No true excision is necessary as long as appropriate drainage is achieved. In type IV cysts, the CBD abnormalities are treated as type I anomalies. Additional procedures may be necessary in order to provide adequate decompression of the intrahepatic portions of the ductal malformation. It is important in the treatment of all type I cysts to excise or eliminate the entire cystic mucosa wall prior to reconstruction, as malignant transformation of the choledochal cyst lining has been reported (12).

The most problematic complications of choledochal cyst excision occur at the time of primary operative intervention. In cases identified in infancy, inflammation surrounding the cyst and the other critical portal structures (hepatic artery and portal vein) is less common. Complete excision of the cyst and its entire wall can usually be carefully accomplished without incurring significant risk of damage to the portal vein in particular. In patients who present later in life having undergone multiple bouts of cholangitis, the connection between the posterior wall of the choledochal cyst and the anterior surface of the portal vein can be both intimate and ill-defined. Under these circumstances, attempts to completely excise the posterior wall of the cyst as it is juxtaposed to the portal vein can lead to critical injury of the portal venous structures. In these cases, complete excision of the cyst mucosa, leaving the posterior fibrous wall of the cyst in continuity overlying the portal vein, is a wiser surgical option (13) (Fig. 2). Damage to the portal vein incurred while undertaking dissection of this difficult inflammatory plane can result in portal vein stenosis and subsequent portal hypertension.

Indications for Reoperation

Stenosis at the hepatico-jejunostomy is the most common anticipated complication of biliary reconstruction. This is directly related to the level of the anastomosis within the biliary tree (common hepatic duct versus right/left hepatic duct), the size of the reconstructed bile duct, and the degree of inflammation within the wall (14). In most cases, a short section of the proper hepatic duct at the superior extent of the type I cyst can be identified for anastomosis. Rarely is it necessary to excise the cyst to the level of the right and left hepatic ducts (15). The primary anastomosis reconstructs the end of the divided bile duct to the side of a 35–40 cm isoperistaltic retrocolic Roux-en-Y. Care must be taken to place the anastomosis as close as is possible to the end of the Roux-en-Y, so that there is no blind ending sac at the end of the Roux, which can dilate with time and lead to blind limb syndrome or cholangitis. The anastomosis can be completed using either a single layer of dissolving monofilament sutures, 5.0 or 6.0 in most cases and 6.0 in all infants. At times, the bile duct wall is thick enough that several supporting sutures incorporating the bile duct wall and the seromuscular intestine can be placed to take tension off of the primary anastomosis. The size of the anastomosis is usually sufficient, making internal or externalized bile duct stents unnecessary. In cases where individual reconstruction of the right and left hepatic duct is necessary, a short multifenestrated silastic internal stent can be placed and secured using a dissolving monofilament suture, allowing the stent to pass into the

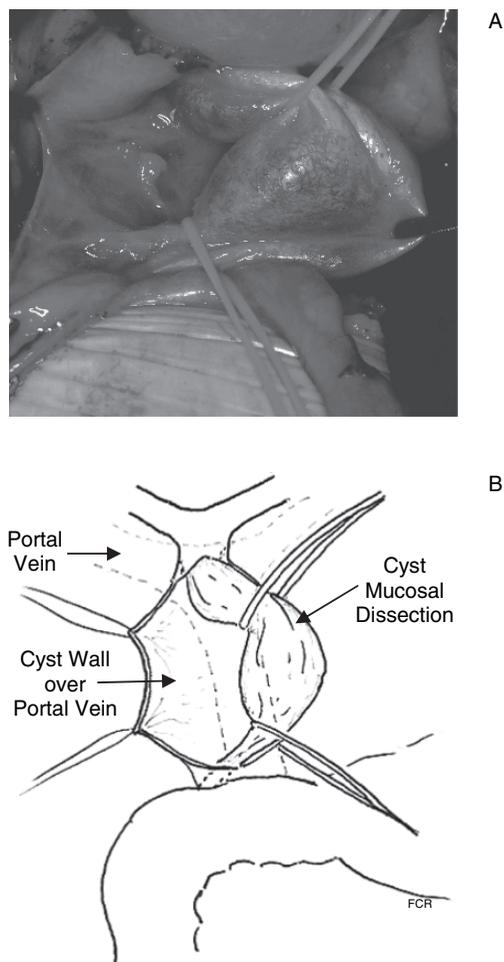


Fig. 2. Choledochal cyst excision using mucosectomy of the posterior wall to prevent hilar structural damage. (A) Operative photograph; (B) schematic diagram.

intestinal tract in several months. Recurrent cholangitis and biliary anastomotic stenosis related to reflux into the Roux-en-Y can be decreased by constructing the Roux-Jejunal anastomosis to the side of the functional jejunal segment, and narrowing the angle between the inflow jejunum and the Roux-en-Y with several sutures to approximate the limbs forming a functional “Y” rather than the “T”-shaped anastomosis that often occurs with the conventional techniques (Fig. 3).

In an effort to identify potential stenosis of the reconstructed bile duct prior to the onset of cholangitis, routine surveillance in the early postoperative period using ultrasound is recommended. Initial ultrasound visualization of the hilar anastomosis 3 to 4 weeks following cyst excision is sufficient to establish a new baseline anatomic assessment after postoperative edema has resolved. Follow-up examinations at 3 months, 6 months, and 1 year to ensure there is no progressive dilatation of the intrahepatic bile ducts. If anastomotic narrowing is suggested by ultrasound examination or the development of clinical signs including recurrent cholangitis or intermittent jaundice, initial treatment could be instituted by a percutaneous transhepatic

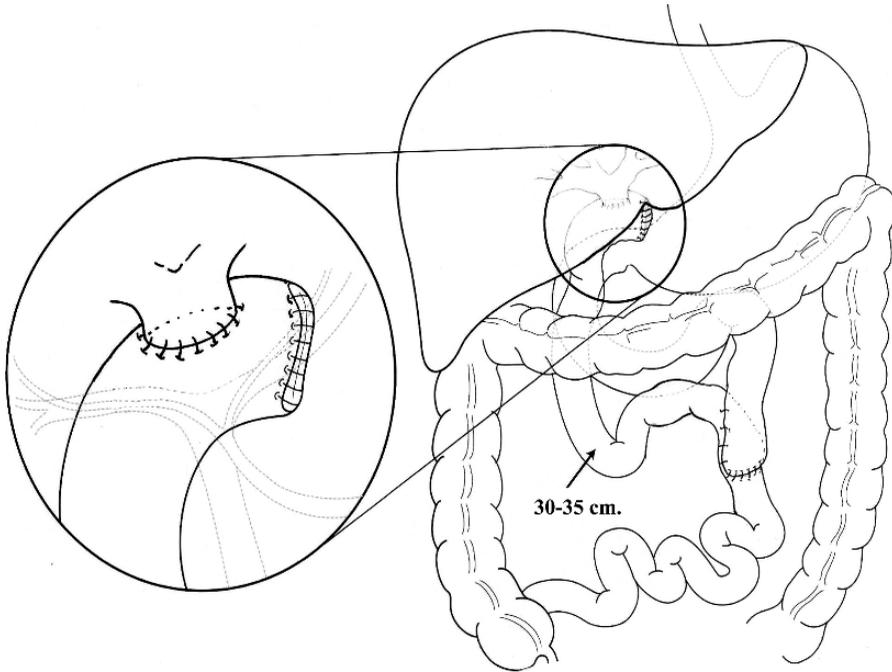


Fig. 3. Schematic diagram of Roux-en-Y anastomosis to prevent reflux of enteric contents into the biliary drainage limb. Authors' technique.

balloon dilatation of the anastomotic site. In cases where serial balloon dilatation was unsuccessful, surgical exploration and reanastomosis is indicated.

Development of a remnant cyst within the head of the pancreas can also require reoperation owing to the formation of stones, or the development of recurrent pancreatitis. This is also managed best by prevention at the time of the initial operation. The distal portion of the cyst is always best-excised after the cyst has been opened and divided, allowing excellent visualization of the lower cyst wall and identification of the distal CBD orifice. In the saccular cysts, the distal ductal remnant is often very small in diameter and a small portion of the cyst is present within the pancreatic tissue, making recurrence unlikely. In the fusiform cysts, the distal CBD must be carefully dissected to the lowest margin of the cyst. In cases where there is dilated duct up to the pancreatic duct junction, careful division just superior to the pancreatic duct is necessary. Residual ductal remnants can dilate with time and develop a recurrent cyst (Fig. 4). Re-resection of this remnant cyst is complicated by residual inflammation and scarring from the primary operation, making identification of this critical duct anatomy more difficult. Mucosectomy and closure of the distal ductal remnant can be successfully used in difficult cases.

The incidence of biliary carcinoma in association with choledochal cysts is 20 times greater than that of the general population (12). Although malignant bile duct tumors have been reported in relatively young children, the typical onset of disease is during adulthood (76%). The typical malignancy is an adenocarcinoma of the bile duct and the youngest reported patient to date has been 17 years of age. The risk increases with increasing age in patients who have not been treated with complete duct mucosal excision. The etiology of a malignancy is postulated to be from chronic irritation and

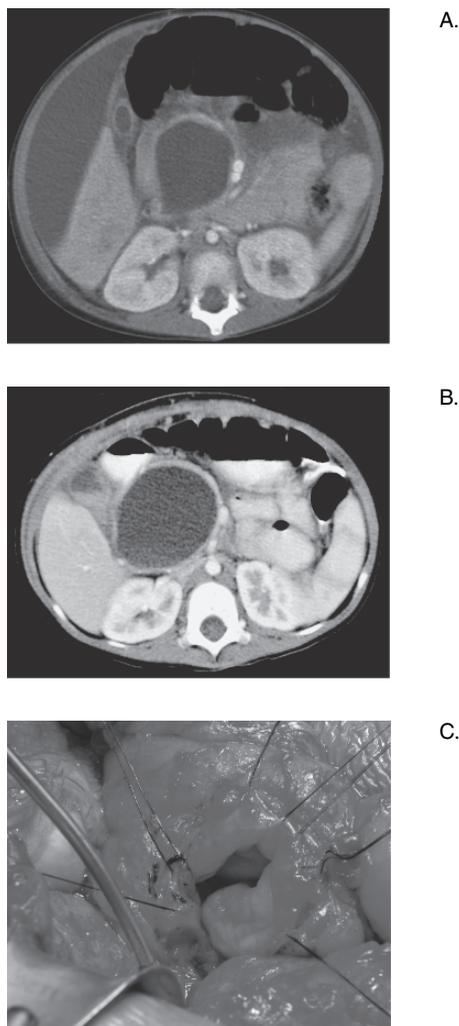


Fig. 4. Recurrent cyst formed from distal remnant of fusiform choledochal cyst. (A) 2.5-year-old at presentation with ruptured choledochal cyst and bile peritonitis. (B) CT Scan of remnant cyst in head of pancreas 4 weeks after primary CDC resection. (C) Operative photograph of remnant cyst resection secondary to enlargement and pancreatitis.

inflammation. Reflux of pancreatic enzymes into the area of the cyst with injury to the cyst mucosa and disruption of the cyst wall, and attendant stasis and superimposed infection, may set the stage for emergence of malignant cells within this milieu. Dysplasia of the bile duct mucosa without carcinoma is frequently seen. This hypothesis is supported by the emergence of intestinal metaplasia within the choledochal cyst wall with advancing age (16).

BILIARY ATRESIA

Biliary atresia is a progressive obstructive cholestasis of unknown etiology leading in most cases to progressive biliary cirrhosis and portal hypertension. In the late 1950s, the Japanese surgeon, Morio Kasai, undertook radical excision of the atretic

extrahepatic biliary remnants in an effort to expose patent microscopic biliary channels and achieve effective bile drainage. This operation is the foundation for the initial operative treatment of biliary atresia today. Many children have achieved long-term survival and function following this procedure, but the progressive nature of biliary atresia and the rapid development of portal hypertension and liver failure makes biliary atresia the most common indication for liver transplantation in the United States.

The clinical forms of biliary atresia can be divided into two subtypes: a postnatal form that progresses after birth and a fetal/embryonic form that is present at birth and is often associated with other congenital anomalies (17). The postnatal presentation accounts for 75–80% of cases of biliary atresia. These infants present with jaundice, which appears after “physiologic jaundice” should have resolved, and increases in severity within weeks. Hepatic histology is significant for the presence of bile duct remnants with acute and chronic inflammation within the portal triads. This inflammation appears to be progressive. Associated anomalies are uncommon. The fetal/embryonic subtype occurs in 15–20% of infants with biliary atresia, and is characterized by jaundice that is recognized and progresses during the first few days of life. In most cases, no jaundice-free interval can be identified. The probable in-utero onset of this is suggested by the absence of patency in the biliary structures, and a frequent absence of bile duct remnants at early laparotomy. Associated congenital anomalies are common and follow one of three patterns: (1) Most common group (60%)—cardiovascular (atrioventricular canal defects, septal defects, valvular abnormalities, tetralogy of Fallot, patent ductus arteriosus), genitourinary (kidney or collecting system), and gastrointestinal (Meckel’s diverticulum, midgut anomalies); (2) laterality sequence anomalies (30%)—polysplenia, asplenia, situs inversus, intestinal malrotation, cardiovascular anomalies; and (3) isolated intestinal malrotation (10%). Most studies have demonstrated a poorer prognosis with the fetal/embryonic subtype, although this experience is not universal (17).

To be successful, the Kasai procedure should be done before irreversible sclerosis of the intrahepatic bile ducts has occurred. Prompt evaluation is necessary for any infant with jaundice older than 14 days to determine if conjugated hyperbilirubinemia is present. If infectious, metabolic, and endocrine disorders are excluded, then hepatic biopsy most accurately identifies infants with suspected biliary atresia from those with alternative diagnoses. In these selected infants, exploratory laparotomy and intraoperative examination/cholangiogram should be done expeditiously by a surgeon who has experience undertaking the Kasai procedure.

Preliminary laboratory studies to identify biliary atresia are rarely definitive. Abdominal ultrasonography excludes choledocholithiasis, perforation of the bile ducts, and choledochal cyst. In patients with suspected biliary atresia, the gallbladder is either not visualized or atretic and the fibrous biliary ductal remnant often forms a “triangular cord” appearance. Cholescintigraphy is rarely helpful even with phenobarbital induction.

Age at operation has been recognized as a critical determinant of both early bile drainage and long-term survival. Infants undergoing operation prior to 60 days of life have high likelihood of successful bile drainage (60–80%), and 70–80% continue to enjoy a successful result at 10 years. This contrasts to infants younger than 90 days of age at operation, where successful bile drainage is achieved in 40%, with 10–15% enjoying more than 10-year survival without liver transplantation. Histologic examination of the liver by percutaneous liver biopsy is the most helpful step in

evaluating children with suspected biliary atresia, and selecting appropriate candidates for laparotomy. The degree of portal fibrosis is especially important in infants who present at a later age for evaluation. The combination of late presentation (>4 months of age) and severe fibrosis makes successful bile drainage unlikely; in these cases primary liver transplantation should be considered to be the preferred option.

We believe all patients younger than 4 months of age at referral should undergo an initial Kasai portoenterostomy. In individuals referred late (>4 months) who present with signs of progressive fibrosis on biopsy and clinical portal hypertension, primary liver transplantation is indicated. There are individuals who are referred late with minimal liver injury who are still good primary Kasai candidates, but these should be selected on an individualized basis. Primary surgical intervention should occur at a center with experience in the Kasai procedure, and complex hepatobiliary disease. Both the presence of an experienced surgical team, and an institution skilled in complex liver disease have been shown to improve overall primary operative success (18,19).

Indications for Early Reoperation

Reoperation is reserved for infants who have had active bile drainage following their initial operation, leading to an anicteric state, followed by abrupt cessation of bile excretion, and who have favorable hepatic histology and biliary ductal remnant histopathology from their primary procedure. Favorable histology would include the absence of progression of intrahepatic fibrosis/cirrhosis and the absence of hepatocyte degeneration (20). The disappearance of jaundice is best represented by a sustained total bilirubin of less than 2 mg/dL during the postoperative period (21). Although cessation of bile flow most often occurs within the first several postoperative months, other factors such as age, liver enzymes (aspartate amino transferase [AST], alanine transferase [ALT]), and coagulation status did not predict success. Reconstitution of bile flow is successful in more than one-half of patients using these selective criteria. Infants with inadequate or no initial bile flow are poor reoperative candidates, and should not be reexplored. Successful reoperation in this group with poor initial bile flow approaches only 10%. Multiple reoperative attempts to reconstruct patients with poor drainage only increase the degree of intraperitoneal scarring and vascularized adhesions surrounding the liver. Multiple studies have documented the increased complexity and risk of liver transplantation following repetitive unsuccessful reoperative attempts after the initial Kasai procedure (22). This risk is manifest by increased bleeding from portal hypertensive collaterals within dense adhesions and an increased risk of intestinal injury at the time of liver mobilization for transplantation. Although these factors have not increased posttransplant mortality in experienced hands, they substantially increased the postoperative morbidity and risk of bleeding, intestinal perforation, and infectious complications in this already difficult patient group.

Reoperation may also be necessary secondary to obstruction or complications within the draining Roux-en-Y limb. A period of enthusiasm existed for constructing “antirefluxing” valves within the draining Roux-en-Y limb. Subsequent studies have shown this modification did not significantly decrease the incidence or severity of cholangitis (23–25). At times, these intussuscepting valves can lead to mural fibrosis or luminal stenosis, creating intestinal stasis and paradoxically increasing the risk of cholangitis. This can also occur if stenosis develops at the Roux-en-Y–jejunal anastomotic site. Because this obstruction exists within the defunctionalized Roux-en-Y intestine, conventional contrast studies of the gastrointestinal tract are most often nondiagnostic.

In patients with sufficient hepatic function, a technetium-99 HIDA scan will show sufficient drainage from the liver but delayed flow and obstruction within the Roux-en-Y. This unique form of intestinal obstruction warrants immediate operative intervention; stenotic antirefluxing valves should be resected with primary reconstruction of the intestine.

The unique anatomic circumstance of a patent distal CBD and cystic duct in association with proximal hepatic duct atresia has often been treated by porto-cholecystostomy ("gallbladder Kasai"). This reconstruction, using the gallbladder and its patent outflow to drain the biliary fibrous remnant, has the advantage of maintaining normal distal ductal anatomy and the ampulla of Vater, thereby decreasing the risk of ascending cholangitis (26,27). In cases where the resistance to outflow within the patent CBD remnant precludes sufficient drainage, persistent jaundice can occur in spite of an adequate and potentially functional hilar dissection (28). Identification of this patient is critical, as early conversion from porto-cholecystostomy to conventional Roux-en-Y drainage is often successful. Appropriate initial operative assessment is the key first step to avoiding this complication. The presence of mere patency within the distal CBD and cystic duct structures is not sufficient to assure success. Ducts of sufficient diameter and low resistance must be present, and are best selected by intraoperative cholangiography and assessment of resistance to flow at the time the duct is flushed in the operating room. In cases deemed to be adequate for this reconstruction, we also leave a 5-F feeding tube within the porto-cholecystostomy lumen, which is externalized to allow both early low-pressure decompression of the reconstruction as well as assessment of biliary drainage (29). This indwelling tube can also be used to carry out a postoperative cholangiogram several days following the initial reconstruction to confirm patency and document flow into the duodenum prior to its discontinuation. Excessive drainage of bilious material from this decompression tube in the early postoperative period in conjunction with poor flow into the duodenum suggests the need for early conversion to Roux-en-Y porto-enterostomy.

Similar difficulties resulting from higher resistance to drainage from the hilar dissection site have been encountered with the appendico-duodenostomy (30,31). Although the appendix has been successfully used for years for urinary drainage, its success for primary reconstruction in patients with biliary atresia or choledochal cysts is less predictable. When undertaking primary reconstruction of the bile duct with the appendix, the cecal patch is placed at the porta hepatis fibrous remnant site, making the resulting reconstruction antiperistaltic to normal biliary flow. In addition to being prone to postoperative ischemic stenosis, manifestations of cholestasis such as increased gamma-glutamyl transpeptidase level (GGT) and histologic findings of liver fibrosis can occur. When used for patients with primary biliary atresia, clearance of jaundice was significantly impaired (28%) (32). Although this reconstruction has been successful in individual cases, any patient having undergone appendico-duodenostomy should be carefully observed for acholic cholestasis or persistent jaundice and be considered as a candidate for Roux-en-Y reconstruction if these findings occur in the framework of adequate long-term liver functional status.

A final indication for reoperation exists in patients who were initially reconstructed using any of the multiple stoma drainage options at the time of their initial Kasai porto-enterostomy. These procedures were also advocated as a method to prevent or ameliorate the effects of ascending cholangitis. However, with the development of portal hypertension, these stomas become a significant site of portosystemic collaterals,

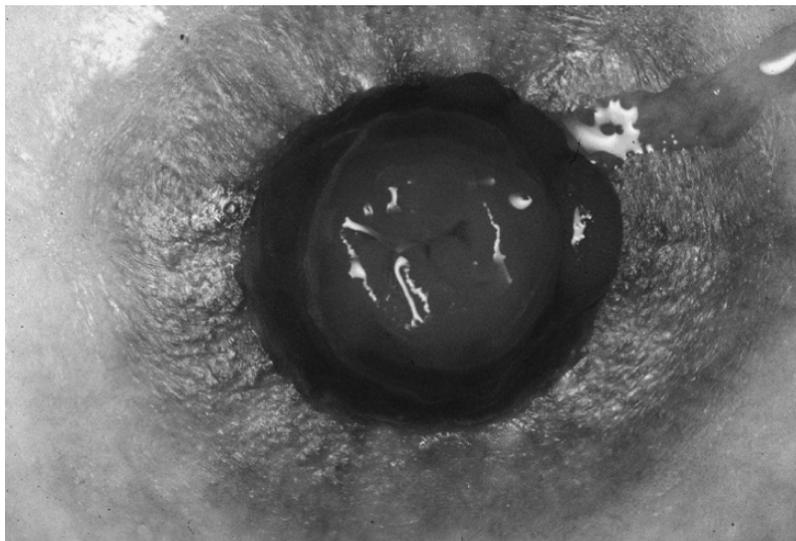


Fig. 5. Portosystemic varices surrounding a stoma within the Roux-en-Y following portoenterostomy.

leading to varices in the stoma and its underlying bowel, and a site of significant hemorrhage (Fig. 5). At the time of initial operation, we use an undivided 35- to 40-cm isoperistaltic retrocolic Roux-en-Y (original Kasai procedure) without stoma diversion. When patients who have undergone initial diversion are encountered, we choose to close all stomas as soon as possible after the first 3 postoperative months, favoring anatomic reconstruction of the Roux-en-Y (33). Early closure prevents the complications of bleeding, and allows intestinal and peritoneal healing prior to any need for transplantation.

Indications for Late Reoperation

Indications for late reoperation are extremely limited. Recurrent cholangitis occurring in a patient with stable and suitable liver function, in the absence of portal hypertension, requires careful evaluation and consideration. Imaging of the liver and biliary structures using high-resolution magnetic resonance cholangiopancreatography (MRCP) and computed tomography (CT) may identify individual patients with partially obstructed biliary ducts who may benefit from modifications of their biliary hilar drainage (Fig. 6). In our experience, careful identification of biliary draining radicals after the anterior wall of the Roux-en-Y is detached from the liver will allow ductal dilation or ductoplasty, similar to sphincteroplasty, and temporary internal ductal stenting. Candidate selection for this procedure should be extremely limited, as most individuals with recurrent cholangitis also have progressive liver disease and portal hypertension, being better served through transplantation. Patients with intrahepatic biliary cysts on “lakes” following Kasai portoenterostomy frequently present with recurring cholangitis. Most represent progressive intrahepatic disease rather than specific ductal obstruction and are best treated by transplantation

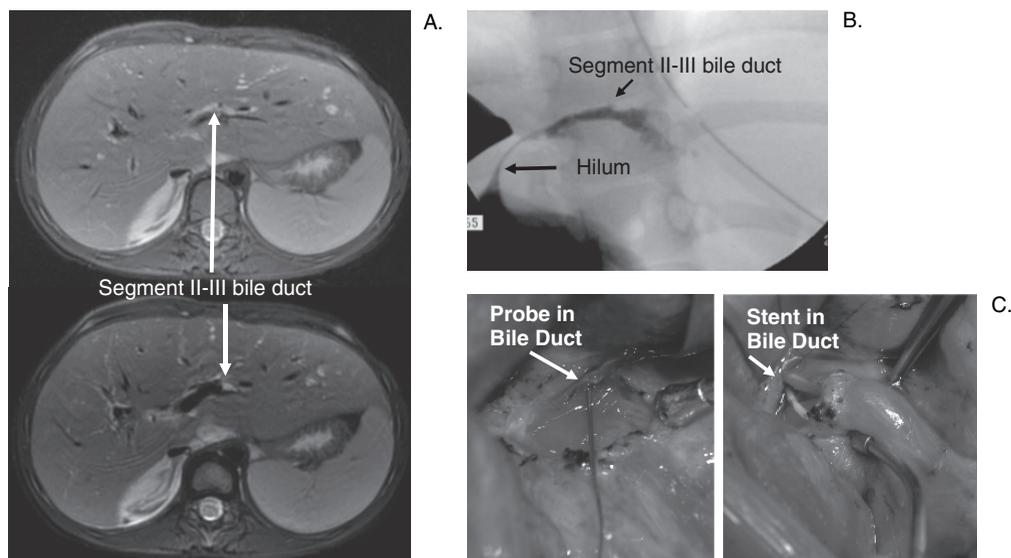


Fig. 6. Dilated segment II/III bile ducts in patient with recurrent cholangitis, 5 years following a successful Kasai portoenterostomy. (A) MRI scan with dilated bile duct; (B) operative cholangiogram showing dilated Segment II/III bile duct; (C) postdilation and stent placement within the intrahepatic bile duct remnant.

CBD INJURY

Clinical Presentation

Cholelithiasis and cholecystitis have been recognized with increasing frequency in the pediatric population. In addition to the common risk factors of obesity, pregnancy, and the use of oral contraceptives, there is a propensity to develop gallstones in patients with hematologic disorders, following ileal resection, and in the setting of long-term use of parenteral nutrition. Laparoscopic cholecystectomy has become routine in the management of such patients regardless of age. Although laparoscopic cholecystectomy is associated with decreased postoperative morbidity compared to open cholecystectomy, CBD injury remains a serious and dreaded complication of the minimally invasive approach, and injury rates approach 1% in some series.

Only approximately 10–20% of bile duct injuries are recognized at the time of initial cholecystectomy, and many remain unrecognized prior to hospital discharge as well (34,35). This diagnostic delay further increases postoperative morbidity and can lead to death. Any patient with mild nonspecific abdominal symptoms following laparoscopic cholecystectomy must be considered to have a potential bile duct injury, particularly in the setting of abnormal white blood cell counts or “liver function” studies. The most common presentation of bile duct injury following cholecystectomy is abdominal tenderness, fever, nausea, emesis, and ileus. These symptoms progress to peritonitis and sepsis when unrecognized. Cholestatic jaundice (depending on the injury type) and leukocytosis are common. Ultrasound examination may be performed, revealing a right upper quadrant fluid collection or dilated bile ducts, although nondilated ducts are the more common finding. CT or MR scanning is superior to ultrasound in the detection of bilomas or abscesses. A hepatobiliary iminodiacetic acid (HIDA) scan can compliment

ultrasound or CT scanning by providing evidence for a complete biliary obstruction or bile leak. Percutaneous drainage of a right upper quadrant collection that yields bilious fluid will confirm an injury. Endoscopic retrograde cholangiopancreatography (ERCP) can localize the site and extent of bile duct injury, and is also useful as a therapeutic modality for nasobiliary stent placement and sphincterotomy in the case of ductal stricture or leak. If ERCP fails to visualize the upper biliary ductal system, such as in the case of a complete obstruction, percutaneous transhepatic cholangiography (PTC) is utilized to visualize the upper tracts.

Clinical Classification

The spectrum of bile duct injuries following laparoscopic cholecystectomy includes biliary leaks, strictures, ligation, and transection. Several classification systems have been proposed to describe this wide array of bile duct injuries. Originating in the time of open cholecystectomy, the Corlette-Bismuth classification system is based on the proximal biliary stump length, but does not allow for description of the type or extent of injury (36). More recently devised, the Strasberg classification system offers a more descriptive and detailed schema: Type A, bile leak from cystic duct or liver bed; Type B, partial occlusion of the biliary tree (often an aberrant right hepatic duct); Type C, bile leak from a duct not communicating with the CBD; Type D, lateral injury to the biliary tree without loss of continuity; and type E, circumferential injury to the biliary tree with loss of continuity (37) (Fig. 7).

Operative Management

If a bile duct injury is detected at the time of laparoscopic cholecystectomy, and the surgeon is not comfortable with biliary reconstruction, dissection should be minimized, drains placed to control leak, and the patient transferred to an institution with sufficient experience in reconstructive hepatobiliary surgery. If biliary reconstruction is considered at the time of initial cholecystectomy, the procedure should be converted to an open operation in order to provide optimal exposure of the biliary system for determination of injury extent and severity. Intraoperative cholangiography is indicated to accurately identify the biliary anatomy. Small lateral ductal injuries can often be repaired primarily over a T-tube. Complete transection of a major bile duct should be repaired with a Roux-en-Y hepatico-jejunostomy, or in circumstances in which there is little associated damage or tissue loss, may be suitable for a primary end-to-end anastomosis over a T-tube. CBD injuries that involve resection of part of the duct are not amenable to primary repair, and in most such cases, adequate drainage with referral for elective reconstruction is advised.

More than 75% of patients with bile duct injuries related to laparoscopic cholecystectomy will present with a delay, ranging from days to months (34,38). Once bile duct injury is identified, the initial management includes control of sepsis with antibiotics, percutaneous drainage if a biloma or leak is present, and decompression of the biliary system by ERCP or by percutaneous transhepatic biliary drainage catheter placement. Following stabilization and treatment of sepsis, classification of the injury and identification of the biliary anatomy should be performed prior to reexploration for reconstruction. MRCP has emerged as a biliary imaging modality that rivals the anatomic detail provided by the more invasive ERCP or PTC (39).

The timing of hepatobiliary reconstruction after delayed presentation of bile duct injury is an important contributor to surgical outcome. In patients identified within

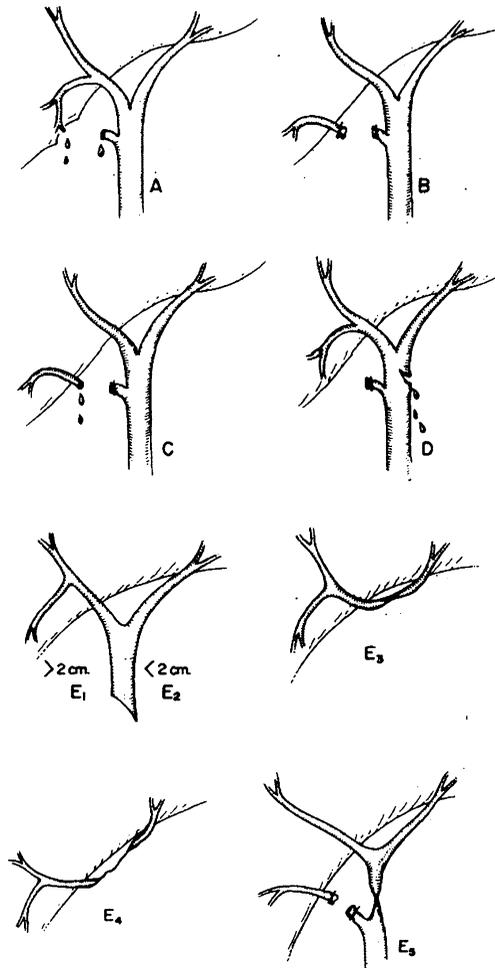


Fig. 7. Strasberg classification system of biliary ductal injuries (37).

1 week of injury, in the absence of associated biliary leak or sepsis, early reconstruction may be performed. However, only lesions distal to the hepatic duct bifurcation should be considered for early repair, as the absence of biliary dilation may make reconstruction technically difficult. If injury involves the hepatic ductal bifurcation, or if significant associated biliary leak or evidence of sepsis is present, delayed repair (after approximately 6–8 weeks) is encouraged. The advantages of delayed repair include resolution of inflammation and infection, demarcation of bile duct ischemia, and a potentially larger bile duct for anastomosis. At the time of elective delayed reconstruction, dense fibrosis is often encountered around the area of prior injury. Mobilization of the proximal duct stump is performed, but division of the hilar plate to lower the hepatic duct remnant may be necessary to avoid dissection between the fibrotic stump remnant and portal vein (40). Reconstruction is performed by Roux-en-Y hepatico-jejunostomy. Temporary internal stenting may be utilized in cases requiring individual reconstruction of the right and left hepatic ducts, or in cases when a percutaneous biliary drainage catheter is already in place (41).

Long-Term Issues

Factors that predict a successful outcome following operative repair of bile duct injuries include a tension-free repair, anastomosis involving a viable and well-vascularized duct, and preservation of the hepatic duct bifurcation. The occurrence of late postoperative strictures months to years following biliary reconstruction is related to several factors: local ischemia, thermal injury caused by electrocautery use near the bile duct, and inflammation and fibrosis owing to bile leak (42). In addition, approximately 50% of patients who undergo end-to-end biliary anastomosis over a T-tube will develop a late stricture. Anastomotic stenosis or stricture can be asymptomatic, or it may present with jaundice, cholangitis, and portal hypertension. In the most advanced cases, the liver dysfunction progresses to secondary biliary cirrhosis and eventual end-stage liver disease. The mainstay of management of late strictures following biliary reconstruction is percutaneous biliary dilation. In one recent study, approximately 60% of patients with biliary-enteric anastomotic stenosis after reconstruction for bile duct injury were successfully managed by dilation and did not require subsequent intervention over a mean follow-up period of 76 months (43). Up to 5% of patients may require eventual liver transplantation after failed operative revision or balloon dilation of the hepaticojejunostomy.

RETAINED COMMON DUCT STONES

Clinical Presentation

Cholelithiasis is uncommon in the pediatric population, with an incidence of approximately 6–13% in children with cholelithiasis (44–46). Congenital CBD anomalies, such as choledochal cysts, pancreatobiliary maljunction, and preduodenal position of the CBD, are often found in association with cholelithiasis in this population of patients. In adults undergoing CBD exploration for cholelithiasis at the time of cholecystectomy, the incidence of retained CBD stones postoperatively ranges from 8% following laparoscopic cholecystectomy to 14% following open cholecystectomy (47,48). The overall incidence of retained CBD stones following laparoscopic cholecystectomy in adults is approximately 0.7%. The incidence of retained CBD stones in children remains unknown at this time. Furthermore, there is little consensus with regard to the management of this problem in the pediatric population, and suggested guidelines have been applied based on the experience in adults.

Cholelithiasis can present with a variety of symptoms, and diagnosis on the basis solely of history and physical examination is often unreliable. Symptoms of intermittent right upper quadrant pain, often postprandial, with associated nausea and emesis, and a history of fatty food intolerance, are suggestive of biliary tract pathology, but are nonspecific. The spectrum of clinical presentations of cholelithiasis includes obstructive jaundice, cholangitis, and acute pancreatitis, and in advanced disease, CBD stones may present with sepsis. The classic triad of right upper quadrant pain, jaundice, and fever and chills, known as Charcot's triad, is highly suggestive of cholangitis, and cholelithiasis is a common predisposing cause. When cholangitis becomes severe, patients may also present with hypotension and mental status changes, known as Reynold's pentad.

Diagnostic Evaluation

In patients who present following cholecystectomy with symptoms suggestive of retained CBD stones, a complete blood count, “liver function” studies, amylase, and lipase should be evaluated. Elevations in alkaline phosphatase, GGT, and conjugated bilirubin are highly suggestive of a retained common duct stone. It should be noted, however, that the negative predictive value of normal laboratory studies in choledocholithiasis is low, and a high index of suspicion must be maintained based on clinical presentation (49). The white blood cell count and serum amylase level may be elevated in choledocholithiasis, even in the absence of cholangitis and pancreatitis.

In patients in whom a retained common duct stone is suspected following prior cholecystectomy, transabdominal ultrasound may be performed as an initial radiologic tool. In the appropriate clinical setting, the presence of a dilated biliary system on ultrasound is highly suggestive of a retained common duct stone, and in some cases, a CBD stone may be identified (Fig. 8). A CBD diameter of 10 mm or greater on ultrasound examination has been found to be highly prognostic of choledocholithiasis (50). The accuracy of transabdominal ultrasound in the diagnosis of retained common duct stones is closely related to the experience of the examiner (51).

Magnetic resonance cholangiography (MRC) and helical computed tomography cholangiography (HCT-C) are more sensitive for the diagnosis of CBD stones. MRC is noninvasive and not operator-dependent, and its sensitivity and specificity in the diagnosis of common duct stones range from 88–98% and 84–100%, respectively (52–55). Helical CT scanning is another noninvasive option; however, risks of this

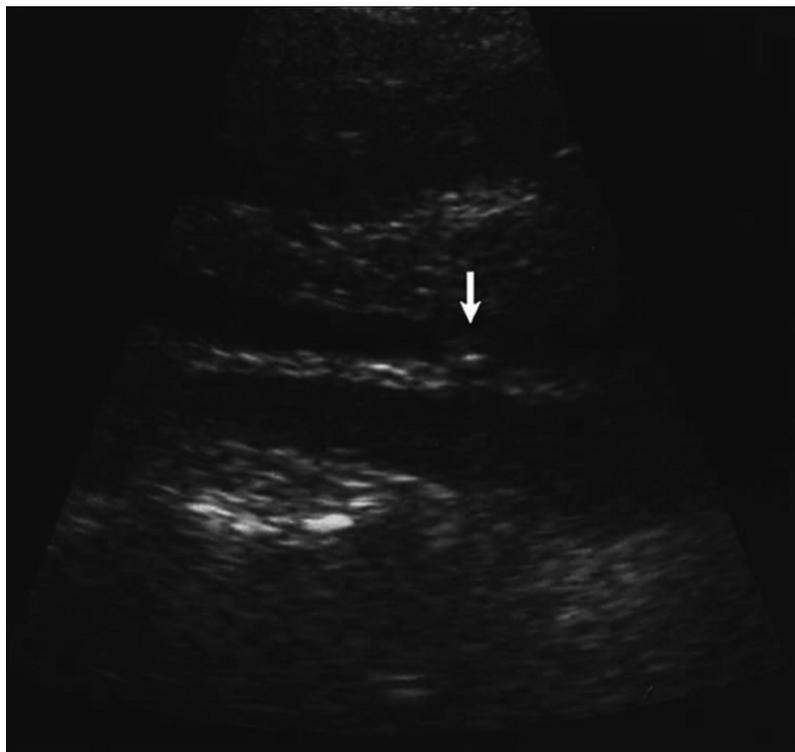


Fig. 8. Transabdominal ultrasound demonstrating gallstone (arrow) in the CBD.

modality include radiation exposure and allergic reaction to intravenous contrast material. Its sensitivity (85–88%) and specificity (88–96%) are similar to MRC, and this modality may be considered in the diagnosis of choledocholithiasis in centers without MR capability (54,56,57).

ERCP remains the gold standard in the diagnosis of choledocholithiasis, and is also an important therapeutic modality for the management of retained CBD stones (Fig. 9). Both the sensitivity and specificity of ERCP in the detection of choledocholithiasis range from 95–100% in the adult population (58,59). A disadvantage of ERCP is its invasiveness and risk of complications, including bleeding, perforation, pancreatitis, infection, and adverse events related to sedation and anesthesia. These factors have led to the recommendation that MRC be used when there is a low index of suspicion for choledocholithiasis, and ERCP be used in patients with a high likelihood of having common duct stones (60). Despite the low incidence of biliary disease in the pediatric population and limitations related to equipment size, ERCP success rates and complication rates are comparable in children compared to adults (61,62).

Management

The initial step in the management of patients with retained CBD stones following cholecystectomy is the initiation of antibiotic therapy if cholangitis is present. Antibiotic coverage should consist of an agent that is active against enteric microorganisms, as

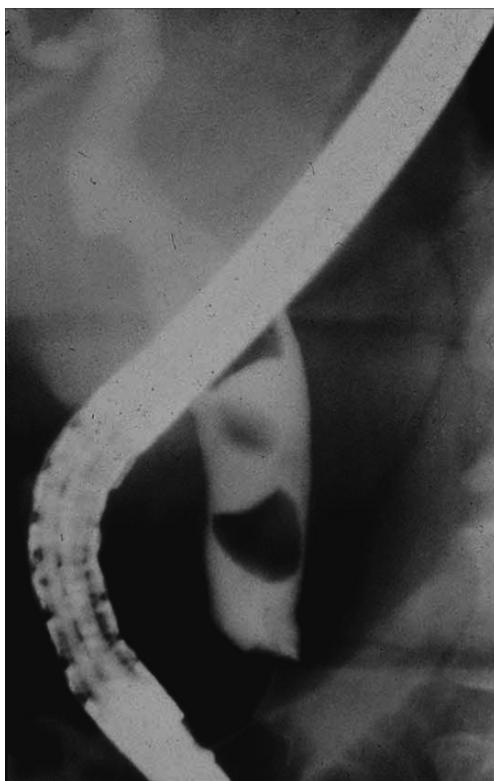


Fig. 9. Endoscopic retrograde cholangiogram demonstrating choledocholithiasis and CBD dilation.

the bacteria that are typically identified include *Escherichia coli*, *Klebsiella* species, *Clostridium* species, *Streptococcus faecalis*, and *Enterobacter* species.

In patients who present post-cholecystectomy with retained common duct stones, decompression of the biliary tree is critical, and must be performed urgently in the setting of cholangitis, pancreatitis, or sepsis. The condition of the patient will often dictate the best approach to acute ductal decompression. In patients who are clinically stable without pancreatitis, ERCP can be considered in experienced hands. Patients suffering from clinical instability from sepsis are best addressed by PTC and external stent placement. This allows access to the obstructed biliary tree and decompression, allowing the treatment of sepsis and pancreatitis prior to instrumenting the ampulla of Vater and injecting the pancreaticobiliary ductal system.

ERCP with endoscopic sphincterotomy and stone extraction is commonly performed for the acute treatment of the obstructed duct in stable patients. Several techniques have evolved for stone extraction, including balloons, baskets, and lithotripsy devices. Using conventional endoscopic techniques, the success rate for the complete clearance of the CBD is approximately 90% (63). The largest diameter stone that can be retrieved is dependent on the size of the distal CBD and the sphincterotomy (64). Large stones require fragmentation by lithotripsy techniques (mechanical, laser, or electrohydraulic) prior to extraction (65,66).

If retrieval of a retained common duct stone fails by endoscopic techniques, a second option is to return to the operating room for a laparoscopic or open CBD exploration (CBDE). CBDE may be performed via a transcystic approach, in which Fogarty balloon catheters and Dormia wire baskets may be advanced into the distal common duct via the cystic duct stump. Intraoperative fluoroscopy is often used as an adjunct, and if these maneuvers fail, then they may be repeated under direct vision with passage of a choledochoscope also placed via a transcystic approach. The ducts may be forcefully irrigated with saline to promote passage of small stones and debris, and large stones (>1 cm) may require fragmentation by lithotripsy techniques. Common hepatic or intrahepatic duct stones are difficult to retrieve via the transcystic approach because of the acute angle between the cystic duct and common hepatic duct. Transcystic CBDE is successful in stone retrieval in more than 80% of patients (67–69).

CBDE may also be performed via a choledochotomy, and this approach is often used in the setting of large stones, a CBD that is greater than 10 mm in diameter, or a cystic duct that cannot be dilated large enough to allow passage of the choledochoscope. This approach allows more effective stone clearance of the biliary tree proximal to the insertion of the cystic duct, and probably is a better option for the more difficult cases of retained stones, such as a stone impacted at the ampulla. A longitudinal choledochotomy is made as small as possible, the choledochoscope is inserted, and similar techniques described for transcystic CBDE are utilized to remove stones (70) (Fig. 10). Antegrade sphincterotomy may be performed if necessary through the choledochoscope (71,72). The technique of closure of the choledochotomy is critical in order to avoid late biliary stricture. Closure of the CBD is performed using interrupted absorbable 5.0 or 6.0 monofilament suture, and the use of a T-tube remains controversial (73,74).

Overall, the success rate of laparoscopic CBD clearance ranges from 73–100%, and the literature suggests that there is no significant difference in success rate between the transcystic versus choledochotomy approach (75,76). Conversion to an open CBDE is required in approximately 5% of cases. A cystic duct tube or a T-tube choledochotomy closure may be performed in cases of incomplete clearance of the bile duct, and

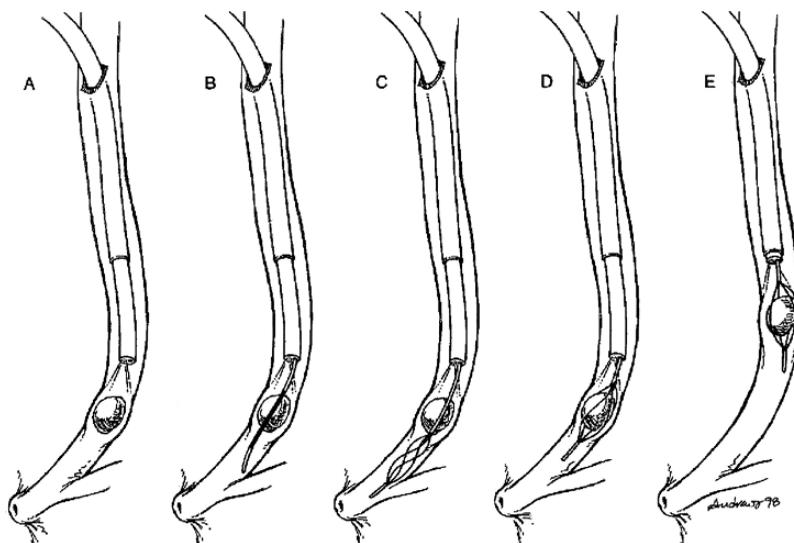


Fig. 10. CBD stone extraction under direct vision by a wire basket advanced through a choledochoscope (70).

postoperative endoscopic or percutaneous approaches may be undertaken. If later extraction is necessary, suitable healing of the T-tube tract is necessary prior to attempts at extraction to prevent intraperitoneal leakage of bile. The risks of laparoscopic CBDE include bile leak, pancreatitis, bile duct injuries, and cholangitis. It should be stressed that if ERCP for a retained common duct stone has failed and the surgeon is not technically able to perform a laparoscopic CBDE, open CBDE is a viable option. The majority of data published regarding CBDE has been from studies in adults; however, one recent series in the pediatric population reported successful clearance of common duct stones in two-thirds of children undergoing laparoscopic CBDE (77). Nevertheless, the small caliber of the common duct in children provides a significant challenge in the management of retained common duct stones by CBDE.

Percutaneous transhepatic therapies are a third option for clearance of the CBD of stones (78). Under ultrasound guidance, the biliary tract is accessed percutaneously, and a balloon catheter is advanced into the CBD. Lithotripsy, stone extraction, and an antegrade sphincterotomy may be performed by this approach to clear the common duct. If multiple sessions are required to clear the duct, a percutaneous biliary drainage catheter is left in place to decompress the system.

On rare occasion, such as nonremovable impacted distal CBD stones, surgical biliary bypass may be required to decompress the system. Options include choledochoduodenostomy and choledochojejunostomy.

When a common duct stone is detected intraoperatively, CBDE as described previously may be performed for clearance. However, in a subset of patients, it is not possible to clear the duct of stones. In such scenarios, postoperative ERCP with sphincterotomy and stone retrieval is the procedure of choice.

Long-Term Issues

CBDE is a technically demanding procedure, particularly via the laparoscopic approach. Extensive manipulation of the CBD and laparoscopic suturing of the duct

may result in late complications, such as CBD stricture. Factors that may contribute to late stricture formation following CBDE include devascularization of the duct during dissection and the creation of a choledochotomy in a common duct that has a diameter of less than 7 mm. Studies have also indicated that late strictures may occur following endoscopic sphincterotomy for the management of choledocholithiasis, and the very long-term outcome of patients who have undergone sphincterotomy is not yet known (79). Strictures following CBDE or endoscopic sphincterotomy are typically addressed by endoscopic balloon dilation and biliary stent placement.

Other groups have reported a small incidence of late recurrent CBD stones following laparoscopic CBDE (80–82). However, on the basis of high initial success rates in complete duct clearance and the low rate of morbidity following laparoscopic CBDE (ranging from 3–7%), these authors have strongly advocated this “single-stage” approach to managing choledocholithiasis detected intraoperatively at the time of cholecystectomy.

Although these evolving techniques have shown good success in experienced hands, the potential for injury and a poor outcome is high when these advanced laparoscopic skills are not well developed. It is incumbent on the operating surgeon to honestly assess their personal experience and address these difficult cases in the fashion that allows for the greatest chance for an initial excellent outcome, regardless of the nature of the surgical approach and exposure.

PORTAL HYPERTENSION

Portosystemic Shunts

Numerous surgical procedures have been devised to divert portal blood into the low-pressure systemic venous circulation, thereby decreasing the portal venous pressure. Enthusiasm for the use of portosystemic shunting in children was limited by early reports suggesting that children less than 8 years of age, and those with vessels for the shunt anastomosis less than 8 to 10 mm, would be unsuitable candidates because of the high risk of shunt thrombosis (83). In addition, Voorhees et al. suggested a high incidence of neuropsychiatric disturbances following nonselective shunts in children (84). Recent experience in centers skilled in pediatric vascular reconstruction has established that a high rate of success can be achieved with minimal complications even in small pediatric patients. To achieve these goals, we have found that the following principles should be followed: (1) anastomosis should be constructed using fine (6.0–7.0) monofilament sutures with provision for growth; (2) sufficient mobilization of vessels is necessary to prevent kinking or twisting of the shunt after the viscera are returned to their normal location; (3) predivision marking of the vessel wall along the long axis will prevent unappreciated rotation of the vessel prior to the anastomosis, (4) postreconstructive venography should be performed in order to ensure division of all collaterals and adequate shunt flow; (5) selective postoperative anticoagulation should be performed and antiplatelet drugs administered; and (6) the surgical team should be skilled at pediatric portal vascular reconstruction.

Primary Surgical Options

In general, portosystemic shunts can be classified into three groups: non-selective and selective shunts, and direct reconstructions of the portal circulation using vascular grafts (shunts).

Non-selective shunts are constructed to communicate with the entire portal venous system, and therefore have the potential to divert blood from the normal antegrade perfusion to the liver. Historically, the most commonly used shunt in children was the mesocaval shunt (Clatworthy Shunt), where the distal inferior vena cava (IVC) was ligated, divided, and its proximal portion was then anastomosed to the side of the superior mesenteric vein (SMV). This shunt has now been replaced by the H-graft mesocaval shunt, which is constructed using a short segment of internal jugular vein or, less frequently, ringed Gore-Tex graft material is used to connect the SMV and the IVC, retaining the advantage of a larger vessel for the anastomosis, and avoiding ligation of the IVC (85) (Fig. 11). Excellent patency (93%) and no episodes of encephalopathy support its use in pediatric patients. The limited intraabdominal dissection needed to complete this shunt contributes to its technical ease, and if liver transplantation is needed, the shunt can be easily occluded at that time. Other nonselective shunts have significant disadvantages in children because of the need for splenectomy (proximal splenorenal), or dissection of the main portal vein, which compromises liver transplantation (end-to-side, side-to-side portocaval shunts).

Selective shunts are constructed to divert the “gastrosplenic” portion of the portal venous flow into a systemic vein, most frequently the left renal vein or the immediately adjacent IVC. Communication between the “central” mesenteric portal circulation, which perfuses the liver, and the gastrosplenic portal circulation is severed by dividing the gastroepiploic veins, the coronary vein, and the retroperitoneal pancreatic collaterals (86). The most common and successful selective shunt, the distal splenorenal shunt (DSRS) (Warren Shunt), preserves antegrade perfusion to the liver within the mesenteric portion of the portal circulation, while decompressing the esophageal venous

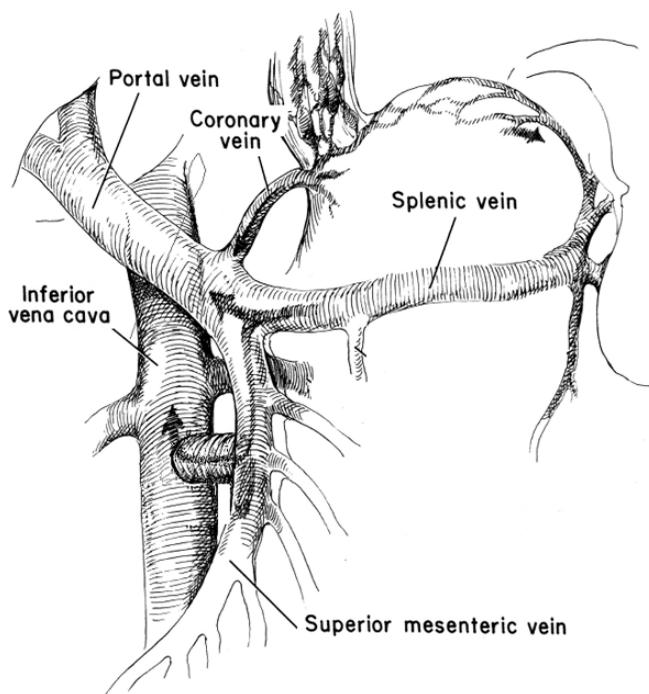


Fig. 11. H-graft Mesocaval Shunt, depicted using internal jugular vein graft (89).

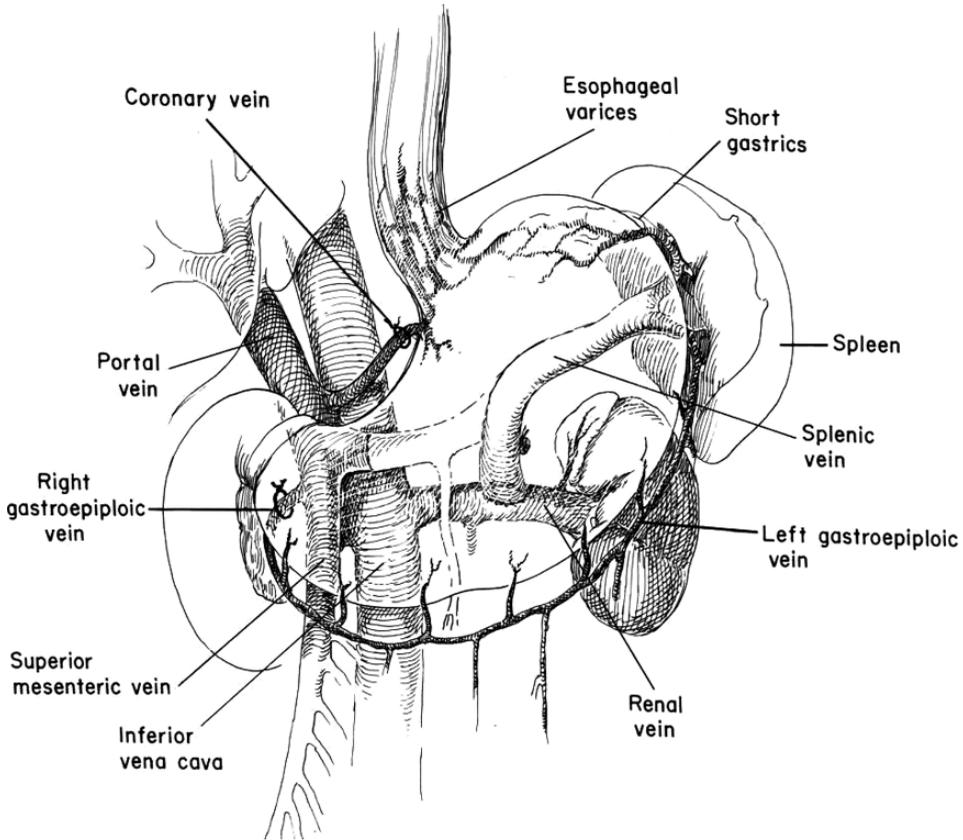


Fig. 12. Distal splenorenal (Warren) shunt (89).

plexus through the short gastric veins and splenic vein (Fig. 12). We use this shunt as our primary option in children where direct reconstruction is not possible. When the adrenal vein is appropriately located and dilated, it serves as an alternative anastomotic site to access the left renal vein (87,88). When performed in centers experienced in complex vascular reconstruction of the portal system, as is necessary in pediatric liver transplantation, shunt patency has ranged from 83–100% (86,89–91).

Direct reconstruction of the portal circulation in children with extrahepatic portal vein obstruction into the left branch of the portal vein represents the ideal solution (92–95). This mesenterico-portal shunt (REX Shunt) reestablishes normal portal inflow into the intrahepatic portal vein, using either an interposition jugular venous graft or the dilated coronary vein (Fig. 13). Candidates for this procedure should fulfill three conditions: the liver parenchyma must be normal, they must not have a hypercoagulable state, and the umbilical portion of the left hepatic vein must be accessible and patent. Patients with diffuse intrahepatic portal vein thrombosis are not good candidates for this reconstruction. Doppler ultrasound and direct portography would suggest, however, that at least two-thirds of children with extrahepatic portal venous obstruction (EPVO) have sufficient left portal vein patency to undergo this procedure. Experience with this shunt is less extensive. Initial patency seems excellent; however, later revision secondary to stenosis or occlusion is somewhat more common (92–98). Although this is a more complex technical challenge, in these selected patients, this option should be

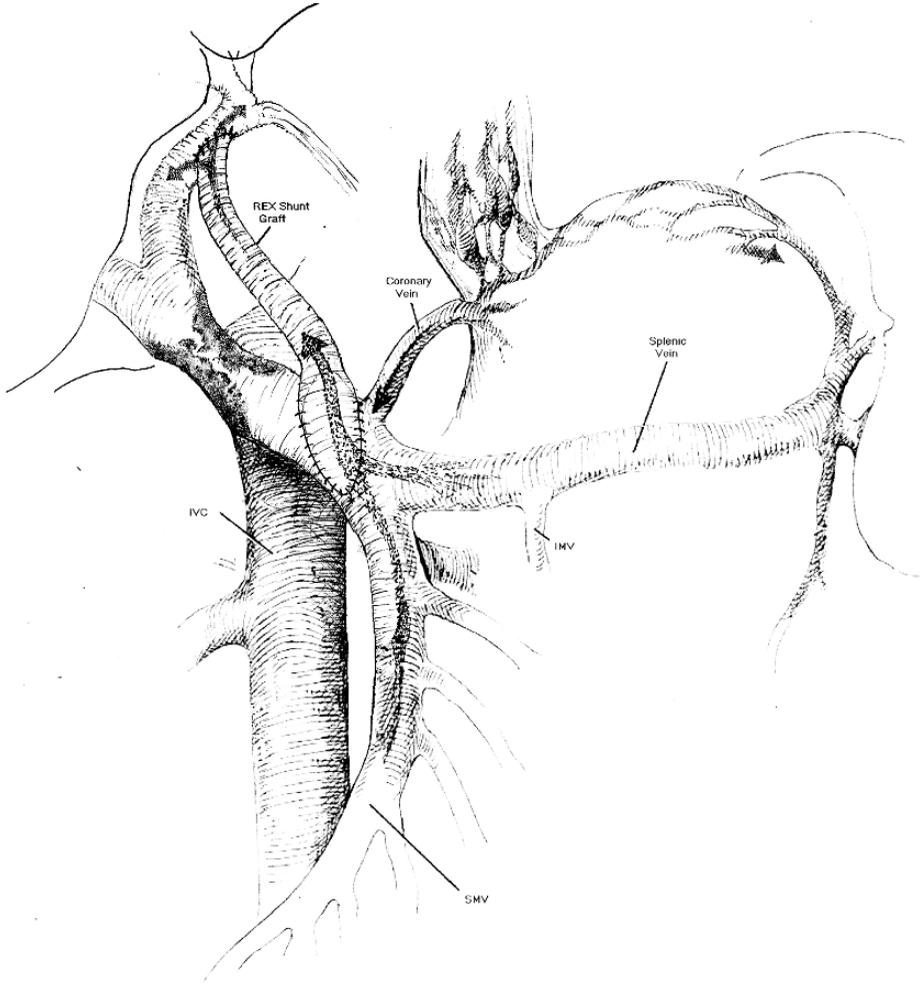


Fig. 13. REX mesenteric portal venous reconstruction (89).

used in preference to other true “shunts” that divert portal blood rather the reconstruct the normal portal flow patterns.

Indication for Reoperation

The most common technical problem encountered following portacaval shunt construction is shunt thrombosis. When the reconstructive techniques outlined previously are carefully followed, this is fortunately an uncommon event occurring in less than 5% of all children undergoing primary portacaval shunting. Because of the technical complexity of the Rex shunt, this appears to be slightly more common in these patients.

Appreciation of postoperative shunt thrombosis can occur either through routine Doppler ultrasound surveillance or less commonly postoperative bleeding. It is our practice to do routine immediate postoperative ultrasound with Doppler vascular studies to assure adequate blood flow following abdominal closure and periodic surveillance during the first postoperative week.

When approaching a patient with documented thrombosis, a thorough reevaluation of the most likely responsible factors should be undertaken. The first of these factors is vascular inflow. Because portal flow is substantial, it is very uncommon to have a limitation of portal inflow as a primary etiology of shunt thrombosis. Unique circumstances such as patients with diffuse multifocal mesentery venous thrombosis may limit portal inflow but are rarely the primary factor inducing thrombosis. Appropriate assessment of inflow at the time of primary shunt placement should avoid this rare circumstance. However, disturbance of shunt in flow rate because of twisting or kinking within the shunt itself is a significant contributor to early shunt thrombosis. This technical abnormality is best prevented by careful orientation of the vessels prior to undertaking and the initial anastomosis. It is our practice to mark the longitudinal surface of any vein or graft that is used so that appropriate orientation can be maintained at the time of the anastomosis. In circumstances where kinking or a rotational twist of the vessel has occurred, immediate thrombectomy and revision of the anastomosis to remove this obstruction is necessary. It is important to carefully survey the shunt following the initial anastomosis to assure that subtle rotational abnormalities have not occurred. In circumstances where an extremely difficult anastomosis is anticipated, such as the left portal vein anastomosis with the Rex shunt, we find that construction of the distal anastomosis first allows more successful orientation of the interposition graft prior to completing the less complex proximal portal anastomosis at the level of the mesentery. In cases where a difficult distal anastomosis would be difficult to revise, the free portion of the mobilized splenic or mesenteric vein can be divided, rotated, and reanastomosed at a safer location within the shunt. Although this is rarely necessary, rotation and reconstruction of the shunt in a safer middle position of the vessel is preferential to attempting to revise or redo a delicate and technically complex distal anastomosis.

The second technical factor related to shunt thrombosis occurs at the level of the anastomosis. Anastomotic stenosis or poor endothelial approximation contribute to thrombosis in portacaval shunts as in all vascular reconstructive procedures. Again, prevention at the time of the primary operation plays a key role. The thin vessel wall and extreme flexibility of the portal venous system make it uniquely prone to anastomotic stenosis unless specific technical points are followed. In addition, a provision for growth should be incorporated into each of these anastomoses in children. Meticulous attention to these technical details will substantially decrease the need for anastomotic revision. We follow several principles in constructing or critical venous anastomoses: (1) we use monofilament 6.0 or 7.0 sutures for all portal anastomoses; (2) if we use a running suture for the difficult to access posterior anastomotic suture line, we apply lateral tension on both lateral ends of the anastomosis at the time the sutures is tied to prevent "telescoping" of the suture line, which creates posterior wall shortening and stenosis; (3) in all patients where growth is anticipated, the anterior anastomotic suture line is constructed using interrupted sutures. In cases where the anastomotic contour is unclear following reconstruction, an intraoperative venogram may be helpful.

The final factor that may compromise shunt flow and hence precipitate thrombosis is shunt outflow. The patients with highest risk for elevated post shunt venous pressure are those undergoing Rex reconstruction. In these cases, initial capacitance of the portal veins within the liver is somewhat unpredictable and often limited. Studies have documented rapid and substantial improvements in the outflow portal venous size and

capacity following the Rex shunt, but this initial limitation of outflow undoubtedly contributes to the risk of early thrombosis in this operation. Portal venography of the intrahepatic portal vein may be helpful at the time of initial operation in assessing this capacity; however, it does not appear to be predictive of success or risk of thrombosis. In patients undergoing distal splenorenal or H-graft mesocaval shunts, outflow pressure is determined by central venous pressure within the IVC. Unique circumstances causing IVC pressure to be elevated can compromise outflow. These include patients with a cirrhotic compression of the retrohepatic IVC, a condition most commonly seen with Budd-Chiari syndrome. In this unique circumstance, caval-atrial bypass may be necessary as a second component to portacaval shunting. Children with elevated right heart pressures secondary to congenital heart disease can present a similar challenge. In these patients, control of the central venous pressure is best undertaken through improved medical management of their cardiac disease.

When portal reconstruction is undertaken respecting the previously described principles, postoperative anticoagulation is rarely necessary. Our present protocol includes the use of postoperative antiplatelet treatment with low-dose aspirin for 30 to 90 days. Direct anticoagulation using intermittent subcutaneous or intravenous heparin is restricted to patients with preexisting hypercoagulable states until their baseline anticoagulation can be reestablished. In addition, in patients where shunt reconstruction was necessary because of initial thrombosis, intravenous anticoagulation for several postoperative days may be undertaken at the surgeon's discretion. All patients with primary portal/mesenteric venous thrombosis and those with postoperative thrombosis should undergo a complete evaluation of their coagulation system to identify hypercoagulability and institute appropriate treatment prior to shunting.

Transjugular Intrahepatic Portosystemic Shunt (TIPS)

The introduction of TIPS has added another therapeutic option for the physician confronted with complex portal hypertension. This procedure uses interventional radiographic techniques to place an intrahepatic expandable metallic shunt between a portal vein branch and the hepatic vein, forming a central nonselective portocaval shunt. The procedure is undertaken using access via the right internal jugular vein. The hepatic veins are identified and a fluoroscopic/ultrasonographic-guided puncture from the hepatic vein into an intrahepatic portion of the portal vein is undertaken. This tract is then dilated and an expandable mesh stent is placed, forming a communication between the intrahepatic portal vein and the hepatic vein branch (Fig. 14).

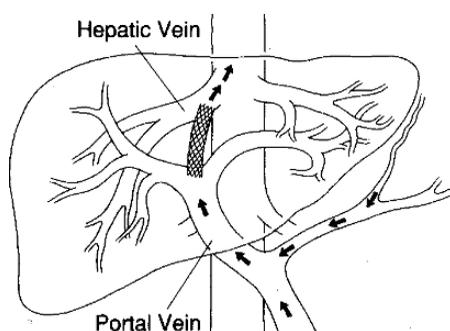


Fig. 14. TIPS (transjugular intrahepatic portosystemic shunt) (89).

Technical difficulties in establishing a safe but large enough tract for sufficient shunt flow limit the usefulness of this procedure in infants. In children with biliary atresia, the close proximity of the biliary Roux-en-Y conduit to the portal vein and the often diminutive size of the portal vein increase the risk of stent perforation, malposition, or perforation (89). This procedure has great benefit in the control of refractory portal hypertensive bleeding unresponsive to the pharmacologic methods, and in patients needing temporary portal decompression prior to liver transplantation (99). The ability to embolize bleeding varices from the coronary vein at the time of TIPS placement assists in achieving primary control of bleeding sites.

The two principal long-term complications of TIPS are encephalopathy and shunt occlusion. Being a central nonselective shunt, this procedure can precipitate hepatic encephalopathy, especially when used in patients with severe intrinsic liver disease. The overall risk of encephalopathy ranges from 5–35% in adult patients, a rate similar to that seen with side-to-side surgical shunts. Most episodes of encephalopathy can be controlled with dietary protein restriction and lactulose administration. Postprocedural encephalopathy seems to be less common in children, although limited clinical experience and the difficulties in diagnosing subtle encephalopathy in children makes this observation tentative. Selection of a shunt size to allow sufficient portal decompression without shunting excessive amounts of blood from the liver is also a theoretical solution.

Stenosis of the shunt or shunt thrombosis remains a major complication following TIPS. Shunt stenosis occurs in 25–75% of cases, with shunt patency decreasing with the length of time that the shunt is in place. Intimal hyperplasia or incorrect shunt placement most commonly causes the stenosis. Regular monitoring for shunt patency and periodic shunt dilation or restenting is necessary (100). Shorter stent lengths and smaller diameter stents have been constructed for pediatric applications; however, the risks of hepatic perforation and stent malposition are greater in small patients. The complications of shunt stenosis are equally problematic, and patient growth over time may cause the initial shunt to be too short, requiring revision or restenting to maintain access to both the portal and hepatic venous circulation. These limitations and risks make TIPS a reasonable and suitable treatment for acute unresponsive variceal hemorrhage in children with established intrinsic liver disease, often while awaiting liver transplantation. It is particularly helpful when used as a bridge to achieve stability by controlling refractory hemorrhage in patients awaiting liver transplantation. Long-term decompression is better achieved through surgical shunts at the present time, and TIPS is not indicated in patients with EPVO.

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Reoperation of the Pancreas

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INTRODUCTION

Pancreatic pathology is rare in children. The most common neonatal diagnosis that may require surgical intervention is persistent hyperinsulinemic hypoglycemia of infancy (PHHI). Fortunately, this disease is relatively uncommon, with an incidence of one in 50,000 newborns per year (1). In addition, many patients with this diagnosis can be adequately treated with aggressive medical therapy. Pancreatic neoplasms occur in children but are also quite rare. At St. Jude Children's Research Hospital, 890 newly diagnosed patients with solid organ tumor were seen over the last 5 years. Only two of these patients had pancreatic tumors. Only one had a malignant tumor. Traumatic injury is the most common reason surgical intervention is required on the pancreas of a child. Accidental pancreatic injury is rare in infants, so child abuse must be considered in young children who present with injury in this location. Severe pancreatic trauma is most common in males over the age of 6 years old. These patients are often cared for by adult trauma surgeons. Epidemiology and personal experience, therefore, suggest that most pediatric surgeons will only operate on the pancreas sporadically throughout their careers. One result is that literature concerning management of many of the complications associated with pancreatic diagnoses and pancreatic surgeries in children is quite limited, and many recommendations for management of pancreatic problems have been developed from more extensive experience with adult patients, who suffer from different disease processes.

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Reviewing the combined experience at our institution over the course of the last 5 years, we have performed over 17,000 pediatric surgical procedures. Only seven of those procedures have been pancreatic operations. Five of the seven procedures performed were for traumatic injuries.

In this chapter, we will discuss management options for the following pancreatic complications in children: persistent hypoglycemia after pancreatectomy for PHHI, traumatic pancreatic pseudocyst, recurrent pancreatitis, and pancreatic fistula.

PERSISTENT HYPOGLYCEMIA AFTER PANCREATECTOMY FOR PHHI

Neonatal hypoglycemia is a frequently observed problem that requires quick diagnosis and effective treatment in order to prevent seizures and neurological sequelae. With the exception of prolonged fasting, hyperinsulinism is the most common cause of hypoglycemia in infancy. Transient hyperinsulinism can be caused by maternal factors, such as maternal diabetes, intravenous glucose administration during labor and delivery, and maternal use of medications (oral hypoglycemics, terbutaline, propranolol). Perinatal stress from factors such as birth asphyxia, maternal toxemia, preeclampsia, or prematurity can also cause prolonged hyperinsulinism. Persistent hyperinsulinism is caused by congenital endocrine or metabolic disorders, of which PHHI is one etiology. Other etiologies include hypopituitarism, adrenal insufficiency, and Beckwith-Wiedemann syndrome. PHHI represents a group of clinically, genetically, and morphologically heterogeneous disorders characterized by dysregulated insulin secretion, resulting in severe and persistent hypoglycemia (2). It is an uncommon diagnosis with an incidence of one in 30,000 to 50,000 live births (1).

Five proteins have been shown to cause PHHI when mutations occur: the sulfonylurea receptor 1 (SUR-1), Kir6.2 (potassium inward rectifying channel), glutamate dehydrogenase, glucokinase, and mitochondrial enzyme short-chain-3-hydroxyacyl-CoA dehydrogenase (2). SUR-1 and Kir6.2 combine to form the β -cell adenosine triphosphate (ATP)-sensitive potassium channel (K_{ATP} channel). The K_{ATP} channel is inhibited by sulfonylurea drugs (used therapeutically to stimulate insulin secretion in type 2 diabetes) and activated by diazoxide (the main medical treatment for PHHI). Loss of function mutations in the K_{ATP} channel secondary to mutations in SUR-1 or Kir6.2 result in K_{ATP} channel hyperinsulinism—the most common and severe form of PHHI. Most of these mutations are autosomal recessive in nature, although autosomal dominant forms have been reported. These patients usually present in infancy with symptoms secondary to hypoglycemia, such as seizures, lethargy, and loss of consciousness. Patients are usually macrosomic at birth. Typically, affected patients do not respond to medical treatment and require pancreatic resection. There are two distinct histological forms of K_{ATP} hyperinsulinism—focal and diffuse. Focal disease accounts for 30–60% of cases of hyperinsulinism that progress to surgery, and identification and excision of focal lesions is curative in these patients (3). Patients with diffuse PHHI require 95% pancreatectomy to control their hyperinsulinism.

The second most common form of PHHI, also known as hyperinsulinism and hyperammonemia syndrome, is caused by a gain-of-function mutation in mitochondrial glutamate dehydrogenase (GDH). The mutation reduces the sensitivity of the enzyme to inhibition by guanosine triphosphate (GTP), resulting in excessive flux through GDH and excessive insulin release. In the liver, increased GDH activity leads to excessive

ammonia production and impaired urea synthesis. These patients present with recurrent episodes of hypoglycemia and asymptomatic hyperammonemia. A less frequently seen form of PHHI is caused by an activating mutation in β -cell glucokinase, which results in increased affinity of glucokinase for glucose, closure of K_{ATP} channels and inappropriate insulin secretion. Mutations in both GDH and glucokinase have an autosomal dominant inheritance pattern and represent a milder form of PHHI (2,3). Affected patients usually are not macrosomic at birth. They usually present at 6–9 months of age and generally respond to medical therapy.

Recently, a mutation in the gene encoding the mitochondrial enzyme short-chain 3-hydroxyacyl-CoA dehydrogenase has been found to be associated with congenital hyperinsulinism. This is an autosomal recessive disorder, can present early in the neonatal period or late with mild hypoglycemia, and generally responds to medical therapy with diazoxide (2).

The diagnosis of hyperinsulinism is made when the circulating insulin concentration exceeds $5 \mu\text{U/mL}$ while the glucose concentration in the same sample is less than 45 mg/dL (3). Genetic testing can be performed to identify four of the five genes known to be associated with PHHI through commercial labs (2). The initial goals of management for these patients are: (1) to prevent brain damage from recurrent hypoglycemia; (2) to establish a specific diagnosis and therapy; and (3) to encourage normal feeding behavior while assuring safe fasting tolerance. The first step in management is correction of hypoglycemia with the goal of maintaining plasma glucose levels above 70 mg/dL . This process often requires placement of central venous access and continuous infusion of glucose solution. Once PHHI is diagnosed, medical treatment is instituted with diazoxide (starting with $5\text{--}15 \text{ mg/kg/day}$ (2) to a maximum of $15\text{--}20 \text{ mg/kg/day}$ (4)) with concomitant use of a diuretic (i.e., chlorothiazide or furosemide) to prevent fluid retention. The second line of medical therapy for patients minimally responsive to diazoxide is octreotide ($5\text{--}20 \mu\text{g/kg/day}$ (2), $25\text{--}100 \mu\text{g/kg/day}$ (4)). Glucagon (1 mg/day) has also been used as an adjunct to help maintain euglycemia (2). Although not routinely used, there have been reports of successful treatment of PHHI with nifedipine ($0.3\text{--}0.8 \text{ mg/kg/day}$ dosed orally four times a day) (4,5).

Failure of medical therapy, commonly referenced but often undefined, can range from recurrent episodes of hypoglycemia and seizures during treatment to an inability to safely control glucose levels in an outpatient setting. It is the definitive indication for pancreatic resection. The extent of pancreatic resection for PHHI refractory to medical treatment differs depending on whether the patient has focal or diffuse PHHI. Unfortunately, this diagnosis is difficult to make preoperatively. Traditionally, the literature has recommended performing preoperative pancreatic venous sampling or intraarterial calcium stimulation to help define and locate focal disease. However, both these procedures have only modest success and are technically difficult and highly invasive. More recently, positron emission tomography (PET) scans with fluorine-18 L -3,4-dihydroxyphenylalanine (^{18}F -fluoro- L -DOPA) (with concurrent magnetic resonance imaging [MRI] for anatomical correlation) have been shown to accurately discriminate between focal and diffuse PHHI (6). Although this is a relatively new procedure, it shows promise for future use in the preoperative diagnosis of focal versus diffuse PHHI.

Differentiation of focal versus diffuse PHHI can also be made at surgery by intraoperative biopsies. Suchi et al. (2004) published a series of 52 patients who underwent surgery for persistent hyperinsulinism (7). At surgery, the pancreas was inspected for

a potential focus of adenomatosis, then biopsies (<1 cm diameter) were obtained from the pancreatic head, body, and tail. After hematoxylin and eosin (H&E), the frozen sections were evaluated for presence of islet cell nuclear enlargement (defined as a nucleus that has an area three times larger than the surrounding nuclei) and focal adenomatosis (defined as a confluent proliferation of endocrine cells that occupies at least 40% of lobules). When islets with unequivocally large nuclei were identified in multiple biopsies, diffuse PHHI was diagnosed, and a near total pancreatectomy was performed. When normal pancreas was seen, the search for a focal lesion continued. Using this method, 17 of 18 diffuse hyperinsulinism cases were correctly diagnosed by frozen section, and 26 of 30 focal hyperinsulinism cases were correctly diagnosed by frozen section.

When a focal pancreatic lesion is found and fully resected, this procedure is curative. However, some focal lesions may be small and difficult to identify. In such cases, some authors have recommended performing an approximately 75% pancreatectomy, namely resection of the body and tail of the pancreas to the right of the superior mesenteric artery (1). The specimen is submitted for meticulous histopathological examination in search of a focal lesion, which may be of microscopic dimensions. Unfortunately, focal lesions have been found in all locations in the pancreas. Adzick et al. (2004) reported their experience with partial pancreatectomies for focal PHHI (8). Of the 36 patients reported, 19 patients had focal lesions in the pancreatic head, 15 were in the neck, body, or tail of the pancreas, and four had extensive involvement of the pancreas. Although most patients underwent distal pancreatectomies, five patients required complete resections of the head of the pancreas with preservation of the body and tail and Roux-En-Y pancreaticojejunostomy.

If a patient fails medical treatment and is diagnosed with diffuse PHHI, a near-total pancreatectomy is performed. A 95% pancreatectomy, the procedure of choice, involves resection of the neck, body, and tail of the pancreas as well as the uncinate process posterior to the superior mesenteric vein (SMV), and excision of most of the pancreatic head, leaving only a sliver of pancreas on the medial wall of the second part of the duodenum and around the common bile duct. This procedure requires a high level of technical sophistication and carries a high risk of future diabetes mellitus. Of all patients undergoing 95% pancreatectomy, 50–75% will develop insulin-requiring diabetes mellitus (3,9). At the same time, approximately one-third of patients who undergo this procedure will suffer from persistent hypoglycemia (3). Therefore, careful postoperative care and close long-term follow-up of these patients are imperative.

Two recent publications have reported single institution, long-term follow-up of patients who have undergone subtotal or near-total pancreatectomies for persistent hyperinsulinemic hypoglycemia of infancy. Cherian and Abduljabbar (2005) published their data from Saudi Arabia, where the incidence of PHHI is higher secondary to a higher consanguinity rate (10). They followed 10 infants (four boys and six girls) diagnosed over a 20-year period who underwent 95% pancreatectomy for PHHI. All patients failed initial medical therapy. After surgical resection, two patients suffered transient episodes (lasting a few weeks to 6 months) of hypoglycemia that responded to diazoxide treatment, and all patients had pathologic diagnosis consistent with diffuse hyperinsulinemia. Unfortunately, all of their patients eventually developed diabetes mellitus. Three patients developed persistent hyperglycemia immediately after surgery, and seven patients developed diabetes between 7–11 years after surgery.

Ismail and Werther (2005) published their institutional outcome in Australia over a course of 16 years (11). They followed 14 patients with PHHI. A group of 10 patients responded to medical treatment with diazoxide. Of these patients, eight were still on medication at the time of the last follow-up (range of 6 months to 16 years). The most common side effect of diazoxide treatment was hirsutism. Four patients failed medical therapy and underwent surgery. One patient underwent an 80% pancreatectomy for focal adenomatosis. Three patients underwent 95% pancreatectomies for diffuse hyperinsulinemia. One of these patients developed hyperglycemia immediately after surgery, which persisted. None of the other three surgical patients had developed diabetes mellitus or recurrent hypoglycemia at a follow-up of 5–8 years.

Outcomes and rates of morbidity in patients who undergo subtotal or near-total pancreatectomies for PHHI can differ significantly from institution to institution. Although reasons for these differences in outcomes are unknown, one possible reason may be that differing degrees of pancreatic resections are performed from patient to patient. This may be owing to the fact that different surgeons define subtotal and near-total pancreatectomies differently. It may also be owing to the fact that pancreatic tissue distribution may differ from patient to patient, even when anatomic relationships are used in defining resection margins. Reyes et al. (1993) illustrated this problem in a study of 13 pediatric patients who underwent pancreatic autopsies within 24 hours of death (12). He showed that pancreatic tissue from the tail to the right of the superior mesenteric vessels (what is traditionally considered a 75% pancreatectomy) can weigh between 39–72% of the total pancreatic weight (average = 54%). Pancreatic tissue to the left of the pancreaticoduodenal vessels can weigh between 44–96% of the total pancreatic weight (average = 69%). Just the pancreatic head tissue in the duodenal sweep and the uncinata process can weigh between 4–57% of the total pancreatic weight (average = 32%). Therefore, in order to perform an adequate subtotal (60–90%) or near-total (95%) pancreatectomy, the entire gland including the uncinata process must be visualized at the time of the resection.

Recurrent hyperinsulinism after surgical resection is attributed to residual, nonexcised pathologic pancreatic tissue. Some patients who develop recurrent hyperinsulinism after surgery can be adequately treated with long-term medical therapy with diazoxide. However, some patients may require additional surgery in order to control recurrent hypoglycemia (8,13). In these patients, reexploration with resection of remaining pancreatic tissue is necessary. Martinez-Ibanez et al. (2002) published a series of 12 patients who suffered recurrent PHHI after subtotal pancreatectomy for diffuse PHHI (13). Two patients required total pancreatectomies in order to control their disease. Both patients subsequently developed diabetes; one died and one was alive at follow-up. Often, recurrence results from initial difficulty in fully excising diseased tissue from the pancreas around the common bile duct or the pancreatic duct (8). Therefore, this area must be carefully inspected during second surgery. There is minimal information in the literature addressing usefulness of preoperative imaging for patients who require reexploration. It is possible that with increased use of PET scans in the future, this imaging modality will be helpful in locating residual pathologic tissue prior to surgical reexploration.

Although surgical management of PHHI can often be successful, the morbidity associated with this procedure is significant. Therefore, medical treatment is still considered the first line of treatment for this disease with surgical resection serving as an adjunct to failed medical therapy.

TRAUMATIC PANCREATIC PSEUDOCYST

Pancreatic pseudocysts in children are the results of blunt abdominal trauma in greater than 60% of cases (9). The pancreas is the fourth most commonly injured intraabdominal solid organ in children with blunt trauma (9). The mechanism of injury usually involves anterior compressive forces applied to the pancreas, such as from a bicycle handle bar, off-road vehicles, or contact sport. As stated before, child abuse must be suspected in young children with upper abdominal trauma. Pseudocysts form as a result of egress of pancreatic enzymes after injury, which cause autodigestion of surrounding tissues, ultimately resulting in the formation of a cavity of enzymatic fluid lined by fibroblasts and inflammatory cells.

Traumatic pseudocysts occur most commonly after a grade III injury to the pancreas (distal transection or parenchymal injury involving the pancreatic duct) (14). Controversy exists on management of this type of injury in children. Some surgeons advocate a nonoperative approach initially, arguing that not all pancreatic ductal injuries develop into pseudocysts and those that do may be amenable to percutaneous or endoscopic drainage (15,16). Wales et al. (2001) published a retrospective study of nine patients with computed tomography (CT)-documented traumatic pancreatic transections who were treated without surgery (16). Only four of the nine patients developed pseudocysts. Although CT scans may not produce a 100% accurate diagnosis of pancreatic transection, this data does raise a question as to whether all pancreatic ductal transections require surgical intervention. Some surgeons favor early distal pancreatectomy, arguing that early surgery allows for timely definitive treatment of ductal injury, shortening hospital stay and length of abdominal symptoms. Meier et al. (2001) reported a series of nine patients who underwent initial operation for pancreatic transection (17). The mean hospital stay for these patients was 11 days with a mean total perenteral nutrition (TPN) use of 8.3 days. They compared this to two patients who were initially observed. Both patients eventually developed pseudocysts and required drainage procedures. They also required 20 and 45 days of TPN, respectively. Nadler et al. (1999) reviewed 51 patients treated for blunt pancreatic injuries at their institution (18). They showed that patients who underwent laparotomy within 48 hours of injury for pancreatic transection had a significantly shorter length of hospital stay than those who underwent laparotomy more than 48 hours after injury. At our institution, we have treated five patients with traumatic pancreatic transections over the last 5 years. Four patients underwent immediate spleen-sparing distal pancreatectomies, with a mean hospital stay of 8 days. One patient was initially managed nonoperatively. He developed a pseudocyst, which was treated with a cystojejunostomy. He ultimately required a second surgery, a distal pancreatectomy, for treatment of recurrent pancreatitis. The question as to whether nonoperative management should play a role in the initial management of grade III pancreatic injuries remains to be answered. If one prefers early surgical intervention, it should be prompt (within 24–48 hours of injury). If the patient is stable and observation is selected, further evaluation with noninvasive or minimally invasive techniques, such as magnetic resonance cholangiopancreatography (MRCP) and endoscopic retrograde cholangiopancreatography (ERCP), should be considered as needed. At this point, initial management should be individualized depending on site of injury, timing of referral, associated injuries, and institutional expertise.

Once a pseudocyst forms, there are multiple management options. Some pseudocysts resolve spontaneously. In adult studies, patients who were unlikely to experience spontaneous resolution of pseudocysts included patients with chronic pancreatitis, patients with pancreatic ductal abnormalities, and patients with cysts lasting greater than 6 weeks (19). Size did not predict likelihood of subsequent cyst resolution (19). Persistent pancreatic pseudocysts in children are thought to be less common than in adults because of a lower incidence of chronic pathologies causing pancreatic ductal and parenchymal abnormalities (20).

Early treatment with somatostatin analogs may have some therapeutic effect on cyst resolution in children, as illustrated by a few case reports. Mulligan et al. (1995) reported successful resolution of traumatic pseudocysts in two children after 28 and 35 days of subcutaneous octreotide treatment (21). Bosman-Vermeeren et al. (1996) reported one case of a patient who developed a pancreatic pseudocyst after exploratory laparotomy for traumatic pancreatic rupture (22). The pseudocyst resolved with somatostatin treatment, thereby avoiding the need for a second surgical intervention. However, there are no prospective studies on this subject. Therefore, at this point, somatostatin analog is not part of standard of treatment for pancreatic pseudocysts in children.

Chronic pseudocysts are defined as those persisting 4–6 weeks. In individual series, the incidence of chronic pseudocyst formation after trauma in children ranges from 0–69% (23,24). Persistent pseudocysts, especially those that cause gastrointestinal symptoms, require drainage in order to prevent possible complications associated with bleeding, rupture, or infection (Fig. 1).

The standard treatment for drainage of chronic pancreatic pseudocysts is a cystogastrostomy or a Roux-en-Y cystojejunostomy. Hemorrhage is the most serious complication associated with this procedure and is seen more commonly with

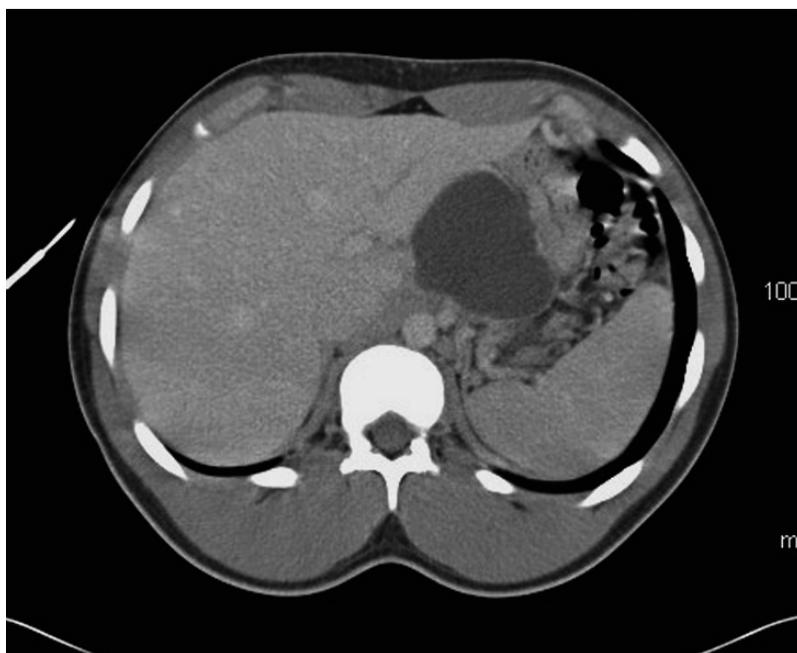


Fig. 1. CT scan of a patient with a pancreatic pseudocyst 8 weeks after blunt abdominal trauma.

cystogastrostomy than cystojejunostomy (20). The most common site of bleeding after a cystogastrostomy is at the anastomotic site along the submucosa of the gastric wall (20). Precise hemostasis with ligation of vessels in the gastric wall at the time of anastomosis can decrease the risk of this complication. If bleeding occurs, the most effective treatment is reexploration and ligation of the bleeding vessel. If the bleeding rate is relatively slow and the patient is hemodynamically stable, gastroscopy and/or angiography can be useful adjuncts for localization and possible therapeutic treatments of the bleeding site. No data is available on pseudocyst recurrence rates after surgical drainage procedures in the pediatric patients. In the adult literature, recurrence rates of 3–13% have been reported (25–27).

Pancreatic resections have also played a role in the treatment of pancreatic pseudocysts in the adult population, although little is known about its efficacy in children in comparison to drainage procedures. Schlosser et al. (2005) compared 169 adult patients who underwent a duodenum-preserving pancreatic head resection for pancreatic pseudocyst to 37 patients who underwent pseudocystojejunostomy for the same pathology (25). They found that pain relief was better after resection in comparison to the drainage procedure (94 versus 75% of patients), pseudocyst recurrence was lower after resection (13 versus 1%), but endocrine function was worse after resection (65 versus 33% diabetics). There have not been similar reports in the literature comparing resection versus drainage procedures in children.

Although internal surgical drainage procedures have been advocated as a safe, time-tested, and effective treatment for chronic pseudocysts, there has recently been increasing utilization of and an accepted role for external cyst drainage. In patients who have evidence of cyst infection, external drainage has traditionally been advocated to prevent further spread of infection (9). External drainage is also advocated when the cyst requires drainage sooner than 4–6 weeks after cyst formation, because the lack of a mature pseudocyst wall makes formation of a reliable anastomosis difficult (20). Risks associated with external drainage of pseudocysts include infecting the residual pseudocyst, hemorrhage associated with the drain, and development of a persistent pancreatic fistula. Pseudocyst infection from a drain usually implies inadequate cyst drainage (20). Ultrasound, CT, or fistulograms may be necessary to identify the etiology of the inadequate drainage, and drain adjustment and replacement may be required in order to correct this problem.

Even though risk of fistula formation after external drainage of pseudocysts also exists in children, there is no published data on the incidence of this complication. Most available case reports and published individual center reviews have shown a relatively high success rate of external drainage of pseudocysts in children. Burnweit et al. (1990) published a review of their experience with pancreatic trauma (23). Pancreatic pseudocysts developed in 45% of their patients who suffered blunt pancreatic trauma. Of these 13 children, six patients experienced resolution of the pseudocysts with bowel rest and TPN. Two patients underwent standard operative procedures (one for cystogastrostomy and one for distal pancreatectomy) because the surgeon felt that spontaneous resolution was highly unlikely based on the size of the pseudocysts. Five patients underwent percutaneous drainage of the pancreatic pseudocysts either under CT or ultrasound guidance. All patients were able to eat after the procedure, and the drains were removed 6–15 days after the procedure once drain output was minimal and pseudocyst decompression was confirmed by ultrasonography. No recurrence was documented at follow-up, which ranged from 8 months to 5 years. Lucaya et al. (1998) reported use of

external drainage and somatostatin treatment for a traumatic pancreatic pseudocyst in a 13-year-old girl (24). ERCP showed a distal pancreatic ductal disruption. The drain was removed after 45 days, and the patient did not suffer a recurrence. In the study by Wales et al. referenced earlier, three of the four patients who developed pseudocysts required intervention (16). All three patients underwent percutaneous catheter drainage with a length of 14–60 days. No recurrences were reported. Conversely, Rescola et al. (1990) published a series of four cases of pancreatic pseudocysts in which percutaneous catheter drainage was used (28). In three of the four cases, percutaneous drainage failed to resolve the problem and distal pancreatectomies were required.

An alternative treatment that has gained wider use in the pediatric population recently with increasing expertise in endoscopic procedures is the use of endoscopic cyst decompression. Patty et al. (2001) reported three cases of pseudocysts in pediatric patients (two traumatic, one secondary to pancreatitis) that were drained by endoscopically placed cystogastrostomy stents (29). The stents were removed at 12 weeks, 7 weeks, and 4 months, respectively. There were no recurrences at the 2-year follow-up. Al-Shanafey et al. (2004) reported three cases of pancreatic pseudocysts (secondary to medical pancreatitis), all successfully treated with endoscopically placed transpapillary drainage or endoscopic cystoenterostomy (30).

There is no good data in either the pediatric or adult literature on incidence of fistula formation after external drainage procedures. However, a recent report published in *Annals of Surgery* by Nealon and Walser (2005) evaluated a group of adult patients who failed endoscopic or percutaneous drainage of pancreatic pseudocysts, defined as persistent fistula drainage (31). They evaluated 79 patients who were managed by their institution's surgical service after failed minimally invasive procedures. Of these 79 patients, 91% had at least one episode of sepsis with 46% requiring intensive care unit (ICU) stays, and 12% had episodes of hemorrhage. Of the 79 patients, 66 ultimately required surgical procedures (59 drainage procedures, seven distal pancreatectomies). Out of 66 patients, 27 required surgery urgently mostly because of sepsis. A majority of these failed patients (80%) had evidence of main pancreatic ductal disruption or stricture along with a pseudocyst on ERCP evaluation. Their conclusion was twofold: Patients who fail endoscopic or percutaneous drainage of pancreatic pseudocyst can suffer severe complications from this procedure; and secondly, failure of these procedures may be associated with severity of main pancreatic ductal disruption. Therefore, preprocedural evaluation of pancreatic ductal anatomy with ERCP in patients with persistent and symptomatic pseudocysts may help direct choice of surgical versus endoscopic intervention.

It is interesting that there are successful reports of external drainage of pancreatic pseudocysts in pediatric patients with documented pancreatic ductal disruption (16,24,32). The presumption is that the pancreatic duct recannulates during the healing process once the pseudocyst is drained. These minimally invasive procedures may be more successful in children than in adults, because most children with pancreatic injury do not suffer from generalized pancreatic parenchymal and ductal pathology, which may make chronic fistulization and recurrent inflammation an additional problem that must be addressed. Nevertheless, even though there are several reports of successful use of endoscopic methods of pseudocyst decompression in children, additional investigation is still needed in order to answer the question of what anatomic criteria will predict success of these procedures in children in comparison to open surgical drainage or resection.

ACUTE RECURRENT PANCREATITIS AND CHRONIC PANCREATITIS

Although acute pancreatitis results from a single insult to the pancreas causing inflammation and injury and is often associated with trauma when seen in children, acute recurrent pancreatitis and chronic pancreatitis represent a stepwise or gradual destruction of pancreatic parenchyma, involving pancreatic exocrine acinar cells and ductular structures, as well as endocrine islet cells. This destruction results in atrophy and fibrosis and ultimately leads to pancreatic insufficiency and diabetes mellitus. The etiology of acute recurrent pancreatitis may include the following: unrecognized pseudocyst, biliary diseases such as cholelithiasis, sphincter of Oddi dysfunction, choledochal cyst, abnormalities of pancreatic drainage such as pancreatic divisum or ductal strictures, hypertriglyceridemia, or genetic predisposition as in certain abnormalities of the trypsinogen gene and cystic fibrosis transmembrane regulator gene (33). If the inflammation of the pancreas persists without full remission then chronic pancreatitis is the appropriate term.

Chronic pancreatitis in children is most commonly associated with systemic disease processes involving genetic abnormalities. In pediatric patients, cystic fibrosis is the most common cause of chronic pancreatitis. Depending on the type of mutation seen in the cystic fibrosis transmembrane conductance regulator, patients present with either pancreatic insufficiency or chronic pancreatitis. Patients with one severe and one mild variable mutation have a 40-fold increase in the risk of developing chronic pancreatitis over the general population (34). Hereditary pancreatitis is another cause of pediatric chronic pancreatitis. Hereditary pancreatitis is defined as recurrent inflammation of the pancreas that occurs in families for two or more generations without other known predisposing factors (35). Trypsinogen mutations have been associated with the majority of hereditary pancreatitis kindreds. These patients usually present with recurrent bouts of acute pancreatitis in childhood with a median age of onset of approximately 10 years of age (34). Patients with this diagnosis have a 50- to 60-fold increase in their risk of developing pancreatic cancer (33).

Initial management of pancreatitis in all patients is principally medical and supportive in nature, including aggressive hydration, management of metabolic complications and pain, and minimization of pancreatic stimulation by fasting with institution of nutritional support by parenteral or jejunal enteral feedings. Somatostatin analogs have been used by some centers as a part of initial therapy, but its efficacy is anecdotal and unproven (33,36,37). Paran et al. (1995) reported a prospective study of patients treated with octreotide for severe pancreatitis (37). There was a trend towards decreased rates of complications in patients treated with octreotide, although not all parameters reached statistical significance. Antibiotic therapy has also been shown in some studies to decrease rates of infectious complications and to confer survival benefit when used in patients with severe pancreatitis, although conflicting data still exists on this subject (38). Early surgical intervention should be avoided if possible, as early surgery has been associated with increased mortality (39). Early surgical debridement of the pancreas should be reserved only for patient with evidence of infected pancreatic necrosis confirmed by percutaneous aspiration of peripancreatic fluid (40). If pancreatic insufficiency develops over time as patients recover, pancreatic enzymes are added to the medical regimen.

Patients with pancreatitis secondary to pancreatic ductal or sphincter abnormalities may benefit from ERCP, pancreatic ductal stone removal, and/or sphincterotomy. There

are no randomized trials of this therapy in children; however, there are several case reports of its efficacy. Kozarek et al. (1993) reported five patients with chronic recurrent pancreatitis who underwent ERCP and papillotomy, with concurrent stone extraction in three patients (41). Four of the five patients experienced marked improvement in their chronic abdominal pain. Two patients were symptom-free at 9 and 11 months, one patient had recurrent pain at 6 months that responded to repeat ERCP and stone removal, and one patient underwent elective surgery for cyst drainage. Varadarajulu and Wilcox (2006) reported a prospective evaluation of six patients with recurrent pancreatitis or persistent right upper quadrant pain diagnosed with sphincter of Oddi dysfunction by manometry (42). These patients underwent either pancreatic sphincterotomy or pancreatic and biliary sphincterotomy based on location of their pathology. Four of the six patients were asymptomatic at a follow-up of 329–1445 days. One patient suffered recurrent symptoms at 525 days, and one patient had only partial symptom relief at 305 days.

Open surgical procedures for pancreatic ductal drainage and pancreatic resection are generally reserved for patients with long-term complications of chronic pancreatitis, such as pseudocyst formation, ductal strictures, intractable pain, and pancreatic failure (Fig. 2). The choice of surgical procedure and the timing of the procedure are dependent on multiple factors including the quality of the pancreatic parenchyma and pancreatic duct, the location of fibrosis and strictures, the presence of exocrine or endocrine insufficiency, and the severity of patient's abdominal pain. Surgical options include minimal procedures such as open sphincteroplasty to improve proximal ductal drainage, to lateral pancreaticoenterostomy with or without pancreatic resection to drain more extensive disease, to distal pancreatic resection to control pain from small ductal disease. Although no extensive studies have been performed to evaluate the efficacy of these procedures in children, there are several informative retrospective reports documenting single institution experiences with these procedures.

Neblett and O'Neill (2000) reported their experience over a 20-year period with 10 pediatric patients with recurrent pancreatitis secondary to pancreatic divisum (43). The average age of first symptoms in these patients was 6 years old. All patients were diagnosed by ERCP at an average age of 11.8 years old. All patients had evidence of obstruction at the level of the minor ampulla or at the junction of the two ducts. Eight patients had complete pancreatic divisum, and two had incomplete variants. Nine of the 10 patients underwent initial transduodenal sphincteroplasty of the accessory duct. One patient underwent primary lateral pancreaticojejunostomy (LPJ) because of a stricture at the junction of the main and accessory pancreatic ducts. Of the nine patients who underwent sphincteroplasty, two patients reported excellent results (no ongoing episodes of pain or pancreatitis), one reported improved results (occasional attacks of pain requiring hospitalization every 2–3 years), one patient was lost to follow-up, and five patients failed the procedure. Of these five patients, one refused further surgery and four underwent lateral pancreaticojejunostomy with improved results. The mean duration of follow-up was 7.3 years. None of these patients ever developed exocrine or endocrine insufficiency and none require chronic analgesics. This study showed that although some patients may benefit from sphincteroplasty for treatment of proximal ductal obstruction from pancreatic divisum, many patients will require more extensive drainage procedures over time.

The pathogenesis of pain in chronic pancreatitis is incompletely understood and is probably multifactorial in nature. One prominent theory is that pancreatic pain is caused

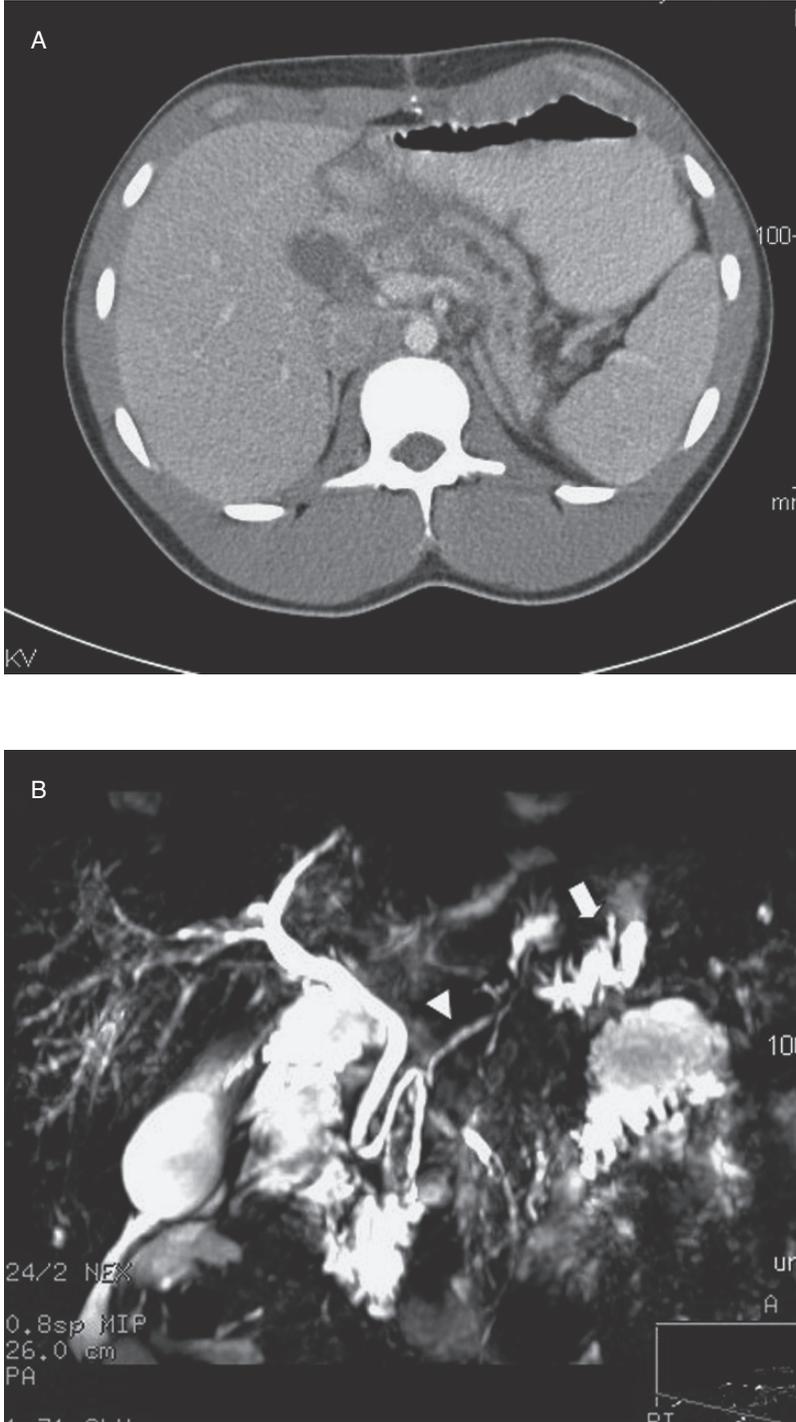


Fig. 2. (A) CT of a patient with recurrent pancreatitis after blunt abdominal trauma. (B) MRCP shows a normal proximal pancreatic duct (triangle), a high-grade mid-ductal stricture, and a dilated, tortuous distal duct (arrow).

by increased intraductal and interstitial pressures caused by fibrosis and obstruction. This is reflected in the fact that in patients with main pancreatic ductal dilation and/or pancreatic ductal stricture(s), ductal drainage procedures have been shown to provide some relief of chronic pancreatitis pain. An additional goal of surgery in children is to facilitate enzymatic drainage, thereby preserving pancreatic function and contributing to normal growth and development (44). Although many variations of pancreatic ductal drainage procedures are used, lateral pancreaticojejunostomy, or the modified Puestow procedure, is one of the most commonly used procedures and has been shown to result in immediate pain relief in 80% of adult patients (45). There are a few retrospective outcome reports of this procedure in pediatric patients.

DuBay et al. (2000) reported their experience over a 25-year period treating pediatric patients with hereditary pancreatitis with the modified Puestow procedure (44). There were 12 patients in their retrospective study. The mean age at first hospitalization was 5.5 years old. The mean duration of symptoms before surgery was 3.9 years. The presenting symptom was abdominal pain in 10 patients and failure to thrive in two. Nine patients also had a greater than 10% weight loss, and eight had a history of pseudocyst formation. All patients had dilated main pancreatic ducts diagnosed by CT scan ($n = 7$), ERCP ($n = 4$), or at surgery ($n = 1$). The pancreatic ducts were identified at surgery by various techniques: a 1- to 2-cm distal pancreatectomy ($n = 6$), via a communicating pseudocyst ($n = 3$), a blind incision ($n = 2$), or intraoperative needle localization ($n = 1$). Distal pancreatectomy for ductal identification prolonged operative time and increased blood loss in comparison to the other techniques. There were no operative mortalities, but there were four postoperative complications: superior mesenteric artery syndrome, postoperative bleeding requiring reexploration, subphrenic abscess requiring operative drainage, and Roux stasis until the tenth postoperative day. Postoperative follow-up showed that hospitalizations for pancreatitis were significantly reduced in all patients over both a 1- and 3-year period. All patients experienced an increase in their percentile body weight postoperatively. No patients required insulin pre- or postoperatively. Five patients were noted to have steatorrhea preoperatively, which resolved in four patients and was controlled in the fifth patient with enzyme supplements. The median follow-up was 15 years. At follow-up, seven patients ranked their surgical outcome as excellent, five as good, and one as poor. Eight of the 12 patients had had symptoms of pancreatitis, six patients had needed hospitalizations, four patients had been placed on pancreatic supplements, and none of the patients had had diabetes mellitus. Their conclusion was that in children with complicated hereditary pancreatitis, the modified Puestow procedure improved the quality of life by decreasing hospitalizations, increasing ideal body weight, and improving pancreatic function.

Chiu et al. (2006) reported their experience over a 6-year period with four pediatric patients who underwent LPJ for chronic pancreatitis (46). There were no mortalities but a significant number of complications including: bowel obstruction caused by internal herniation 1 month after surgery, incisional hernia requiring operative repair, and severe respiratory complications requiring a prolonged stay in the ICU. The follow-up ranged from 2–6 years. There were no recurrences of pancreatitis within this period and all of the patients were able to eat a normal diet. No patients required pancreatic enzyme supplementation, and only the patient who required ICU stay needed insulin postoperatively.

Although the modified Puestow procedure has been shown to significantly decrease symptoms associated with chronic pancreatitis in many children, there have been

noted surgical failures and recurrences. Surgical failures in these patients have been attributed to inadequate decompression of the head of the pancreas. Therefore, Rollins and Meyers (2004) reported their experience with a change in their institution from performing the LPJ to the Frey procedure (addition of anterior resection of the head of the pancreas to the LPJ) (47). They reported a 7-year retrospective study that included 11 pediatric patients (age range from 8–18 years old) followed over a period ranging from 1.3–8.8 years. All patients underwent preoperative evaluation with ERCP. A group of 10 patients had disease involving the head of the gland, and five patients had disease involving the entire gland. Three patients underwent LPJ and eight patients underwent the Frey procedure. One of the LPJ patients underwent conversion to a Frey procedure after a pseudocyst developed in the head of the pancreas 6 months after the initial procedure. There were no mortalities and four postoperative complications: central line infection, upper gastrointestinal bleeding secondary to gastritis, pleural effusion requiring thoracentesis, and renal insufficiency. Of the three LPJ patients, one patient's outcome was considered good (occasional pain not requiring hospitalization) and two patients' outcomes were considered poor (pain causing limitations in activity, regular narcotic use, or repeated hospitalizations). Of the eight Frey procedure patients, six patients' outcomes were considered excellent (patient reported no pain and had returned to normal activity), one patient's outcome was considered good, and one patient's outcome was considered improved (pain caused occasional absence from school or work or required occasional use of pain medication). The authors concluded that the Frey procedure addressed some of the limitations of the LPJ procedure and is currently their preferred procedure for surgical management of chronic pancreatitis in children.

In patients with chronic pancreatitis and no evidence of large duct disease, pancreatic pain is thought to be neuropathic in origin, secondary to chronic inflammation of the pancreatic perineural sheath caused by chronic or recurrent enzyme and cytokine release. Although this is a rare pathology to find in children, data on adult patients suggest that distal pancreatectomy, resection of the diseased pancreatic parenchyma harboring the involved inflamed nerves, can be an effective option. Sakorafas et al. (2001) reported a retrospective study of 38 adult patients diagnosed over a 20-year period with obstructive chronic pancreatitis who underwent distal pancreatectomies (48). The diagnoses were made by ERCP and CT scans and by pathologic confirmation at surgery. Patients were selected who had isolated, dominant major pancreatic ductal stricture or cutoff and changes of chronic pancreatitis in the gland distal to the stricture. In these patients, the head of the pancreas typically appeared relatively normal on imaging tests and lacked parenchymal calcification. The mean time from onset of symptoms to operation was 4 years. All patients underwent a distal pancreatectomy. In five patients, more than an 80% resection was required to reach the stricture. There were no postoperative deaths. Significant morbidity occurred in five patients: hemorrhage in three patients (all requiring reoperation) and two bowel obstructions (one patient requiring reoperation). Follow-up was complete in 31 patients. Mean follow-up was 6.7 years. A group of 15 (49%) patients had complete pain relief, 10 (32%) patients had partial pain relief, and six (19%) patients had inadequate pain relief. Preoperatively, only nine patients were able to work or function normally; postoperatively, 23 patients were able to do so. Diabetes mellitus developed in 14 (45%) patients, and exocrine insufficiency developed in 15 (47%) patients. The

authors' conclusion was that distal pancreatectomy achieved pain relief in a significant percentage of patients with low morbidity and no mortality.

Management of complications of chronic pancreatitis can be challenging. Many surgical options are available with varying degrees of success in relieving symptoms. Choice of surgical procedures depends on location of pancreatic injury and involvement of the main pancreatic duct. Reoperation for persistent or recurrent symptoms usually involves further resection of diseased pancreas.

PANCREATIC FISTULA

A pancreatic fistula is an abnormal passage of pancreatic juice through a pancreatic ductal disruption that exits the pancreatic parenchyma. This fistula can reside totally within the capsule of the pancreas and can be minimal and self-healing, or it can breach the capsule of the pancreas resulting in pancreatic juice entering the retroperitoneal or peritoneal cavities (49). The most important aspects of early management of pancreatic fistulas are similar to management of acute pancreatitis: intensive resuscitation with intravenous fluids and maintenance of adequate nutrition by parenteral hyperalimentation. Broad-spectrum antibiotics are usually added to this regimen (49). Most low-output fistulas will close spontaneously. Somatostatin analogs can decrease amount of fistula output and accelerate fistula closure; however, they will not induce closure of fistulas that would not otherwise close (9). Endoscopic placement of transpapillary endoprotheses can decompress the pancreatic ductal system, relieve downstream obstruction, and aid in fistula closure.

The most common cause of pancreatic fistula in children is as a postoperative complication after pancreatic surgery (9). In adult studies, most pancreatic fistulas resulting from pancreatic resections will close with medical management (50,51). Proactive closure of the pancreatic duct at initial surgery has been shown to decrease the rate of postoperative fistula formation (52). If pancreatic fistulas persist over time, a Roux-en-Y drainage procedure may be necessary once the fistulous tract has matured.

CONCLUSION

Management of pancreatic diseases and injury in children continues to be a challenging proposition. Complications associated with pancreatic diseases can cause significant morbidity. Although experiences with adult patients have provided useful guidelines in the care of pediatric patients, care of the pediatric patient with pancreatic surgical complications must be individualized to the patient and the presenting clinical situation.

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Rhabdomyosarcoma

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BACKGROUND

Rhabdomyosarcoma (RMS) is one of the more common solid tumors in children, with approximately 250 new cases diagnosed each year (1). The treatment of RMS has improved significantly during the last 30 years, with the overall long-term survival rates improving from 25% in 1970 to more than 70% with current multimodality treatment (2–4). The roles of pediatric surgeons in the treatment of RMS have changed significantly through the years, as other adjuvant therapies have become more efficacious.

RMS arises from primitive mesenchymal cells that are present throughout the body, even in areas that are usually not associated with striated muscle (5). However, all tumors show some degree of striated muscle differentiation. RMS may invade local structures and frequently metastasize early through lymphatics or hematogenous spread. Tumors are usually firm, nodular, and variable in size and consistency; however, they are not encapsulated and frequently invade surrounding soft-tissue structures. There are essentially four types of RMS: embryonal, alveolar, pleomorphic, and undifferentiated (3). Embryonal is the most common and is usually found in children less than 8 years old. Overall, patients with embryonal RMS have good prognoses, with a 5-year survival rate of 60%. For spindle cell and botryoid subvariants, the survival rate increases to 95%. Alveolar RMS is found in older children and is associated with

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tumors in the extremities and trunk. Patients with alveolar RMS have worse prognoses than those with embryonal, with an average 5-year survival rate of 54%.

SURGICAL GUIDELINES

The basic principle of wide and complete resection of the primary tumor with a surrounding “envelope” of normal tissue should be followed at the initial, or subsequent, operations whenever possible (6). A somewhat arbitrary margin of 0.5 cm circumferentially, or an uninvolved fascia margin, is adequate. This size of margin generally is more easily obtained in the extremities or trunk than in head and neck tumors. Adequate margins of uninvolved tissue are required unless excision involves sacrifice of normal tissue that cannot be resected, would result in an unacceptable loss of function/cosmesis, or is not technically feasible. Therefore, the resection of vital structures is probably not warranted for most tumors. Also, when the RMS arises from a somatic muscle, excision of the entire muscle of origin or the entire compartment usually is not necessary (7). The surgeon should mark all margins and orient the specimen at the operative field, so that margin evaluation is precise. Narrow margins are unavoidable in some sites. In those situations, the surgeon should take several separate biopsies of the “normal” tissue around the margins of resection and these should be marked and submitted separately for pathologic review.

Any suspected microscopic or gross residual tumor should be marked in the tumor bed with small titanium clips to aid radiotherapy simulation and subsequent reexcision. A clear margin and no residual disease (Group I) is superior to microscopic margins (Group II) or gross residual disease (Group III) in all outcomes analyses (3,8–10).

Exceptions to this general operative approach would include primaries in the orbit, head and neck, biliary, and geritourinary (GU) sites (11–17). For patients with primary disease at these sites, the primary treatment modality employed is chemotherapy and radiotherapy. This approach is appropriate given its usual embryonal histology, good response to treatment, and the poor likelihood of achieving complete operative resection without significant morbidity.

Clinical or radiographic evaluation of regional lymph nodes should be done during diagnostic work-up and is an important component of pretreatment staging. Clinically positive nodes should always be confirmed pathologically. For multiple clinically positive nodes, radical debulking may be useful, with radiotherapy, to obtain regional control (18,19). During biopsy of the regional lymph nodes, a “distant” node should be harvested for pathologic study. Involvement of these distant nodes is analogous to metastatic disease. There is no benefit from extensive nodal dissection if distant node or metastatic disease has already occurred.

Pathologic evaluation of clinically uninvolved nodes is site-specific; it is required in extremity sites and for children older than 10 years with paratesticular tumors (20). Aggressive regional lymph node sampling is the most appropriate method of surgical evaluation because resection is diagnostic but not therapeutic. For this reason, prophylactic radical node dissection, as used for some other malignancies, is not necessary in childhood RMS except in children older than 10 years of age with paratesticular RMS, who should undergo a retroperitoneal nerve-sparing lymph node resection.

Sentinel node mapping using blue dye, radioactive-labeled colloid, or both is helpful in determining regional node status (21). Preliminary data from the Intergroup Rhabdomyosarcoma Group (IRSG) suggest that sentinel node biopsy may be effective.

DIAGNOSTIC EVALUATION

Usually in this patient population, before enrollment, patients will have had an incisional biopsy to provide diagnosis or document metastatic disease histologically. As part of the staging process, patients undergo radiographic assessments with computed tomography (CT) of the chest, CT or ultrasound of the liver, magnetic resonance imaging (MRI) or CT of the head, bone scintigraphy, and bone marrow aspiration and biopsy. Additional staging studies such as abdominal CT scans and cerebral spinal fluid (CSF) cytology are individualized, depending on clinical findings and primary tumor location (Table 1).

PRETREATMENT REEXCISION (PRE)

PRE is a wide, nonmutilating procedure carried out in patients with soft-tissue sarcomas (STS) when microscopic residuals are left after initial excision or when there is insufficient data on the completeness of resection. PRE means wide reexcision of the previous operative site, including an adequate “envelope” of normal tissue, with careful marking and examination of all margins.

Frequently, the initial surgical procedure (a biopsy or excision as used for benign tumor) may have been performed prior to establishing the diagnosis and/or the involvement of the oncology team and may result in a situation in which there is: (1) gross residual tumor; (2) microscopically involved margins; or (3) uncertainty as to margins or residual disease. This applies even if the microscopic margin is thought to be clear, or if the operation and pathologic study were not done as a “cancer” operation.

Table 1
Components of Diagnostic Evaluation for Newly Diagnosed RMS

Hx/PE (Ht/Wt), Clinical assessment
CBC/Diff/Plt
PT, PTT, fibrinogen
Urinalysis, creatinine, SGPT, alkaline phosphate, Ca/Phos/Alb
Electrolytes (Na, K, Cl, CO ₂)
Creatinine clearance or GFR
Bilateral BM aspirate/biopsy
CT chest
EKG or MUGA for cardiac monitoring
MRI or CT of primary tumor ³
CT or US liver (for abdominal/pelvic tumors)
MRI or CT head
CT, US, MRI retroperitoneum for lower extremity, GU, pelvic and abdominal tumors only
Lumbar puncture (for metastatic parameningeal tumors and if multiple intracranial metastases at initial diagnosis)
Bone scan
Lymph node biopsy for extremity and paratesticular tumors

CBC, complete blood count; PT, parathyroid; PTT, partial thromboplastin time; SGPT, serum glutamate pyruvate transaminase; BM, bone marrow; CT, computed tomography; EKG, electrocardiograph; MUGA, multigated angiogram; MRI, magnetic resonance imaging; US, ultrasound; GU, geritourinary.

Under these circumstances, the concept of PRE is advisable and should be applied wherever feasible, unless the resulting disability would be considered unacceptable (i.e., resulting in loss of function or an unacceptable cosmetic result). A lymph node evaluation is indicated using the same criteria as outlined previously in the general surgical guidelines. PRE is particularly applicable to extremity and trunk lesions, although it should be applied wherever possible.

Prior to PRE imaging, a CT or MRI should be performed to visualize any residual tumor but, more importantly, to identify adjacent structures and determine if the residual area can be resected without causing significant morbidity. PRE, as its name suggests, should be done prior to the initiation of chemotherapy or radiotherapy. Clinical group assignment is determined on the basis of pathology from the definitive operation prior to the start of multimodal therapy. The prior procedure(s) will be considered a biopsy.

The initial report of PRE was published in 1989 by Hays et al. (8), looking at 41 children who had undergone PRE for extremity/trunk group II disease. These patients were converted to group I and subsequently had a 91% survival. This was significantly better than the survival for patients initially classified as group I patients (74%) and group II patients that had not undergone PRE (74%). Approximately 36% of pathologic specimens did not contain viable tumor. Additionally, 29% of PRE pathologic specimens had positive margins and required a third procedure. The local tumor recurrence rate (30%) was similar between all three arms of the study. This was not a prospective study and so does suffer from some selection bias. In addition, it did not look at group III patients. And last, pathologic error in initial margin classification is proposed for why the group II PRE patients did better than the group I patients when they should have been equivalent.

A recent study by Cecchetto et al. (22) evaluated the role of PRE in patients with STS enrolled in two consecutive Italian studies between January 1988 and September 1999. Of the 126 patients with grade IIa tumors (microscopic margins positive but no nodal disease), 53 patients (42%) underwent PRE. In this cohort, there were 23 patients with RMS and 30 patients with non-RMS STS. The median interval between primary surgery and PRE was 36 days. Of the 53 patients undergoing PRE, only eight (15%) continued to have positive margins postoperatively. Viable tumor was identified in 21 pathologic specimens from the 45 patients in whom the tumor was completely excised by PRE. Currently, 39/45 patients are in their first complete remission with a median follow-up of 53 months. Interestingly, the presence of residual viable disease in the PRE specimen did not correlate with outcome. PRE was effective especially in extremity, trunk, and paratesticular sites; however, its role was uncertain in large sarcomas over 5 cm in size. The authors reported that patients with complete resection had an 89% 5-year over-all survival. Unfortunately, they did not compare this result with a similar cohort that did not receive PRE to quantify the benefit achieved with PRE. It is also unfortunate that they did not look at group III patients (macroscopic residual), who make up the largest percentage of patients that may have benefited from PRE.

REOPERATION OF PRIMARY TUMOR

These resections occur after initiation of treatment with chemotherapy and may occur at any point in relation to radiotherapy. They are usually performed for group III patients who have macroscopic residual disease but no evidence of metastatic

extension. The most common scenario is for patients that have initially had a biopsy (group III) for large unresectable disease who have undergone adjuvant therapy and now the mass is decreased in size allowing it to be resected safely. Reoperation of the primary tumor in this setting can be broken down into three categories: delayed primary excision, second look operation (SLO), and end of therapy resection.

Resection During Therapy

Resection during therapy consists of two scenarios; delayed primary excision is the attempted complete resection occurring after induction chemotherapy (usually after two to four cycles of chemotherapy) and SLO is performed at any point during therapy and may consist of attempted resection or biopsy. Delayed primary excisions are performed if chemotherapy has decreased the tumor size sufficiently to perform a safe resection without significant morbidity. The purpose of delayed excision is to improve local control with the objective to completely excise the tumor. SLOs, after chemotherapy and radiotherapy, may be considered for patients who are deemed to have partial responses, and selected nonresponders. The purpose of SLO, in patients with controlled metastatic disease, is to confirm clinical response at the primary site, to evaluate pathologic response to therapy, and to remove residual local tumor whenever and wherever it is feasible in order to achieve local control (23,24). Similar to delayed primary excision, SLO occurs most commonly in group III patients. Optimally, complete resection is the objective of SLO if this is anatomically and functionally possible. SLO is not recommended immediately after irradiation, because within this timeframe local changes following radiotherapy could cause false positive interpretations of imaging. In addition, time must be allowed after the completion of radiotherapy for the tumor to achieve its final response status. Both delayed primary excision and SLO are performed using the standard surgical guidelines previously described, including wide reexcision of the previous operative site, an adequate “envelope” of normal tissue, and examination of all margins. A lymph node resection/sampling is indicated when appropriate. These operations are most efficacious for lesions of the trunk and extremities, tumors smaller than 5 cm, children over 10 years of age, lower stage disease, and children with negative nodes.

When these procedures should be performed in relation to other therapy is a question that should be determined in discussion with the child’s oncologist and radiotherapist. There are no strong data from which to draw any firm conclusions concerning timing of operative procedures in relation to chemotherapy and radiotherapy, yet consensus would suggest several guidelines that can be followed. As a general rule surgical resection may be performed once platelet counts have recovered from chemotherapy. The next cycle of chemotherapy may proceed 1–2 weeks postoperatively for trunk operations and 2–3 weeks for extremities, thus allowing another week after chemotherapy for white blood cell (WBC) counts to reach a nadir. Similarly X-ray therapy (XRT) should be delayed 2–4 weeks after resection. However, resection should be delayed 2–6 weeks after the completion of XRT so that response to XRT may be adequately quantified. Each of these guidelines should help to minimize the morbidity of the procedure that can be associated with the adjuvant therapy. For both delayed primary excision and SLO, either CT or MRI is most commonly used to evaluate patients for extent of residual local and metastatic disease prior to the procedure.

The importance of SLO to outcome is debatable. In support of SLO, results from Intergroup Rhabdomyosarcoma Study (IRS) III showed that 75% of patients classified as having partial responses by imaging were found to have complete responses during SLO or were converted to complete responses by excision of residual tumors (24). Converting them to complete response improved their survival. The SLO were most effective in extremities and trunks compared with head and neck lesions. Flaps and/or grafts were occasionally required for reconstruction, because prior radiation can affect wound closure and healing. For intracavitary sites, such as abdomen or thorax, it has been suggested that a complete second-look evaluation using open, laparoscopic, or thoracoscopic evaluation can be performed, but evaluation must be complete whatever method is used. If residual tumor persists after SLO, subsequent procedures should be performed after further therapy, to resect if feasible, or to confirm clinical response status.

However, in a more recent review of IRS-IV, SLO showed that only 20% of group III RMS patients with primary sites of extremity/trunk, retroperitoneal/pelvic, bladder/prostate, or head/neck/orbit had SLO during protocol therapy (Raney et al., manuscript in preparation). Those patients that had SLO were more likely to have trunk primaries, with tumors that were larger and a higher stage compared to those who did not undergo SLO. Also, patients who had SLO were less likely to receive appropriate XRT. Given these caveats, there were no differences in failure-free survival and overall survival between patients that had SLO and those that did not, even when adjusted for the known prognostic factors of age, histology, and stage. Some of this could be explained by the observation that only one-half of the pathologic specimens contained viable tumor, and that of those with viable tumor only one-half achieved complete resection with negative margins with SLO. Those patients that did not achieve complete resection were more likely to be infants, with nontrunk disease, at a higher stage with invasive tumor and positive nodes. If SLO was able to achieve complete resection with negative margins, then failure-free survival and overall survival were both significantly improved. Not too surprisingly, those patients with viable tumor had a significantly worse outcome.

Similarly, IRS-V encouraged SLO after 12 weeks of chemotherapy. IRS-V also permitted a radiation dose reduction from the standard 50.4 to 41.4 or 36 Gy, following SLO with gross total excision and either microscopic positive or negative margins, respectively. However, most eligible patients did not have SLO because of the anticipated morbidity of surgical resection at the most common anatomic sites of RMS, such as parameningeal or bladder/prostate. Outcome data from this study is still pending.

In conclusion, the results from these studies indicate that SLO is indicated, and can improve survival, for patients in whom a complete resection can be anticipated (usually patients with extremity or trunk lesions less than 5 cm in diameter in children over 10 years old with lower-stage node-negative disease) and who have viable tumor (usually patients with partial response or no response to chemotherapy). If complete resection is not achieved, then survival is not different compared to biopsy or observation only.

Resection at the End of Therapy

At the end of therapy, some patients with RMS have residual masses despite receiving all planned therapy. A recent review of IRS IV patients with group III (gross residual disease) RMS found the presence of a residual mass at the end of therapy did not correlate with outcome (25). This study looked at 419 group III patients who completed

all protocol therapy without developing progressive disease. At the end of therapy, 341 (81%) of children achieved complete resolution of the mass and 78 (19%) had a residual mass. The 5-year failure-free survival was similar for the two groups (80 and 77%, respectively) even after adjustment for age, nodal status, primary site, and histology. In addition, those patients that underwent operative resection at the end of therapy for a residual mass were likely to have significant morbidity associated with their procedure (26). A total of 63% of patients with a residual mass at the end of therapy underwent an attempted resection. Of these patients, 45% had excision of a vital structure during the procedure with a resultant loss of function in one-half. Unfortunately only 18% of patients had viable tumor detected in the pathologic specimen, and only two of these nine patients had negative margins. In addition, there was no improvement in outcome for patients that had the tumor resected versus biopsy alone. Therefore, given the lack of improvement in outcome and the associated morbidity, aggressive surgical or alternative therapy is probably not warranted for RMS with a residual mass at the end of planned therapy. In summary, the indications for operative intervention for residual masses at the end of therapy should probably be limited to excision of those tumors that would be easily resectable with minimal morbidity or else biopsy alone to document residual viable disease.

Resection for Recurrent Disease

Recurrent RMS can occur in as many as 35% of patients with trunk and extremity primary tumors, with 50–95% of these patients succumbing to progressive disease (3,27,28). In a study by Pappo et al. (29), the median time from tumor recurrence to death was only 0.8 years, with a 17% 5-year survival rate. In this study the best predictor for survival in patients with recurrence was histology with approximately a 30% survival for favorable histology (embryonal and botryoid) compared to 5% for unfavorable (alveolar and undifferentiated).

Recurrent RMS can be locally extensive and is usually treated with surgical resection and adjuvant therapy. The evidence for the efficacy of resection in these patients has been extrapolated from primary tumors, because there has not been good data looking specifically at recurrent disease. Recently, Hayes-Jordan et al. (30) looked at 32 patients with recurrent RMS. Of these, 13 patients had biopsy only with a subsequent survival rate of 8%. Another 19 patients had resection with a 37% survival rate. But, these operations were associated with a 15% major complication rate and an overall complication rate of 35%. The survival data does suffer from a couple of flaws. First, it was a retrospective review and therefore contains a large selection bias. Second, those patients that underwent operative resection had less extensive disease and fewer patients with metastatic disease, compared to those who only had biopsy, and this may explain the survival results rather than any true benefit from resection. Given these confounding factors, it is difficult to draw any firm conclusions. And given the poor prognosis for these patients, it may be reasonable to perform a resection using the same criteria previously established if the tumor can be safely and completely resected.

METASTATIC RMS

Patients with metastatic RMS account for 16% of all cases (3,31,32). Unfortunately, for patients who present with metastatic disease, there is only a 20–30% chance of survival (27,29).

Clinical Presentation and Risk Factors

Metastatic disease is the single most important predictor of clinical outcome in patients with RMS. Metastatic disease is most commonly asymptomatic and is frequently detected during staging or follow-up radiographic imaging. For those patients with symptoms, the symptoms are usually related to mass effect and functional compromise of adjacent structures.

The risk factors for metastatic disease and prognosis for these patients have been well described. In the study by Breneman et al. (33), IRS-IV patients with metastatic disease, compared to patients without, were more likely to be older, have higher incidence of alveolar histology, more commonly have invasive tumors, have larger tumors, have a higher incidence of regional lymph node involvement, and have a greater proportion of extremity and trunk/retroperitoneal primary sites. A total of 59% of patients had metastases confined to single anatomic site. The most common site of metastasis was the lung, followed by the bone marrow, bone, and distant lymph nodes.

IRS-IV patients with only pulmonary metastatic disease were evaluated and compared to patients with metastatic disease at other sites by Rodeberg et al. (34). Of patients with RMS and metastatic disease, 16% had isolated lung metastases. The majority of patients had bilateral pulmonary metastatic disease. Compared to patients with other sites of metastatic disease, patients with lung-only metastases had a greater proportion of embryonal histology, negative nodal involvement, parameningeal primaries, and a smaller proportion of extremity primaries.

Diagnostic Evaluation

The diagnostic evaluation for metastatic disease is similar to that for the primary lesion, including laboratory studies followed by evaluation for other metastatic disease and lumbar tap if there is a suspicion for central nervous system (CNS) involvement, bone marrow aspirate, bone scintigraphy, and CT evaluation of the liver, chest, and head. In addition, for those patients with metastatic disease biopsy, proof of tumor is also recommended unless the lesions are so obviously metastatic disease that pathologic confirmation is not needed.

Operative Therapy

Primary resection of metastasis is rarely indicated unless performed for pathologic confirmation of metastatic disease or with the intent to remove all metastatic disease in the setting of controlled primary disease. A delayed resection of persistent metastatic disease following radiation and chemotherapy should also only be performed if total removal could be performed without significant loss of function, disfigurement, and delay in initiating systemic treatment. There is really minimal evidence to support resection of metastatic disease (35). However if there is proven residual disease 3–6 months after radiation therapy, and if there was no residual unresectable disease elsewhere, then resection may be feasible.

Positive nodes outside of the regional basin (i.e., scalene node for upper extremity sites, inguinal nodes for paratesticular or vaginal/uterine sites, and paraaortic nodes for all sites other than immediately adjacent retroperitoneal or paratesticular sites), should be classified as distant metastatic spread (Clinical Group IV, Stage 4). Nodes that are positive in the regional nodal basin are not considered metastatic disease, but instead are treated as regional disease. This distinction underscores the importance of nodal sampling in the distal nodal basin if regional nodes are positive, to determine

if disease spread is regional or metastatic. This is especially important for RMS that commonly spreads to the lymph nodes such as extremity and paratesticular. Frequently, the nodes are classified positive based on imaging studies. If there is uncertainty as to involvement of these or other similar distant nodes, and the patient is classified as Stage 4 on the basis of these nodes, then biopsy by open or controlled needle techniques should be performed to confirm metastatic disease.

In some rare instances when there are two sites of disease, it can be difficult to be certain which of the two sites is primary. In this setting both should be resected with a curative intent. If there is question as to whether an apparent metastasis is RMS versus other benign disease, involvement should be confirmed by biopsy. Biopsy should be performed using either open, minimally invasive techniques or controlled needle biopsy as appropriate.

Outcomes

In IRS-IV the estimated 3-year overall and failure-free survival for all patients with metastatic disease was 39 and 25%, respectively (33). By univariate analysis, overall survival was significantly worse for alveolar histology and increasing number of metastatic sites. By multivariate analysis, the presence of two or fewer metastatic sites was the only significant predictor of outcome. The combination of embryonal histology with two or fewer metastatic sites identified a subgroup with a significantly improved survival. These results are similar to others (36,37). Similarly, the report by Carli et al. (38) identified primary tumor in parameningeal, extremity, or “other” sites; age younger than 1 year and older than 10 years; bone or bone marrow metastases; multiple metastases; and multiple sites of metastases as unfavorable prognostic factors for both overall and failure-free survival. Two subgroups were identified. Those with fewer than two unfavorable factors had 5-year overall survival of 47%, but for those with two or more unfavorable factors survival was only 9%.

Overall survival at 4 years for lung-only metastases was not significantly different from other single-site metastasis (42 versus 34%) (34). Survival was not improved for unilateral disease or fewer than five metastatic lesions. Factors associated with diminished overall survival included unfavorable histology and age greater than 10 years. Overall, outcome is comparable, although slightly better, for patients with isolated lung metastatic disease compared with other single-site metastatic disease.

ADJUVANT THERAPY

During IRS studies, different chemotherapeutic regimens have been evaluated as phase II window studies to determine new combinations that may improve outcome compared to standard vincristine, adriamycin, and cyclophosphamide (VAC) chemotherapy. The window trials have identified the combinations of ifosfamide and doxorubicin, with a 63% response rate, and topotecan and cyclophosphamide, with a 47% response rate (39,40). Using this information, the new proposed regimen for high-risk RMS patients will include ifosfamide-etoposide and vincristine-doxorubicin-cyclophosphamide.

Other strategies that have been attempted to improve outcome in these children include administration of high-dose chemotherapy (HDCT) followed by peripheral blood stem cell or autologous bone marrow support. In general these interventions have failed to significantly improve outcomes. The role of high-dose chemotherapy alone or combined with stem cell rescue (HSCR) in the treatment of relapse or metastatic

disease appears to be minimal, with survival rates, at best, of 12% at 2 years (41). These results are similar to the 10% 5-year survival rate reported by Pappo et al. (29) for patients who experienced progression of disease or relapse after treatment in IRS III, IV-pilot, and IV studies. Similar results were found in the European trial by Carli et al. (38), who reported that there was no survival difference between HDCT and standard chemotherapy.

Radiotherapy is specified for primary disease sites and any sites of metastases in an effort to improve disease control. Patients receive 50.4 Gy at 1.8 Gy/day to the primary tumor. Patients with metastases in soft tissue or bone also receive radiotherapy doses of 50.4 Gy. Patients who had diffuse pleural or peritoneal metastases received 14.4 Gy (whole lung) or 18.0 Gy (whole abdomen) to those cavities. Modification of radiation dose is made to keep adjacent critical structures within tolerance. In addition, there is evidence that radiotherapy may improve recurrence for both primary disease as well as metastatic disease (34,42).

SECOND MALIGNANCIES AND LATE COMPLICATIONS

Most late complications are related to chemotherapy and radiotherapy and may be site-specific or may be generalized. Most commonly the systems affected include visual, endocrine, cardiopulmonary, and neurologic. An excellent review of the generalized complications was performed by Punyko et al. using the Childhood Cancer Survivor Study Group (43). The relative risk for developing complications was greatest within the first 5 years after the completion of therapy. However, many serious complications occurred beyond 5 years including impairments of the visual, endocrine, cardiopulmonary, and neurologic systems. Examples of site-specific complications include patients with head and neck RMS enrolled on IRS II and III who were reviewed by Raney et al. (44). Over 77% of patients had at least one problem, and the most common included decreased growth and height, hypoplasia or asymmetry of tissues, impaired vision and hearing, as well as learning difficulties. Late effects are even more common for pelvis RMS (45). In this study, 92% of patients experienced some late complication and more than one-half required operative intervention to abate the complications. The main risk factor for complications appeared to be treatment with radiotherapy.

As expected, there is a risk of developing a second malignancy as a result of the treatment used to treat the initial malignancy. This was most recently reviewed by Cohen et al. (46) looking at 1499 RMS survivors. Patients were 7.7 times more likely to develop a second malignancy based on general population cancer rates. Second malignancy was much more common for patients treated with radiotherapy and chemotherapy compared to patients only treated by surgery. The types of cancer most likely to occur were acute myelogenous leukemia, cutaneous melanoma, breast cancer, and sarcomas of the bone and soft tissues.

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Neuroblastoma

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Neuroblastoma is an embryonal tumor of the sympathetic nervous system, arising during fetal or early postnatal life from sympathetic cells derived from the neural crest. It is the most common solid extracranial malignancy of childhood and the most common malignant tumor in infants (1). The overall incidence of neuroblastoma is one case per 100,000 children in the United States, or approximately 700 newly diagnosed patients per year. Neuroblastoma represents 7–10% of all malignancies diagnosed in pediatric patients younger than 15 years of age, but is responsible for a disproportionate percentage of pediatric cancer deaths, approximately 15% (2). However, neuroblastoma is an extremely heterogeneous disease (3); tumors can spontaneously regress or mature, even without therapy, or display a very aggressive, malignant phenotype that is poorly responsive to current intensive, multimodal therapy. A number of the factors responsible for this heterogeneity have been identified recently, and increasing evidence indicates that the biologic and molecular features of neuroblastoma are highly predictive of clinical behavior. The assessment of biological risk can, therefore, be used for treatment assignment and stratification, whereby those at high risk for disease relapse are given intensive multimodal therapy in an attempt to effect a cure, whereas those at low risk for relapse can have treatment intensity diminished in an attempt to avoid therapy-associated toxicity, while still achieving a very high rate of cure. The importance of appreciating the predictive value of these biologic factors is true not only for the oncologist when considering appropriate chemotherapy, but also for the surgeon when considering the nature and extent of an operative procedure for a child with neuroblastoma.

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PROGNOSTIC VARIABLES

The most important clinical variables for children with neuroblastoma appear to be age (4) and stage (5) at diagnosis. The most powerful biologic factors at this time appear to be *MYCN* status (6,7), ploidy (8,9) (for infants only), and histopathologic classification (10). These variables currently define the Children's Oncology Group (COG) risk stratification and therapeutic approach.

Stage

Criteria for staging neuroblastoma are included in the International Neuroblastoma Staging System (INSS) (Table 1) (11). The INSS is primarily a surgical staging system that includes factors such as the extent of surgical resection, the status of ipsilateral and contralateral lymph nodes, and invasion of the primary tumor across the midline for the determination of the stage of localized tumors. Because the INSS system depends to some extent on the skill and aggressiveness of the surgeon, an alternate staging system for patients with localized disease is being evaluated. This system, the International Neuroblastoma Risk Group (INRG) Staging System, is based solely on the radiographic appearance of the primary tumor and a determination of whether certain anatomic "risk" factors (primarily invasion of major vascular structures) are present (12). Lymph node involvement is not a factor in this potential new staging system.

Age

Patient age at the time of diagnosis is the only other clinical variable, in addition to stage, with independent prognostic value. For all stages of disease beyond stage

Table 1
International Neuroblastoma Staging System

1	Localized tumor with complete gross excision, with or without microscopic residual disease; representative ipsilateral lymph nodes negative for tumor microscopically (nodes attached to and removed with primary tumor may be positive).
2A	Localized tumor with incomplete gross excision; representative ipsilateral nonadherent lymph nodes negative for tumor microscopically.
2B	Localized tumor with or without complete gross excision, with ipsilateral nonadherent lymph nodes positive for tumor. Enlarged contralateral lymph nodes must be negative microscopically.
3	Unresectable unilateral tumor infiltrating across the midline ^a , with or without regional lymph node involvement; or localized unilateral tumor with contralateral regional lymph node involvement; or midline tumor with bilateral extension by infiltration (unresectable) or by lymph node involvement.
4	Any primary tumor with dissemination to distant lymph nodes, bone marrow, bone, liver, skin, and/or other organs (except as defined for stage 4S)
4S	Localized primary tumor (as defined for stage 1, 2A, or 2B), with dissemination limited to skin, liver, and/or bone marrow ^b (limited to infants <1 year of age).

^a The midline is defined as the vertebral column. Tumors originating on one side and crossing the midline must infiltrate to or beyond the opposite side of the vertebral column.

^b Marrow involvement in stage 4S should be minimal, that is, less than 10% of total nucleated cells identified as malignant on bone marrow biopsy or on marrow aspirate. More extensive marrow involvement would be considered to be stage 4. The MIBG scan (if performed) should be negative in the marrow.

1 localized tumors, patients less than 1 year of age have significantly better disease-free survival rates than older children with equivalent stages of disease (4). Outcomes for children ages 12–18 months with variable biologic features are currently being reviewed to determine whether there are subsets of patients in this age group who also tend to have a more favorable outcome, similar to that seen in infants less than 1 year of age.

Amplification of the MYCN protooncogene

Overall, approximately 25% of primary neuroblastomas in children have *MYCN* amplification; *MYCN* amplification is present in 40% of patients with advanced disease and 5–10% of patients with low-stage disease (6). The copy number, which can range from 5- to 500-fold amplification, is usually consistent among primary and metastatic sites, and at different times during tumor evolution and treatment (13). Amplification of *MYCN* is associated with advanced stages of disease, rapid tumor progression, and poor outcome; therefore, it is a powerful prognostic indicator of biologically aggressive tumor behavior (6,7).

Histopathology

Shimada and colleagues (14) developed an age-linked classification system of neuroblastic tumors based on tumor morphology. Neuroblastomas were divided into two prognostic subgroups, favorable histology and unfavorable histology, based on patient age and histologic characteristics, including the degree of neuroblast differentiation, the nuclear morphology of neuroblastic cells (mitosis-karyorrhexis index [MKI]), and the presence or absence of surrounding Schwannian stroma (15). This classification system has become widely accepted and has proven to be useful as an independent predictor of disease outcome (10). Therefore, histopathology is included in the current multifactorial risk stratification for patients with neuroblastoma. This determination is particularly important in patients who are older than 1 year and have localized disease. Because the histopathologic pattern within a tumor can be heterogeneous, Shimada has recommended analyzing representative sections from at least 1 cm³ of viable, nonnecrotic tissue to determine histopathologic classification. This recommendation has significant implications for the surgeon considering whether to perform an “open” or percutaneous core needle biopsy on a tumor suspected of being a localized neuroblastoma in a child older than 1 year.

Risk Stratification

The diagnosis of neuroblastoma is generally made by histopathologic evaluation of primary or metastatic tumor tissue, or by the demonstration of tumor cells in the bone marrow combined with elevated levels of urinary catecholamines. On the basis of the previously mentioned clinical and biological prognostic variables, infants and children with neuroblastoma are categorized into three risk groups predictive of relapse; low, intermediate, and high risk (Table 2). The probability of prolonged disease-free survival for patients in each group is 95–100%, 85–90%, and less than 30%, respectively.

LOW-RISK DISEASE

The treatment for patients with low-risk disease is generally surgical resection alone. This is true even in the presence of microscopic residual disease (stage 1), gross

Table 2
Neuroblastoma Protocol Assignment Schema

<i>INSS Stage</i>	<i>Age (days)</i>	<i>MYCN status</i>	<i>Shimada histology</i>	<i>Ploidy</i>	<i>Risk Group</i>
1	ANY	ANY	ANY	ANY	LOW
2A/B	<365	ANY	ANY	ANY	LOW
	≥365	Nonamplified	ANY	–	LOW
	≥365	Amplified	FAV	–	LOW
	≥365	Amplified	UNFAV	–	HIGH
3	<365	Nonamplified	ANY	ANY	INTERMEDIATE
	<365	Amplified	ANY	ANY	HIGH
	≥365	Nonamplified	FAV	–	INTERMEDIATE
	≥365	Nonamplified	UNFAV	–	HIGH
	≥365	Amplified	ANY	–	HIGH
4	<365	Nonamplified	ANY	ANY	INTERMEDIATE
	<365	Amplified	ANY	ANY	HIGH
	≥365	ANY	ANY	–	HIGH
4S	<365	Nonamplified	FAV	DI >1	LOW
	<365	Nonamplified	FAV	DI = 1	INTERMEDIATE
	<365	Nonamplified	UNFAV	ANY	INTERMEDIATE
	<365	Amplified	ANY	ANY	HIGH

residual disease (stage 2A), or ipsilateral, nonadherent lymph node involvement (stage 2B) if the tumor has favorable biologic characteristics. Infants with stage 4S disease who are not experiencing substantial symptoms may undergo an initial biopsy and observation only, if the tumor has favorable biologic factors.

INTERMEDIATE-RISK DISEASE

Intermediate-risk neuroblastoma is further classified into favorable and unfavorable biology on the basis of *MYCN* status, Shimada histology, and DNA ploidy. Treatment for patients with intermediate-risk neuroblastoma includes surgery and the use of four of the most active agents in neuroblastoma: cyclophosphamide, doxorubicin, carboplatin, and etoposide, given for either four cycles (favorable biology) or eight cycles (unfavorable biology).

HIGH-RISK DISEASE

The general approach to treating patients with high-risk disease has included intensive induction chemotherapy, surgery, myeloablative consolidation therapy with stem cell rescue, radiation therapy, and targeted therapy for minimal residual disease.

SURGICAL MANAGEMENT

Localized Tumors

Complete resection of a tumor is definitive therapy with generally excellent outcome for patients with most localized neuroblastomas (16). Biopsy of ipsilateral and contralateral lymph nodes is also currently required for accurate staging, risk assessment, and therapy assignment, although this may ultimately become an unnecessary part of staging neuroblastoma regardless of primary tumor site (*see prior*

discussion). It is important for surgeons to appreciate that regardless of the presence of microscopic or even gross residual disease and/or positive ipsilateral lymph nodes, no further therapy is given to patients with low-risk disease. This is true even if only 50% of the primary tumor is resected. Therefore, heroic efforts at gross total resection, including sacrifice of vital organs, is not warranted for patients with localized, low-risk disease (Fig. 1).

In most instances, performing a biopsy before resection is unnecessary in patients with localized tumors that are likely to be neuroblastoma, appear to be easily resected (based on diagnostic imaging), and have had a negative metastatic work-up. Resection, when performed as the initial intervention, especially for those whose primary site is thoracic, may obviate the need for chemotherapy, as many of these patients will have low-risk disease and an excellent prognosis, even if gross residual tumor remains. If there is any question of resectability, however, preresection biopsy should be performed to confirm the diagnosis of neuroblastoma and to assess tumor biology, as this may influence the surgical plan. For example, patients determined to have intermediate- or high-risk disease, based on unfavorable biologic features of their localized tumors, will receive chemotherapy regardless of the extent of surgical resection; the administration of this therapy in a neoadjuvant setting may make resection of the primary tumor easier.

Although *MYCN* assessment can be performed on a minimal amount of tumor tissue, the COG currently requires 1 cm³ of tissue to accurately assess the tumor's histopathology, which may be necessary for appropriate risk classification. Therefore,



Fig. 1. An abdominal computed tomography (CT) scan of a 15-month-old child with a localized left upper quadrant neuroblastoma. The degree of tumor involvement with the superior mesenteric artery and left renal vessels is uncertain by imaging studies, but this patient warrants an exploration to determine resectability of the tumor because, if the tumor biology is favorable, no other therapy in addition to surgical resection will be required, even if a gross total resection cannot be accomplished.

an “open” tumor biopsy (or potentially a laparoscopic biopsy) is recommended, as opposed to a percutaneous core biopsy, which generally provides smaller samples. Situations in which an open biopsy for accurate determination of tumor biology in patients with localized disease is crucial are: (1) If the patient is older than 1 year and has a nonmetastatic tumor that does not invade across the midline, then consideration might be given to resection of the primary tumor even if the tumor may involve adjacent organs or blood vessels. A gross total resection of all visible and palpable disease would be the goal of surgery. If, however, at exploration, the surgeon determined that this could not safely be achieved without risk to vascular structures or contiguous organs, but knew that the tumor either was not *MYCN* amplified or not of unfavorable Shimada histology, a safe resection of at least 50% of the tumor would be sufficient. This would still allow the patient to be classified as low-risk, stage 2A/2B and avoid adjuvant therapy, while still having an excellent prognosis. (2) All patients older than 1 year with localized, unresectable, nonmetastatic disease that crosses the midline (INSS stage 3) receive neoadjuvant chemotherapy. In these patients, Shimada histology may determine whether their disease is classified as intermediate- or high-risk for relapse, and, thus, the intensity of chemotherapy. Therefore, these patients require an open biopsy. Infants with unresectable stage 3 disease, however, need only a percutaneous biopsy for diagnosis and biology assessment, because only the *MYCN* status of the tumor determines the risk classification for these patients.

Locoregional Disease in Patients With Metastatic Disease

The role of surgery is less clear in the curative treatment of patients with high-risk neuroblastoma, given the very poor overall survival rate, but there does appear to be an association between gross total resection of the locoregional disease and local control. Unfortunately, more than one-half of patients with neuroblastoma present with advanced local or metastatic disease, which requires intensive multimodal therapy in addition to surgery. It is crucial that surgeons consider the heterogeneous nature of neuroblastoma, and the molecular and biologic characteristics associated with good or bad prognoses when determining the role of surgery for any specific case.

INITIAL PROCEDURES

More than 80% of patients who are older than 1 year of age and have metastatic neuroblastoma, will have tumor in the bone marrow (17). The presence of disseminated neuroblastoma is often suggested by a patient’s ill appearance, anemia, “raccoon eyes,” bone pain, or a limp, and can be confirmed by demonstrating neuroblasts in the bone marrow. In addition to demonstrating bone marrow involvement with tumor, the COG requires the documentation of elevated levels of catecholamines in the urine to confirm the diagnosis of neuroblastoma. Therefore, most of these patients do not require a surgical biopsy because they will all be classified as high-risk regardless of Shimada histology (or *MYCN* status). Under the proposed new risk stratification schema, however, Shimada histopathology may be important in determining the risk for patients aged 12–18 months with metastatic disease. Therefore, an open biopsy may be required for these situations. Finally, although some infants with metastatic disease are not at high risk for relapse, the only consideration in determining their risk classification is the *MYCN* status and ploidy, which can be performed on samples of the involved bone marrow or percutaneous core biopsies.

Although accurate diagnosis and risk assessment for children with neuroblastoma can often be accomplished without an open biopsy, generally little tumor tissue is obtained by a bone marrow biopsy or percutaneous core biopsy. These small samples, therefore, provide little additional tumor tissue for scientific studies. This is a very important consideration. The recent advances in understanding the biology of neuroblastoma have significantly impacted the current approach to patient treatment. Risk assessment, treatment assignment, and targeted therapy have all been made using patient-derived tumor specimens, banked only after tumor tissue necessary for diagnostic studies were taken and it is expected that analyzing additional genetic and biologic factors in the future will result in further refinement of the current COG risk group schema and will thereby impact future risk-based approaches to therapy. Because of this and the recognition of the critical importance for advancing the understanding of the biology of neuroblastoma and improving the outcome for patients with this disease, the current high-risk COG neuroblastoma clinical trial recommends that, if feasible and safe, according to the surgeon's judgment, and after appropriate informed consent, an open biopsy be performed in which at least 1 cm³ of tumor tissue be obtained. Because there is concordance between primary and metastatic sites with regard to *MYCN* amplification, the most accessible site should be selected for biopsy, should the surgeon elect to perform one (13).

DELAYED SURGICAL INTERVENTION OF LOCOREGIONAL DISEASE

The role of surgery in the management of children with high-risk neuroblastoma is controversial. Several reports have suggested that patients with INSS stage 3 or 4 disease who undergo gross total resection of their primary tumor and locoregional disease experience improved local tumor control and increased overall survival; however, other reports have not confirmed these observations. LaQuaglia et al. reported that the outcome for patients with stage 4 disease who were older than 1 year at the time of diagnosis was improved with gross total resection of the primary tumor, though the influence of resection was not independent of chemotherapy intensity. More recently, the same group has provided further data suggesting that local control and overall survival are correlated with gross total resection of the primary tumor in high-risk neuroblastoma and should be the goal in current clinical trials (18). Similarly, Haase et al. supported the use of aggressive surgery for patients with metastatic disease in an attempt to improve outcome (19). Grosfeld et al. found improved survival in patients with metastatic disease who underwent complete resection of the primary tumor during delayed second-look procedures (20), and Adkins, describing the CCG experience, found a slight improvement in survival following gross total resection, as reported by the operating surgeon (21).

In contrast, several studies, including a large review by Kiely et al. (22) and recent reports from Castel et al., have found that the extent of surgical resection does not affect survival (23). von Schweinitz et al. found that the extent of surgical resection affected only certain subgroups of patients with high-risk disease (24). In addition, substantial complication rates have also been reported after aggressive attempts at removing all gross tumor from the retroperitoneum (25). Shorter et al. also noted that the outcome for patients with stage 4 neuroblastoma depended more on the biologic characteristics of the tumor than on the extent of surgical resection (26). This conclusion is further highlighted by the results of the CCG 3881 study, which showed that, at least in infants with stage 4 disease and single-copy *MYCN* tumors, survival was excellent,

regardless of whether gross total resection (3-year event-free survival [EFS], 91%) or incomplete resection (3-year EFS, 94%) was performed. Recent data suggest that this favorable outcome for patients with stage 4, single-copy *MYCN* disease likely includes patients up to 18 months of age if the tumor's Shimada histology is also favorable (27). Clearly, the excellent prognosis for these patients should not be jeopardized by overly aggressive surgery. However, survival was poor if the tumor was *MYCN* amplified, again, regardless of the extent of surgery; 3-year EFS after total resection was 14%, and 3-year EFS after incomplete resection was 10% ($P = 0.18$) (28). Perhaps, infants with stage 4 disease and *MYCN*-amplified tumors would benefit from more aggressive surgery despite the attendant risks, given that current adjuvant therapy is unlikely to cure them.

The role of cytoreduction by surgical resection is unclear; however, resection of as much gross tumor as possible in patients who receive autologous or allogeneic bone marrow transplant (BMT) in combination with high-dose chemotherapy and total body irradiation (TBI) may be of some benefit. Despite the uncertainty of the role of surgery, the COG high-risk protocol currently recommends attempting gross total resection of the primary tumor and locoregional disease in patients with high-risk neuroblastoma. Most children undergo delayed surgery after the completion of the fifth cycle of induction chemotherapy, even though tumor volume reduction plateaus after the second or third cycle of chemotherapy (29). Other groups are performing surgery as soon as locoregional disease appears, radiographically, to be resectable (30). Although initial surgical resection is often not appropriate for patients with neuroblastoma, the principle of resection at the earliest feasible time should be considered. Since no prospective, randomized studies have been performed, the influence of the extent of surgical resection is unknown. However, in considering aggressive surgical resection, the risks involved, including vascular injury and significant bleeding, kidney or bowel infarction, infection, delay in chemotherapy, and long-term complications (e.g., renal atrophy and diarrhea), need to be weighed carefully against the uncertain benefits of extensive surgery. Certainly removal of kidneys or other organs is to be avoided, as this may hinder or delay the ability to give potentially effective chemotherapy.

OPERATIVE PRINCIPLES

For abdominal/retroperitoneal tumors, a variety of different types of incisions can be used, including upper transverse, midline, and thoracoabdominal approaches. The tumor and adjacent lymphadenopathy should be carefully exposed to determine the relation between the tumor and normal organs and vessels. For a detailed description of extensive surgical resection of neuroblastoma, *see* reference 22. Briefly, the major points espoused by Kiely include: (1) Approach the operation as a vascular-type operation in which identification and skeletonization of the major intraabdominal vessels is critical. If encasement of major vessels such as the aorta, vena cava, or their branches is found, tumor dissection must be performed to free the vessels completely. Generally tumor can be separated from the vessel by dissecting in a subadventitial plane. (2) The tumor should be removed piecemeal. In particular, undue torque on the renal artery in an effort to clear tumor from behind the renal hilum in one piece may result in injury to the intima of the artery with vessel spasm and/or thrombosis, leading to renal ischemia. (3) Dissection commences distal to the lower edge of the tumor, generally along the common or external iliac artery, and proceeds proximally to encounter the tumor along the aorta identifying the major arterial branches (and

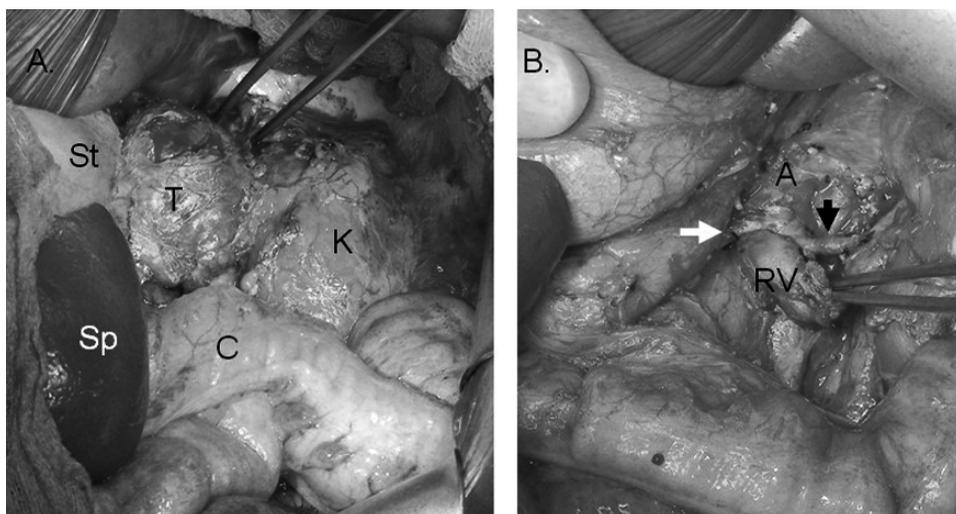


Fig. 2. An intraoperative photograph of a patient undergoing resection of a left adrenal neuroblastoma and adjacent lymphadenopathy. (A) Exposure of the tumor (T) adjacent to the kidney (K) after medial reflection of the stomach (St), spleen (Sp) and pancreas, and inferior reflection of the colon (C). (B) The operative bed after gross total resection of the primary tumor and adjacent lymphadenopathy. With retraction of the left renal vein (RV), the aorta (A) with its superior mesenteric artery (white arrow) and left renal artery (black arrow) can be seen.

left renal vein). With deliberate dissection of the tumor from the mesenteric and renal vessels, injury to the liver, bowel, spleen, and kidneys can be avoided, although this frequently results in piecemeal division and excision of the tumor (Fig. 2). (4) Right-sided tumors are managed similarly and are generally less complicated unless intimately involved with the structures of the porta hepatis. Pelvic tumors may involve the sacral plexus in addition to the iliac vessels. Thoracic tumors are usually more easily cleared of the vascular structures but often extend into the intervertebral foramina. Extraction of tumor from this location is of uncertain benefit and can be associated with significant complications (*see* following section).

Use of the Cavitron ultrasonic aspirator in selected patients may allow for better tumor dissection from the major vessels, with less blood loss and fewer complications (31). Use of the argon beam coagulator and lasers may also help to achieve complete or near-complete resection and, potentially, to reduce operative complications.

Intraspinal Extension of Neuroblastoma

In a subset of patients with paraspinal neuroblastoma, tumor growth may extend into the spinal canal (“dumbbell” tumors) (Fig. 3). If neurologic symptoms result, urgent treatment is required to prevent permanent injury caused by compression of the spinal cord. Each of the three main therapeutic modalities (surgery, radiation therapy, and chemotherapy) has been used in the past. The Pediatric Oncology Group (POG) report by Katzenstein et al. showed similar rates of neurologic recovery in patients treated with surgery or chemotherapy, but significant orthopedic sequelae were seen more commonly in patients treated with surgery (32). Therefore, management generally consists of urgent administration of chemotherapy for patients with stable neurologic symptoms. Emergent surgical intervention is currently reserved for patients

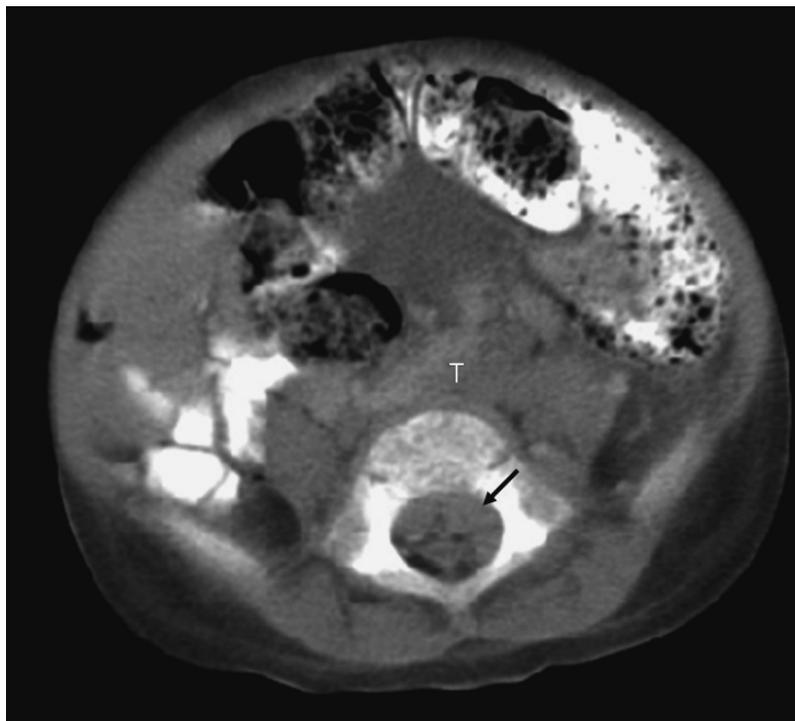


Fig. 3. An abdominal CT scan of a 3-year-old child with disseminated neuroblastoma. The retroperitoneal tumor can be seen (T), as can extension of the tumor within the spinal canal (black arrow). This child presented with lower extremity neurologic deficit.

with an acutely deteriorating neurologic status. Although chemotherapy is probably considered most appropriate for the initial management of these patients, improvements in neurosurgical techniques, including the use of laminotomy instead of laminectomy to access the intraspinal tumor, may necessitate reconsideration of this approach, especially in those patients with acutely progressive symptoms where emergent surgical intervention is the initial treatment of choice.

The appropriate approach for patients with asymptomatic intraspinal tumor extension is also uncertain. For patients with low- or intermediate-risk disease, the risks of attempting to remove the intraspinal component of a paraspinal tumor probably outweigh the benefits. This situation most commonly arises in patients with thoracic primary tumors. The intrathoracic component is resected, and gross residual disease remains in the spinal canal. Care should be taken to minimize surgical complications, such as leakage of cerebrospinal fluid or uncontrollable intraspinal bleeding. Because residual foraminal disease rarely grows to a symptom-developing size, the importance of conservative therapy in this circumstance should be emphasized. In the absence of metastatic disease or unfavorable tumor biology, these patients' disease will be classified as stage 2A/B, low risk, and they have a very favorable prognosis with no further therapy. For patients with high-risk disease, the importance of resecting gross intraspinal disease is uncertain.

Neonatal

Small, localized neuroblastomas in infants younger than 3 months of age tend to regress spontaneously; therefore, the COG has recently opened a protocol (ANBLOOP2) of expectant observation of patients with these lesions to further define their natural history. This study is designed to prove the hypothesis that close biochemical and sonographic observation can be used for safe clinical management of infants with these tumors; surgical resection is reserved for those rare cases in which there is evidence of continued growth.

Stage 4S Neuroblastoma

In 1971, D'Angio, Evans, and Koop reported a number of patients with a “special” variant of metastatic neuroblastoma, termed IVS (now referred to as 4S) (33). These patients were infants who typically had a single, small primary tumor; however, these infants often had extensive metastatic disease in the liver, resulting in significant hepatomegaly, skin nodules (“blueberry muffin” lesions), and small amounts of disease in the bone marrow (<10% of the mononuclear cells). Patients with 4S neuroblastoma were quite remarkable, because the large amount of disease generally underwent spontaneous regression, even without treatment, and the infants ultimately had no evidence of disease (Fig. 4).

Only supportive therapy has been recommended for this stage of neuroblastoma because of the high incidence of spontaneous regression and the resultant good prognosis (34). Most of these patients have tumor with favorable biology (single-copy *MYCN*, favorable Shimada histology, and DNA index > 1); therefore, they are assigned to the low-risk classification and receive no therapy. However, despite the generally benign course of their malignancy, these infants can die of complications caused by

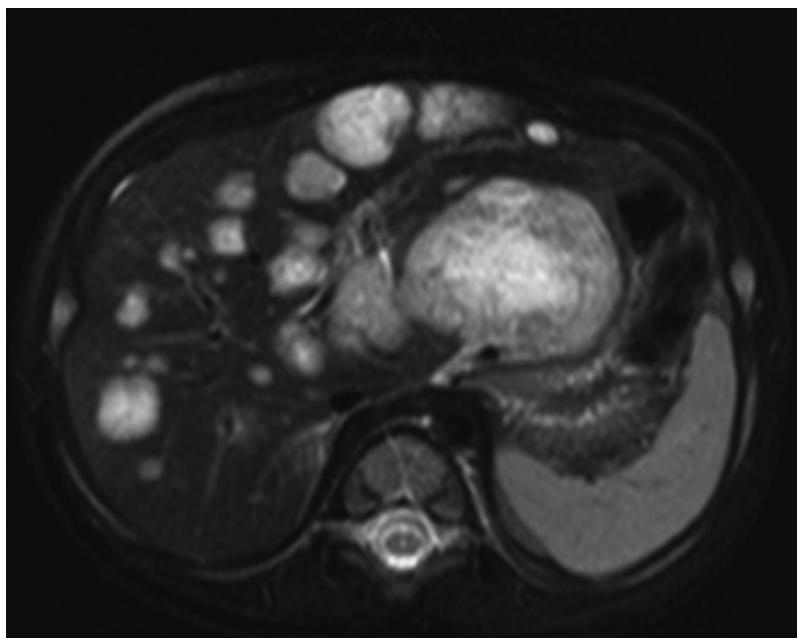


Fig. 4. An abdominal magnetic resonance imaging of a 2-month-old child with stage 4S neuroblastoma demonstrating extensive, multifocal liver involvement.

the initial bulk of their disease. Limited chemotherapy, local irradiation, or minimal resection can be used to treat infants with life-threatening symptoms of hepatomegaly. Operative placement of a Silastic pouch as a temporary abdominal wall may be a choice for those with significant liver enlargement that causes either respiratory compromise secondary to diaphragmatic elevation or obstruction of the inferior vena cava (35). This procedure may help to avoid life-threatening events until shrinkage of the liver is achieved by either spontaneous regression or therapy.

Recurrent Disease

Patients with recurrent high-risk neuroblastoma have a nearly uniformly dismal outcome. Nevertheless, surgery may have a limited role in the management of patients with relapsed neuroblastoma where the recurrence has been documented to be localized and refractory to available chemotherapeutic agents.

REOPERATIVE SURGERY

Reoperative surgery for children with neuroblastoma, caused by either disease progression or recurrence, is rare, other than for delayed resection of the primary locoregional disease after an initial biopsy and neoadjuvant chemotherapy. The reasons for this are twofold. In children with low- or intermediate-risk disease, the likelihood of disease recurrence or progression is extremely low; whereas the appropriateness of surgical resection of recurrent, high-risk disease is very limited, as previously mentioned, given the extremely poor prognosis for these patients and the high probability of widely disseminated disease in this setting. On rare occasion, staged attempts at achieving gross total resection of locoregional high-risk disease prior to undergoing high-dose consolidative chemotherapy with autologous stem cell rescue may be undertaken, although the value of this approach is unproven.

Reoperative surgery may also be undertaken for reasons relating to complications of the surgery. Postoperative bowel obstruction requiring surgical intervention can occur because of adhesions or intussusception (Fig. 5). Occasionally, reoperation for surgical

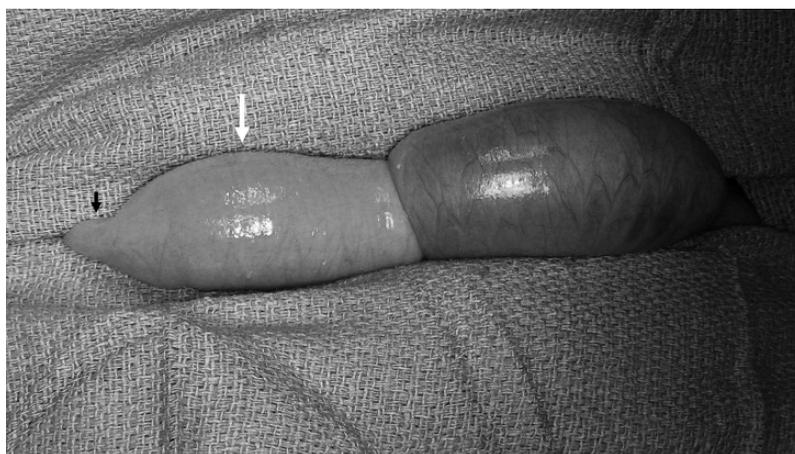


Fig. 5. Midjejunal intussusception in a patient reexplored for a bowel obstruction 6 days after an adrenalectomy for a localized, stage I neuroblastoma. At the right is the dilated, fluid-filled proximal small bowel. The black arrow shows the collapsed, more distal bowel. The white arrow shows the intussusciens containing the intussusceptum.

closure of a lymphatic channel will be required for persistent chyle leak in the abdomen or chest that is refractory to or persistent despite conservative measures.

SUMMARY

The treatment of neuroblastoma requires a multidisciplinary approach. Although surgical resection may be the only therapy required for patients at low-risk of disease recurrence, the surgeon provides but one element of the modern multimodal treatment of children with high-risk disease. However, it is essential that the surgeon be aware of the heterogeneous nature of neuroblastoma and the factors that have, thus far, been identified in predicting the risk for relapse of children with this disease. An appreciation for the different clinical scenarios and how they impact on the need for tissue to perform necessary studies and the role of surgery with respect to timing and extent of tumor resection is essential for the optimal care of these children.

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Wilms' Tumor

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Wilms' tumor, also known as nephroblastoma, is the predominant renal cancer of childhood, affecting approximately 500 new patients in North America per year (1). Nephroblastoma was first recognized and described by Max Wilms in 1899 (2). The tumor arises from embryonic kidney precursor cells that obtain malignant qualities because of various degrees of genetic instability. Even though the majority of diagnosed Wilms' tumor are unilateral and arise from sporadic mutations, approximately 10% of patients with Wilms' tumor have bilateral or multifocal tumors and other associated malformations and/or clinical syndromes. Inherited alterations of the 11p13 locus including the *WT1* gene are responsible for syndromes including WAGR (Wilms' tumor, aniridia, genitourinary abnormalities, and mental retardation), Denys-Drash, and Frasier (3). Additionally, alterations within the 11p15 locus are also associated with Wilms', in particular, Beckwith-Wiedemann syndrome.

Wilms' tumors are typically responsive to chemotherapy; therefore, a strategy consisting of nephrectomy and adjuvant chemotherapy is the basis of most validated treatment protocols. Additional interventions, such as preoperative chemotherapy for unresectable disease and radiation therapy for primary and metastatic disease, have also been employed and shown to improve outcomes in particular clinical circumstances. The surgical standard of care is total nephrectomy of the affected kidney with lymph node sampling. Partial nephrectomy can be used in a small subset of patients but is usually not considered in unilateral disease because of the low occurrence of renal failure after unilateral nephrectomy.

In oncology, reoperative surgery has several meanings depending on the clinical context. The need for additional surgery owing to disease recurrence or metastatic

disease would be considered reoperative surgery. Because some of the primary difficulties of reoperative surgery are inflammatory changes and adhesions caused by the first surgery, a tumor resection after neoadjuvant chemotherapy or radiation therapy must fall under this category because of the multitude of challenges encountered in a posttreatment resection. In this chapter, we will focus on the surgical management of inoperable tumors treated with neoadjuvant therapy, tumor with major venous extension, bilateral Wilms' (synchronous or metachronous), metastatic disease, and disease recurrence.

UNRESECTABLE WILMS' TUMOR

Clinical Presentation

Wilms' tumor that is deemed unresectable has a clinical presentation similar to operable primary nephroblastoma. On average, patients are approximately 3 years of age at diagnosis of Wilms' tumor. Patients who undergo preoperative chemotherapy for an initially unresectable tumor are older, on average, than patients undergoing primary nephrectomies, with 61% older than 4 years of age (4). The typically large, round, and firm abdominal/retroperitoneal mass is detected either by parents during routine care of the child or by the primary care physician during well-child examinations. Common clinical signs include, abdominal pain, anorexia, weight loss, gross or microscopic hematuria, hypertension, anemia, polycythemia, and/or fever. Patients can also have acquired von Willebrand's disease warranting a coagulation evaluation.

Diagnostic Evaluation

Invariably, the first study to be ordered in evaluating an abdominal mass is a plain radiograph of the abdomen. This usually reveals visceral displacement caused by the mass effect of the tumor. Calcification within the mass can sometimes be appreciated as well. Ultrasound is the first test to order in evaluating a suspected Wilms' tumor. This will enable the surgeon to differentiate between a cystic or solid mass and to determine renal vein/inferior vena caval (IVC) extension. On ultrasound, Wilms' tumor typically appears as a solid lesion with a pseudocapsule made up of compressed renal parenchyma (5). In addition, ultrasound screening is suggested in children with Wilms' tumor-associated syndromes at 3-month intervals until 6–8 years of age.

Computed tomography (CT) is very useful in the work up of Wilms' tumor. With intravenous contrast, the mass will have less contrast enhancement than the normal renal parenchyma defining the tumor margin. Because Wilms' tumor arises from the kidney progenitor cells, the tumor appears within the kidney with normal surrounding and medially displaced renal parenchyma and collecting system. Occasionally, curvilinear calcification can be seen within the tumor. CT is also useful for assessment of synchronous, contralateral disease and tumor spread into lymph nodes, renal vasculature, IVC, and adjacent organs. Presence of lung and liver metastasis is also assessed with CT. CT scan of the brain should be obtained in the postoperative setting for rhabdoid variant, and bone scan for clear cell variant of Wilms' tumor. Magnetic resonance imaging (MRI) can assist with intravascular evaluation of tumor extension. Resectability can be assessed with imaging but only as an adjunct to actual surgical exploration.

Surgical Exploration, Biopsy, and Staging

Wilms' tumor is deemed inoperable based upon the following criteria defined by Ritchey et al. (4): (1) tumor found to be unresectable at surgical exploration; (2) inoperability determined by clinical evaluation or diagnostic imaging; (3) extensive intravascular tumor extension above the level of the hepatic veins; and (4) other patient-related morbidities secondary to the tumor. Disease stage is only accurately assessed with surgical exploration to determine extent of tumor, lymph node spread, and obtain a histologic diagnosis with intraoperative biopsy.

The North American-based, National Wilms' Tumor Study group (NWTSG) and the European-based, International Society of Pediatric Oncology (SIOP) have somewhat opposing views on this issue. NWTSG has previously recommended initial surgical exploration to accurately stage the patient prior to initiating a stage-appropriate treatment protocol. If the tumor is deemed operable, a complete resection is performed followed by adjuvant therapies based on staging. SIOP recommends basing the diagnosis of Wilms' tumor solely on clinical history and imaging. From this, the patient is staged and placed on a stage-appropriate, preoperative chemotherapy. The patient then undergoes delayed surgical exploration and resection of primary tumor.

There are some proponents of percutaneous biopsy, rather than surgical exploration, as a strategy for staging patients prior to initiation of preoperative chemotherapy. Saarinen et al. state that percutaneous needle biopsy of Wilms' tumor in children is safe and prevented misdiagnosis and understaging in one patient with unfavorable histology and one patient with rhabdoid variant (6). However, this paper also reports two patients that had subcapsular intratumoral bleeding. The United Kingdom Children's Cancer Study Group (UKCCSG) Wilms' Tumor Study-3 adopted percutaneous needle biopsy for tumor staging prior to initiation of chemotherapy (7). They reported the associated morbidities with needle biopsy, which included: decrease in hemoglobin (20%), local pain (19%), one patient with massive intratumoral bleeding requiring operative intervention, and one patient with tumor rupture who subsequently died. Also, needle track seeding and recurrence have been reported complications of percutaneous biopsy (7,8). Moreover, NWTSG-5 demonstrated that unfavorable histopathologic features are inhomogeneously distributed within a given Wilms' tumor, leading to the suggestion that neither percutaneous nor open biopsy is infallible for accurate staging.

The concept of staging with only clinical and imaging data has not previously been practiced by the NWTSG for two reasons: (1) misdiagnosis, and (2) inaccurate staging. Because many other types of pediatric malignancies can arise in the perirenal, retroperitoneal space, misdiagnosis of Wilms' tumor is a very real possibility. Institutions using the SIOP protocols have mistakenly diagnosed mesoblastic nephroma and renal cell carcinoma as Wilms' (9,10). These misdiagnosed tumors are treated with the assigned preoperative Wilms' tumor chemotherapy regimen and only correctly diagnosed after tumor resection 6–8 weeks later. Although this is less of an issue for renal cell carcinoma, for which there is no effective neoadjuvant chemotherapy at present, there is more concern regarding administration of cytotoxic chemotherapy to patients with benign histology such as mesoblastic nephroma.

The primary concern of the NWTSG is inaccurate staging. The treatment protocols for stage I and II disease currently do not differ; however, in the next Children's Oncology Group (COG) Wilms' tumor protocol, stage I, favorable histology tumors

less than 550 g, will not receive adjuvant chemotherapy. For now, the main concern is understaging a stage III or IV patient as a stage I or II. Gow et al. retrospectively reviewed a series of patients with nonmetastatic Wilms' tumor (stages I–III) to compare preoperative CT staging to histologic and operative staging (11). They found that CT correctly staged only 38% of patients and that 40% of histologically confirmed stage III patients were categorized as stage II by imaging alone. In addition, 20% of the histologic stage I and 33% of histologic stage II patients were staged as stage III by CT scan. Additional studies have shown inaccurate staging of stage III/IV patients as stage I/II based on imaging in 10–14% of the patients studied using the SIOP protocols (10,12).

Regardless of these discrepancies in image-based and histologic staging, outcomes on SIOP protocols do not differ greatly from the NWTSG protocols. This has prompted NWTSG to reevaluate their approach and incorporate some image-based staging into upcoming trials. In all new protocols, any tumor spill, including open or needle biopsy will automatically classify the patient as stage III. In part because of this change, tumor biopsy will not be required at presentation for patients with bilateral Wilms' tumor, *see* following section. Given that any tumor that is deemed unresectable at presentation is stage III, an adequate biopsy should be performed in this circumstance in order to tailor neoadjuvant chemotherapy.

Because imaging is not adequate for assessment of lymph node involvement, patients who have inadequate lymph node sampling may need to be upstaged in order to prevent undertreatment. Specifically, patients in the very low-risk group of age less than 2 years, stage I tumor less than 550 g, and favorable histology will not receive adjuvant chemotherapy or radiation therapy, but in order to qualify for this treatment arm the patient will have had adequate lymph node sampling. Similarly, patients with stage I clear cell sarcoma of the kidney with biopsy proven negative lymph nodes will be spared radiation therapy.

Prognostic Factors

The most significant prognostic factor for Wilms' tumor is unfavorable histology, defined as the presence of anaplasia (reviewed in (13)) (Fig. 1). The results of NWTSG-1 showed that greater than 50% of the patients diagnosed with unfavorable histology Wilms' tumor died as a result of disease versus less than 7% of patients with Wilms' tumor lacking histologic anaplasia (14). Since then, the definition and subclassifications of anaplasia have been redefined to increase prognostic power. Faria et al. describe focal anaplasia as one or a few discrete foci in the primary tumor and diffuse anaplasia as nonlocalized or located beyond the primary tumor capsule (15). With this definition, there was a striking difference in outcomes, with focal anaplasia associated with a 3% mortality and diffuse anaplasia with 96% mortality.

Other prognostic factors have proven useful throughout the various treatment trials for Wilms' tumor. More recently, chromosomal deletions at 16q and 1p causing a loss of heterozygosity (LOH) at these loci have been associated with poorer prognosis in both favorable and unfavorable Wilms' tumor (16,17). In addition, these studies revealed that 16q LOH, which was the most predictive of outcome, was more frequent in unfavorable versus favorable histology, anaplastic Wilms' (32 versus 17%, respectively). NWTSG-1 through NWTSG-3 have reported significant prognostic factors for regional disease (stages I–III), including disease that is inoperable at diagnosis (18–20).

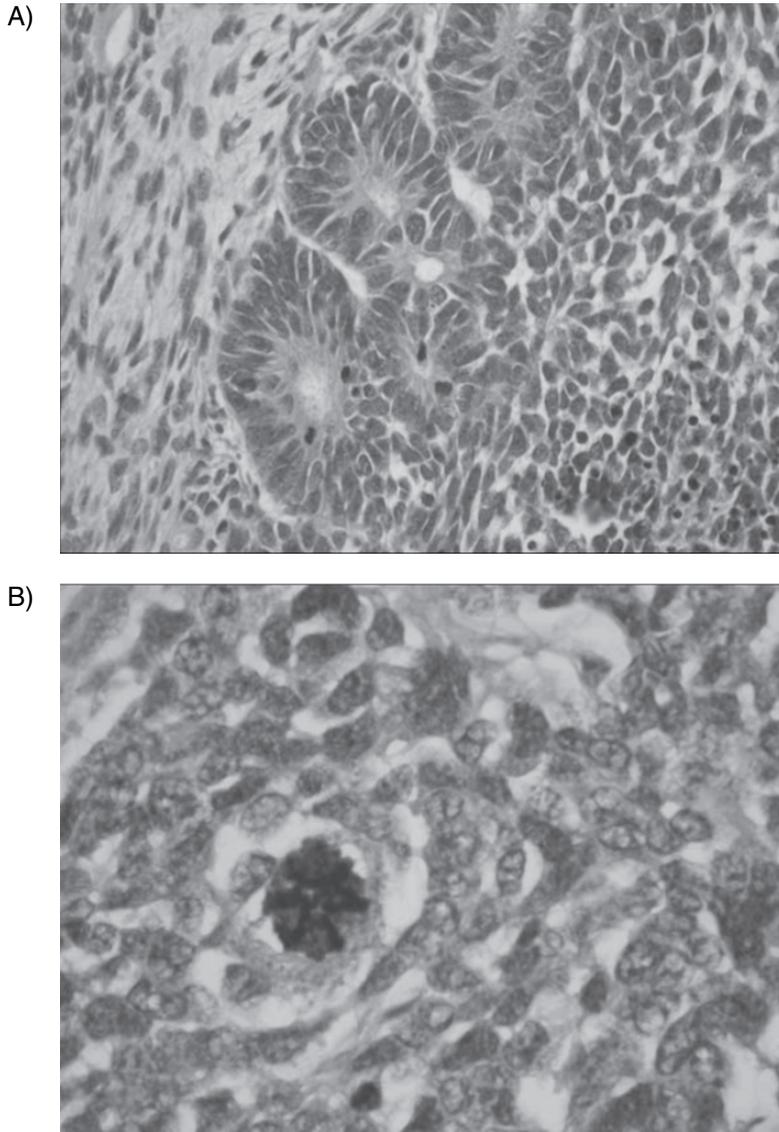


Fig. 1. (A) Favorable histology Wilms' tumor. (B) Unfavorable histology or anaplastic Wilms' tumor.

NWTS-1 identified unfavorable histology, tumor weight (>250g), and lymph node-positive disease as the most important predictors of mortality. Relapse was also associated with these factors, as well as single-drug therapy and age greater than 24 months. The results of NWTS-2 were similar but found that age and tumor weight really had no predictive value. NWTS-3 reiterated that lymph node spread, age at diagnosis, and tumor weight were predictive of outcome; moreover, this trial also identified that microscopic disease at the tumor margin was an independent risk factor for relapse and death in stage II and III patients.

COG Guidelines

The COG guidelines from the NWT5-5 trial for the treatment of inoperable tumors involve a multimodality approach. The first step is to perform exploratory surgery for a first-hand assessment of resectability and to obtain a biopsy. All suspicious lymph nodes and intraabdominal metastatic lesions should be sampled. For treatment centers that choose to stage and treat patients prior to surgery based on imaging and biopsy, patients should be categorized as stage III and treated accordingly. At 5 weeks after initiation of treatment, serial imaging is used in the preoperative chemotherapy or inoperable tumor open biopsy setting to monitor tumor response. Once the tumor size and extent has responded to therapy and the tumor is deemed operable, an operative resection can be planned. If there is no response to therapy, a second open biopsy should be performed to sample the tumor, nodes, and metastasis in order to assist in planning more intense medical therapies. The COG also emphasizes the importance of either pre- or postoperative radiation therapy to reduce intraabdominal recurrence.

Operative Strategy

Very few Wilms' tumors will remain unresectable after neoadjuvant chemotherapy, although the resection may still be difficult, especially if the tumor is adherent to multiple adjacent structures. The choice of incision for a large Wilms' tumor is based on the specific anatomy of the tumor. In the presence of extensive disease in the cephalad retroperitoneum or significant involvement of the proximal aorta or vena cava, a thoracoabdominal approach can offer distinctive advantages over the standard transverse upper abdominal incision. Whichever approach is used, it should be large enough to allow mobilization and dissection of the tumor with minimal risk of intraoperative complications. When an adequate incision is used, it facilitates exposure of the key vascular structures and decreases the tendency toward overly vigorous retraction of the tumor, thereby minimizing the risk of vascular compromise/hemorrhage or tumor rupture.

The initial conduct of the operation is similar to that for an up-front resection in a patient who has not received neoadjuvant chemotherapy. For a right-sided lesion, the lateral and cephalad attachments of the ascending colon and hepatic flexure are divided, and the mesocolon is dissected off the anterior surface of Gerota's fascia. If the tumor arises in the superior pole of the kidney, or if it is adherent to the liver, it is often helpful to divide the triangular ligament of the right lobe of the liver to gain mobility and aide with exposure. For tumors that efface, and possibly invade the vena cava, it is best to obtain control of this structure proximal and distal to the area of concern.

For a right-sided tumor, the fibrous areolar tissue overlying the vena cava is incised longitudinally and the right renal vein is identified, dissected out, and surrounded with a vessel-loop. The right renal artery is located parallel and posterior to the vein. Although it is ideal to control and divide the renal vein and artery prior to any manipulation of the tumor, at times it can be difficult to positively identify the renal artery from an anterior approach. Under these circumstances, it is safer to mobilize the kidney and rotate it medially in order to expose the posterior aspect of the hilum and assure that the artery is in fact entering the renal sinus. Large Wilms' tumors can distort the relationship between the aorta and the renal arteries; therefore, it is incumbent on the surgeon to trace the artery from the involved kidney prior to ligation. After dividing

the renal vessels, the ureter is identified, traced to the pelvic brim, suture ligated, and divided. Finally the tumor and kidney are dissected out circumferentially and removed. It is standard practice to remove the adrenal gland if it is not separated from an upper pole tumor by a distinct tissue plane. The approach to a left-sided tumor is similar to the right side; again, care should be taken to positively identify the renal vessels for large tumors that distort the retroperitoneal anatomy.

Wilms' tumors rarely invade the liver, spleen, pancreas, or colon, although the tumor may be somewhat adherent to these organs. It is not normally necessary to perform en-bloc resection of these organs, but rather it is better to seek a plane of dissection that will allow separation. For nonvital structures such as diaphragm, psoas muscle, or tail of pancreas, it may be advantageous to resect a small portion of these tissues rather than risk entering the tumor.

Outcome

The majority of patients with nonmetastatic, initially inoperable Wilms' tumor have stage III disease based on lymph node involvement, tumor spillage owing to initial biopsy or difficulty of resection, peritoneal involvement, incomplete tumor resection (positive margins on surgical specimen), or piecemeal tumor removal. The NWTSG encourages continued treatment of these patients as stage III regardless of preoperative chemotherapy effects on the tumor. The SIOP treatment protocols use a postchemotherapy staging system to determine the appropriate treatment regimens for each patient. A SIOP trial performed to determine the necessity of abdominal radiation after chemotherapy in patients with stage II, node-negative disease was stopped early because of an unacceptably high rate of local recurrence in those subjects who did not receive abdominal radiation (21).

Patients with stage III Wilms' and favorable histology have a 4-year relapse-free survival of 90% and overall 4-year survival of 95% (22). The treatment recommendation for this group is: nephrectomy with lymph node sampling, abdominal radiation, and a 24-week chemotherapy regimen (NWTS Regimen DD-4A) consisting of vincristine, doxorubicin, and dactinomycin. For patients with diffuse anaplasia on histology, the 4-year overall survival is decreased to 56%. They receive a modified 24-week chemotherapy regimen (NWTS Regimen I) with the addition of etoposide, cyclophosphamide, and mesna (no dactinomycin). In NWTS-3, the complication rate reported for patients who underwent delayed resection after neoadjuvant chemotherapy was 19%, similar to that of patients subjected to primary nephrectomy without preoperative therapy (4,23). However, in NWTS-4, the complication rate was lower, 12.7%, because of decrease in extensive intraoperative bleeding and major intraoperative complications (24).

WILMS' TUMOR WITH MAJOR VENOUS EXTENSION

Epidemiology and Clinical Presentation

It is generally accepted that intravascular extension occurs in 4–10% of patients with Wilms' tumor (25). In NWTS-4, 165 out of 2731 (6%) patients had IVC and/or right atrial involvement (26). Out of these patients, 19% had atrial involvement and 81% had disease extending only into the IVC. Wilms' tumor with caval and/or atrial extension is considered stage II if it is resected en bloc with the primary tumor; however, if the primary tumor and intravascular extension are removed separately, the patient is

considered a stage III (2). Varicocele, heart failure, hepatomegaly, and ascites are unique clinical features of Wilms' tumor with intravascular extension secondary to obstruction of venous return. However, these symptoms are only present in approximately 5% of the patients, with the majority presenting with the common Wilms' tumor symptoms discussed previously (26,27).

Diagnostic Evaluation

Ultrasound is the most appropriate initial study for an infant or child with an abdominal mass. In Wilms' tumor, this modality offers the additional advantage of accurate evaluation of the intravascular extension of the tumor (Fig. 2A). Specifically, amplitude-coded color Doppler sonography is recommended for this particular evaluation (5). CT can also reveal intravascular extension; however, this is a less sensitive test because of artifact from nonopacified blood (Fig. 2B). MRI can be used to map intravascular extension and may be superior to ultrasound for operative planning in patients with intravascular tumor (28).

Operative Strategy

Preoperative imaging will normally identify intravascular tumor extension, but the surgeon should routinely palpate the renal vein and IVC prior to division of the vein to be certain that tumor is not present. If intravascular extension is limited to the renal vein and the vena cava below the level of the hepatic veins, it can normally be removed en bloc with the kidney at the initial tumor resection. In this circumstance, the vena cava is dissected out and surrounded with vessel loops above the level of tumor extension and below the renal veins, and the contralateral renal vein is isolated in a similar fashion. The kidney and tumor are then resected, except for division of the renal vein. Polypropylene stay sutures are placed in the vena cava proximal and distal to the renal vein, the vessel loops are cinched down, and the vena cava is opened at the renal vein junction. In most cases the tumor will be free-floating within the lumen and can be gently manipulated back through the renal vein orifice. A Foley catheter can also be used to aid with tumor extraction. In some circumstances, the tumor will be tightly adherent to the venous intima. In these cases, the venotomy should be extended proximally to near the cephalad extension of the tumor. The technique is similar to a carotid endarterectomy requiring use of a Freer periosteal elevator or fine hemostat to separate the tumor from the wall of the vessel. If at all possible the tumor should not be transected, as this will upstage the patient to stage III. After tumor extraction, the vena cava is closed with running polypropylene suture, with care taken to de-air the vessel prior to tying the suture and releasing the proximal vessel loop.

Primary resection of intravascular tumors that extend above the level of the hepatic veins is associated with a high rate of complications, especially when the right atrium is involved. These patients are treated with preoperative chemotherapy in an attempt to shrink the tumor thrombus below the hepatic outflow. If the tumor does not decrease below this level after chemotherapy, the resection should be performed with the aid of cardiopulmonary bypass. The kidney and tumor are dissected out and the renal artery and ureter are divided. The patient is then heparinized and placed on bypass. After hypothermic circulatory arrest is initiated, the right atrium and vena cava are opened and the tumor is removed either with traction or using a Foley catheter technique. The lumen should be carefully flushed prior to closure to remove any stray tumor fragments. Circulation is then restored.

Outcome

Both NWTSG and SIOP studies have determined that there is no significant survival difference between Wilms' tumor with or without intravascular extension (25,26). The primary differences lie in the frequency of complications. Caval and atrial surgery require more extensive dissection, longer operative time, and, in most cases of atrial extension, cardiopulmonary bypass. Shamberger et al. report a complication rate of 17% with IVC extension and 37% in disease with atrial involvement (26). The most recent summary of surgical complications in NWTS-4 identified caval and/or atrial extension as a significant risk factor for complications in both univariate (odds ratio = 5.66) and multivariate analysis (odds ratio = 3.77) (24). Complications include: intraoperative hemorrhage, bowel obstruction, tumor embolus, IVC obstruction, respiratory distress syndrome, wound dehiscence, aortic thrombosis, paraparesis, hepatic vein injury, cerebral ischemia, pancreatitis, and postpericardiotomy syndrome.

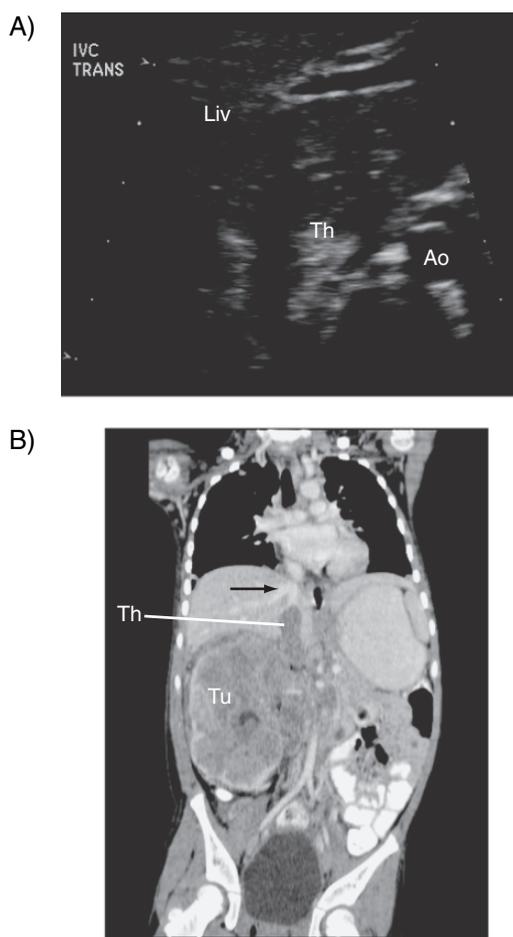


Fig. 2. (A) Ultrasound showing cross-sectional view of the liver (Liv), inferior vena cava (IVC) tumor thrombus (Th) extending from primary tumor, and the aorta (Ao). (B) Coronal reconstruction CT scan showing tumor thrombus (Th) extending from the right kidney primary tumor (Tu) into the IVC. Distal to the thrombus, the patent hepatic vein and IVC (black arrow) can be seen draining into the right atrium.

The most recent NWTSG and SIOP recommendations are for preoperative chemotherapy in patients with tumor extension above the level of the renal veins. This can cause tumor regression from suprahepatic or atrial locations helping to avoid more involved dissections and cardiopulmonary bypass. In NWTSG-4, it was noted that patients who had received preoperative chemotherapy had more adherent intravascular tumor thrombus compared to those who were not treated preoperatively (62 versus 44%, respectively) (26). In approximately one-half of the patients that had received preoperative chemotherapy, unresectable tumor thrombus remained after resection; however, there was no increased risk of relapse. In addition, preoperative chemotherapy was associated with other rare complications, such as tumor embolism and respiratory distress syndrome. In general, the complication rate and overall survival in the primary resection group versus preoperative chemotherapy with subsequent surgery group did not reach statistical significance.

BILATERAL WILMS' TUMOR

Epidemiology and Clinical Presentation

Synchronous, bilateral tumors account for approximately 5% of all nephroblastomas (1). Patients presenting with synchronous, bilateral Wilms' tumor are, on average, 2–3 years of age (29–31). Metachronous lesions occur in approximately 1–2% of all patients presenting with unilateral Wilms' tumor (32). The incidence of metachronous lesions increases to approximately 4% in children diagnosed with unilateral Wilms' tumor at less than 1 year of age.

Symptoms at presentation are very similar to unilateral Wilms' as discussed previously. In a case series by Tomlinson et al., all of the patients presented with a palpable abdominal mass and some patients with palpable mass and macroscopic hematuria (29). Patients with bilateral disease can also present with hypertension and cardiomyopathy (31).

Diagnostic Evaluation

The imaging strategy outlined previously also applies to diagnostic evaluation of bilateral Wilms' tumor. Because 5% of unilateral Wilms' are associated with a synchronous lesion, thorough imaging analysis of the contralateral side must be performed with ultrasound, CT scan, and/or MRI (Fig. 3A). A study by Ritchey et al. showed that CT is more sensitive than ultrasound for detecting synchronous lesions; however, 7% of the patients with bilateral disease would have been missed with imaging alone (33). Because of this, intraoperative evaluation of the contralateral kidney is not reserved for indeterminate imaging and clinical suspicion.

Prognostic Factors

The same histologic, prognostic factors described previously apply to bilateral disease. The most important risk factor for synchronous disease or development of metachronous disease is the presence of nephrogenic rests. Nephrogenic rests are thought to be precursor lesions for Wilms' tumor. These can be subdivided into two categories: perilobar and intralobar. Nephroblastomatosis refers to diffuse or multiple

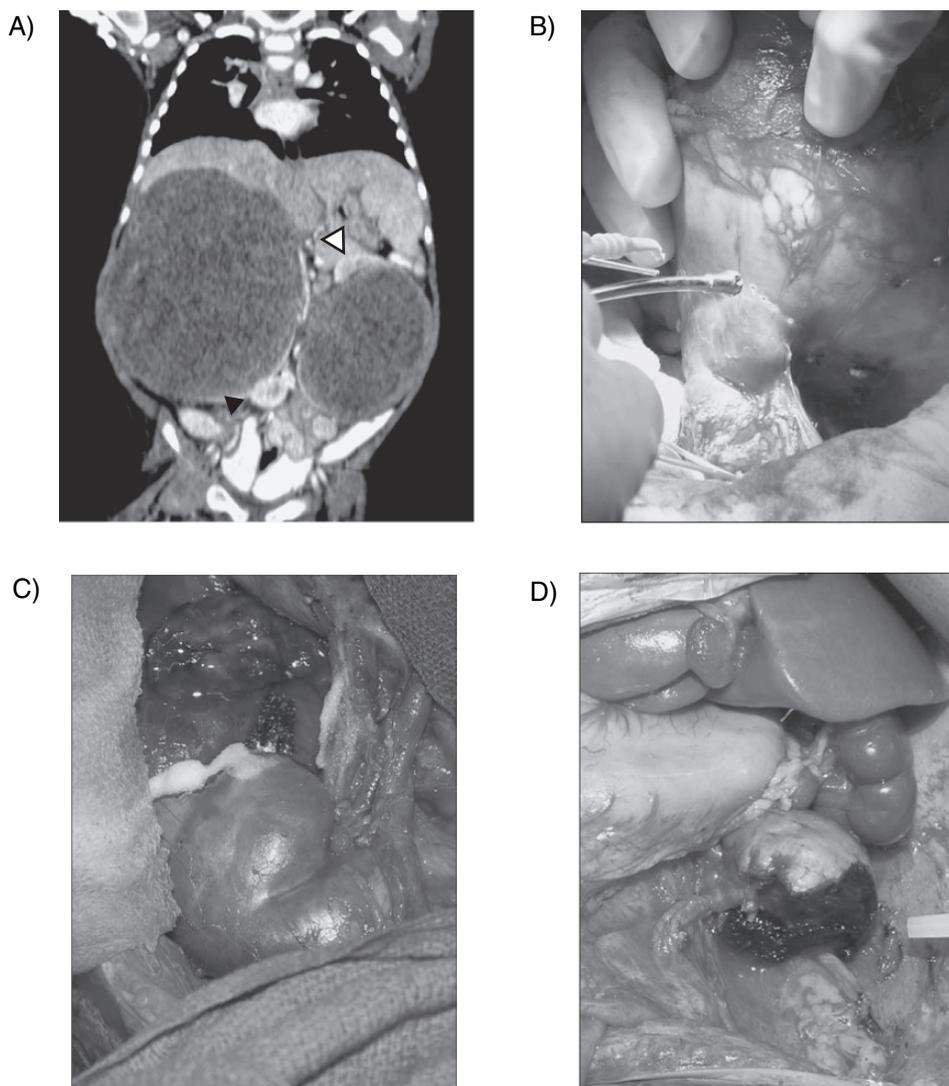


Fig. 3. (A) Coronal reconstruction CT scan showing bilateral Wilms' tumors. Normal kidney parenchyma can be seen on the right (black arrowhead) and left (white arrowhead) sides. (B) Intraoperative image showing dissection of the right-sided tumor from the superior portion of the kidney sparing the remainder of the kidney. (C) Right-sided kidney remnant seen *in situ* with cauterized superior margin with Floseal. (D) Left-sided kidney remnant seen *in situ* with cauterized inferior margin.

rests. Perilobar nephrogenic rests in the resected specimen for unilateral Wilms' are strongly associated with presence of synchronous bilateral disease, a mean age of 3 years, and Beckwith-Wiedemann syndrome (34,35). Intralobar nephrogenic rests were associated with metachronous disease, a younger mean age (16 months), and Denys-Drash syndrome. For unilateral disease with nephrogenic rests, serial ultrasound imaging of the contralateral kidney is recommended until 5–6 years after initial diagnosis, at which time the risk is minimal (36).

COG Guidelines

The NWTS-5 originally recommended performing partial nephrectomy at the initial surgery if the tumors could be completely removed while preserving adequate renal function. The most recent guidelines recommend not performing an initial operation at all. After the diagnosis is made radiographically, the patient should receive 6 weeks of chemotherapy followed by imaging to determine tumor response. If the tumors seem amenable to partial nephrectomy, surgery should be performed at this time. If there is a partial response, the patient can be given more chemotherapy and reassessed after an additional 6 weeks for surgery. If response is poor, bilateral open biopsies can be performed at the 6-week mark to plan more intensive therapy. In the setting of extensive, unresponsive disease in one kidney and responsive or partially responsive disease in the other kidney, therapy should be tailored to achieving a complete resection in the salvageable kidney and radical nephrectomy of the unresponsive kidney. Radiation therapy is reserved for the partially responsive or unresponsive tumors after second-look surgery.

Operative Strategy

CT or MRI with reconstruction of the renal vascular anatomy can be very helpful when planning partial nephrectomy. After an adequate skin incision, the kidney and tumor are mobilized in a standard fashion. The arterial and venous anatomies are defined, and separate branches are tagged with vessel loops, should clamping or ligation be needed. If the extent of tumor invasion into the kidney is unclear, intraoperative ultrasound will help to define a plane of transection that will give an adequate margin of normal kidney, ideally 2 mm. Sonographic imaging can also help predict the involvement of the renal collecting system, which will help the surgeon anticipate the need for repair. The kidney is transected with cautery while the assistant compresses the adjacent parenchyma to minimize bleeding (Fig. 3B). Hemostasis on the cut surface is controlled with both fine Monocryl sutures for larger vessels and Floseal for parenchymal bleeding (Fig. 3C and D). The collecting system is repaired using similar suture material and gently leak-tested by injecting it with dilute methylene blue dye through a fine needle.

Outcome

For bilateral Wilms' tumor, the outcome is worse than the 90% overall survival seen in all stages of unilateral disease. Data from NWTS-3 and -4 show that the 10-year survival approaches 80% for synchronous disease, with no difference in survival noted between preoperative chemotherapy versus immediate resection (37,38). The utility for preoperative chemotherapy in bilateral Wilms' tumor is to increase the likelihood of a successful bilateral nephron-sparing nephrectomy (39). The main determinants of prognosis are similar to unilateral disease: presence of unfavorable histology, age at diagnosis, and highest stage of the individual tumors. Shamberger et al. have recently reported a distinct survival difference between bilateral Wilms' tumors that are responsive to preoperative chemotherapy and those that are not (37). The overall survival for nonresponders is 67 versus 86% in those that respond to therapy. The main reason for nonresponsive disease is undiagnosed anaplastic histology, which is better treated with immediate resection followed by intensive chemotherapy (40). The recommendations from this study are for open biopsy in patients with less than 50% tumor

reduction after two courses of therapy and early surgical resection for nonresponsive disease.

In metachronous disease, survival is much worse than synchronous bilateral disease (41,42). A more recent series of 108 patients with metachronous Wilms' tumor showed a 10-year overall survival of 47%. Paulino et al. further analyzed the survival data according to time interval to second tumor development and found that contralateral tumors arising less than 18 months from the first tumor have a 10-year overall survival of 40%, and those developing at or after 18 months have a survival rate of 55% (43).

Bilateral Wilms' tumor is a risk factor for end-stage renal disease (ESRD). Uremia can be caused by progression of Wilms' tumor, necessitating total nephrectomy, radiation injury, or hyperfiltration in patients with relatively small amounts of renal parenchyma. In contrast to patients with unilateral tumors, in whom the overall incidence of ESRD is 1.3%, the cumulative risk of ESRD in patients with bilateral Wilms' tumor is 15% at 20 years. The incidence is much higher in patients with Denys-Drash syndrome (50%), WAGR syndrome (90%), or with hypospadias or cryptorchidism (25%), whereas patients without these characteristics have an incidence of 11.5% (43).

METASTATIC DISEASE

The most common sites for metastasis in stage IV Wilms' tumor are the lung and liver. Lung metastasis are the most common, occurring in approximately 80% of all metastatic disease; whereas liver metastasis occurs in approximately 15%. Breslow et al. determined that specific metastatic sites at diagnosis had little prognostic value. Survival was dependent on histology and staging of the primary tumor; however, metastasis developing during or after treatment had a significant effect on survival, where liver metastasis conferred a much worse prognosis than lung (44). More recently, Varan et al. reported that metastatic site at diagnosis did effect survival (45). They identified an overall survival of 50% for patients with lung metastasis only and 17% for patients with liver metastasis. The NWTSG reports that the overall survival for patients with stage IV, favorable histology is 80% using a treatment regimen consisting of immediate nephrectomy, postoperative whole lung irradiation, abdominal irradiation according to local stage of primary tumor, and 24 weeks of chemotherapy (22,46). Patients with unfavorable histology of the primary lesion receive the same therapy with the addition of etoposide, cyclophosphamide, and mesna; however, the 4-year survival rate is only 17% (40). In general, resection of metastasis is reserved for persistent or relapsing disease, which will be discussed in the next section.

RECURRENT WILMS'

Disease relapse occurs in less than 20% of patients after treatment of the primary Wilms' (47,48). The characteristics correlating with the best prognosis for relapsed disease include: favorable histology, abdominal recurrence without prior irradiation, recurrent disease confined to the lung, stage I primary disease, primary disease treated with two drug regimen, and recurrence occurring at or after 12 months from diagnosis of primary disease (49). Adverse prognostic factors of relapsed disease include: recurrent abdominal disease in setting of irradiation, tumor positive lymph nodes, two or more recurrent lesions, relapse less than 12 months after diagnosis of primary disease, bone

or brain metastasis, stage IV primary disease, unfavorable histology, and thoracic and abdominal recurrence (50).

The prognosis for recurrent disease has improved over the years; however, overall survival rates are not equivalent to the greater than 90% survival seen with treated primary disease. The 3- to 4-year overall survival of relapsed patients observed in NWTS -1, -2, and -3 is less than 50%, with some subgroups identified with a better prognosis than others (49,51). The latest report of the NWTS-5 focused on a group of patients enrolled on NWTS-5 and subsequently registered on the NWTSG protocol, "Treatment of Relapsed Patients" after recurrence. This protocol incorporates a more intense 24-week therapeutic regimen consisting of radiation therapy, vincristine, doxorubicin, etoposide, and cyclophosphamide (50). Compared to relapsed patients not registered on this protocol, improved 4-year relapse-free was 71 versus 53%, and overall survival rates of 82% versus 62% were observed.

The surgeon's role in recurrent Wilms' tumor lies in both prevention and therapy. The preventative role has to do with local recurrence. A thorough analysis of local recurrence in patients treated in NWTS-4 was completed by Shamberger et al. (52). They identified tumor spill (local or diffuse), age greater than 4 years, incomplete resection, and no lymph node staging at primary operation as factors associated with a high risk of local recurrence. The association of tumor spill and lack of lymph node staging to local recurrence has led to important staging and protocol alterations. As a result of this study, any tumor spillage, local or diffuse, is automatically staged as a stage III in order to receive more intensive postresection therapy to prevent recurrence and improve survival. In addition, resection of stage I tumors without lymph node sampling was associated with a 6-fold higher risk of recurrence compared to node-negative patients; therefore, all Wilms' tumor resections for localized or invasive disease must include a sampling of the hilar and/or paraaortic lymph nodes to recognize patients at risk for recurrence.

The therapeutic role has to do with postrelapse therapy. Green et al. studied recurrent Wilms' confined to the lung in NWTS -1, -2, and -3, and found no benefit of surgical resection over whole-lung irradiation and chemotherapy (53). The guidelines for resection of recurrent disease and metastasis has not been extensively studied; however, it is generally accepted that open biopsy to confirm relapse and attempted resection of intraabdominal recurrence to reduce tumor burden prior to radiation therapy and chemotherapy should be performed (54).

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Peripheral Neuroectodermal Tumors

Michael P. La Quaglia, MD

CONTENTS

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PRIMITIVE NEUROECTODERMAL TUMOR OF THE CHEST WALL

James Ewing described Ewing sarcoma in 1921 as an “endothelioma of bone” (1). He believed at the time that it arose from the osseous blood vessels and later realized that the tumor was more complex, describing it as an “endothelial myeloma” (2). Ewing is credited with differentiating this tumor from neuroblastoma and lymphoma. In 1979, Askin and Rosai described a “malignant small cell tumor of the thoracopulmonary region” in 20 children and adolescents with a median age of 14.5 years and with a female rather than male predominance, as seen in Ewing sarcoma of bone (3). These authors noted a predilection for local recurrence.

In the present era, most texts refer to the Ewing sarcoma family of tumors that include not only the highly malignant small round cell tumors of bone originally described by Ewing but also the chest wall tumors described by Askin, as well as extra-skeletal Ewing sarcoma, and peripheral primitive neuroectodermal tumors (PNET) (peripheral neuroepithelioma). A unifying feature of these tumors is the presence of a reciprocal translocation between chromosomes 11 and 22 (t11;22)(q24;q12) in approximately 85%. This is considered pathognomonic superseding other forms of pathologic analysis, like immunohistochemistry (4). Of great practical importance is that the surgeon must assure that an adequate amount of biopsy tissue is preserved fresh and sterile so that molecular genetic analysis can confirm the diagnosis. This chapter describes the diagnosis, treatment, reconstructive procedures, and late-treatment sequelae of PNET of the chest wall (Askin’s tumor, chest wall Ewing sarcoma). These chest wall tumors comprise 16% of all Ewing sarcomas, with most arising from ribs (63%), followed by scapula (25%), clavicle (9%), and sternum (3%) (5). Ewing sarcoma family of tumors generally metastasize to lung (10%) and bone (10%), or combinations of the two (6%). The rate of metastases for chest wall primary tumors is lower, and was commented upon in the initial paper by Askin (3).

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Diagnostic Evaluation and Initial Therapy

Patients with chest wall tumors may complain of a mass, bone pain referable to the ribs or spine, or radicular pain from infiltration of the intercostal nerves (6). Anterior masses may impinge on the pericardium, and be associated with chest pain in this region or a pericardial effusion. More posterior masses may invade not only the ribs and soft tissue but the bony spine as well. This could give rise to severe back pain. Diaphragmatic infiltration is possible and referred pain to the shoulder could ensue. Large tumors may also be associated with a significant pleural effusion and subsequent dyspnea, tachypnea, and/or orthopnea.

A chest wall mass or fullness is usually detectable on examination. Diminished breath sounds indicate a large pleural effusion, whereas heart tones may be decreased or a rub heard with a pericardial effusion. Because these tumors develop peripherally at the chest wall, the airway narrowing seen with large mediastinal masses like lymphoma is not an issue.

After the initial clinical examination, computerized tomography (CT) of the chest with and without intravenous contrast is recommended. The CT without contrast will delineate the mass, and identify an adjacent pleural effusion or cardiac enlargement, suggesting pericardial effusion. The spine can also be evaluated as tumors near the costovertebral junction can invade a vertebral body. A CT without contrast is also the best study to look for pulmonary metastases, which occur in less than 10% of cases. The addition of intravenous contrast will give further anatomic detail regarding the primary mass and its proximity to the aorta or other great vessels. These tumors arise from a single rib and expand and distort the bone, so that this rib can be identified as the epicenter of the tumor. This will have importance when resection is contemplated. The rest of the mass is extraosseous soft tissue expansion and this can be quite extensive. Magnetic resonance imaging (MRI) of the chest can also be used and it too provides a good deal of anatomic information regarding the primary tumor. Evaluation of pulmonary metastases is less accurate however, and the technique may require 30–60 minutes in the scanner for patients who may be having pain or dyspnea.

Since bony metastases occur in approximately 10% of patients with Ewing sarcoma/PNET, although this rate is probably less with chest wall tumors, a bone scan should also be done. There are recent reports that positron emission tomography with ¹⁸Fluorodeoxyglucose (PET) is more sensitive in identifying bony metastases from Ewing sarcoma/primitive ectodermal tumor (7). PET scans may also be useful in evaluating the response of primitive ectodermal tumors to neoadjuvant chemotherapy (8). Whole-body MRI may also be more sensitive in determining the number of osseous metastases than bone scan (9,10).

Once imaging studies are done, the patient will be ready for biopsy of the primary tumor to definitively establish the diagnosis. Needle core biopsies with or without CT guidance can give adequate amounts of tissue. It should be remembered, however, that much of the tumor before therapy can be necrotic and that an adequate amount of viable tissue must be obtained, not only for histology and immunohistochemistry but also for molecular genetics to document the (11;22) translocation. Molecular genetics alone requires two viable core biopsies. An open approach under general anesthesia may also be elected, and has the advantage of not only getting a larger biopsy but also allows placement of a vascular access device, and bone marrow aspiration and biopsy, which is part of the staging for these patients. Vascular access using an implanted double

lumen port is adequate and may be associated with a lower infection rate and improved quality of life compared to external devices. This initial biopsy should be planned so that the biopsy scar and underlying tissues can be incorporated into the incision for definitive resection and completely excised with the specimen. In addition, the pleural space should not be violated. Untreated chest wall PNETs can be very vascular, and may also have a large necrotic core caused by rapid tumor growth. Frozen sections should be obtained to verify that an adequate biopsy of viable tumor is being obtained. Packing with absorbable hemostatic agents may be required to control bleeding.

There is no formal staging system for Ewing sarcoma/PNET at present, except to describe it as localized or metastatic. Some have used the rhabdomyosarcoma grouping system: Group I—complete gross and microscopic resection; Group II—microscopic residual; and Group III—gross residual. Patients with metastases at diagnosis are given a separate category. This system is based on the results of resection rather than parameters measured at the time of resection and is generally considered unsatisfactory.

Initial therapy for chest wall PNET is systemic, multiagent chemotherapy. Combinations of vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide are used at present. Actinomycin D has not been used in recent studies. No one regimen has been accepted as optimal. Usually 4–5 cycles are administered prior to definitive local control. The rate of complete resection with negative margins for chest wall PNET was 50% when done prior to chemotherapy, and 77% after neoadjuvant chemotherapy ($p = 0.043$) (11).

Surgical Techniques for Excision After Biopsy and Chemotherapy

There is little disagreement that local control and cure from chest wall PNET requires complete resection. Available Pediatric Oncology Group-Childrens Cancer Group Intergroup data has shown that neoadjuvant chemotherapy increases the rate of complete microscopic resection of chest wall PNET, eliminating the need for adjuvant radiotherapy with subsequent pulmonary sequelae and the possibility of secondary tumors (11). The goal should be a complete microscopic resection after neoadjuvant chemotherapy.

Repeat imaging studies should be done prior to resection to document partial response in the primary tumor and make sure the unlikely development of intercurrent metastases has not taken place. At this point, the tumor may be 25–50% of its initial volume and much less vascular. This is illustrated in Fig. 1. The surgeon should know the primarily involved rib and what interspaces are affected as well as whether anterior or posterior. Because a negative microscopic margin is crucial, an adequate amount of tissue surrounding the gross extension of the primary tumor is required. If there is vertebral involvement, appropriate preoperative consultation with orthopedics and neurosurgery must be done so that an adequate contiguous resection is done at the primary resection. Anesthetic management includes placement of a double lumen endotracheal tube, or bronchial blocker, so that one-lung ventilation can be instituted. This will make it easier to excise any attached pulmonary segments in continuity with the primary chest wall tumor. Adequate intravenous access is necessary, as these resections often bleed heavily when the chest wall is cut. Positioning depends on the site of the primary, but the affected side is elevated so that the surgeon has access to all extensions of the tumor. Patients with anterior tumors may undergo less elevation, whereas those with very posterior tumors may be positioned in an almost semi-prone position.

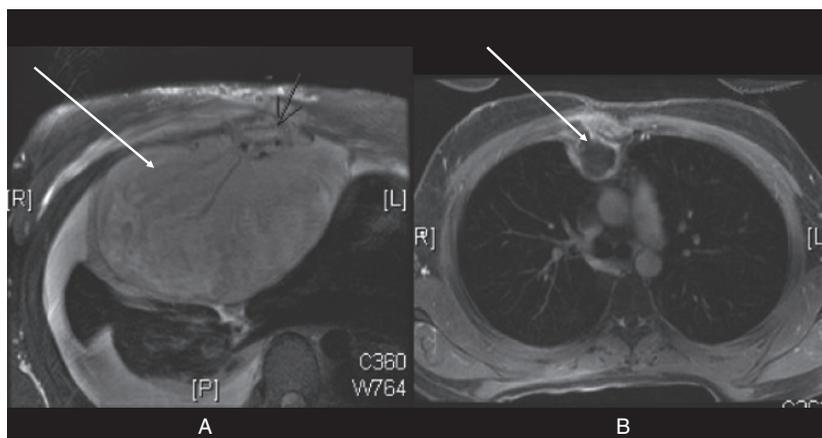


Fig. 1. (A) An MRI showing a large PNET of the upper sternum at diagnosis. A pleural effusion is evident posteriorly. (B) An extraordinary response in the same lesion after neoadjuvant chemotherapy. Intergroup data indicates that survival is not worsened by delaying resection to give chemotherapy, and this approach facilitates complete removal and avoids the need for chest wall irradiation.

Transverse, thoracotomy-like incisions are used and must incorporate the biopsy site and underlying tract, which is resected in continuity with the main specimen. This tract, which includes skin, fat, and the underlying fascia and muscle down to the chest wall tumor, must be handled carefully as it is easy to avulse it from the specimen. An elliptical extension around the tract works best. Once the tract is defined and the incision extended to the chest wall, the surgeon should refer to preoperative imaging studies and enter the pleural cavity anterior or posterior to the tumor and two interspaces away. Entry into the residual tumor will compromise the resection and require postoperative radiotherapy.

Once the pleural cavity is entered, the intrapleural extension of the mass is gently palpated and this usually is straight-forward. There are no generally agreed upon limits to resection of the primary tumor, but excision of an interspace and a rib segment around the gross extension of the tumor superiorly and inferiorly works well and will give a negative microscopic margin. The resection is then centered on the involved rib determined from imaging studies. One palpates the tumor, determines an interspace and rib segment above and below, and begins by cutting the unaffected ribs around the tumor to achieve a margin. Cutting the ribs above and below the affected rib gives mobility to the chest wall and allows visualization of the primary tumor. The intercostal vessels must be ligated when dividing the interspace. Titanium clips, stapling devices, bipolar electrocautery, and ultrasonic devices can be used.

The primarily involved rib, as noted previously, should be the center of the resection and of the residual mass. This is the origin of the tumor, and in theory the marrow cavity could be involved (Fig. 2). Although there is no definitive data, complete disarticulation of this rib at the costovertebral junction is a good surgical practice, especially for posterior tumors. The involved rib can also be separated from the costal cartilage anteriorly. The surgeon must not consider the size of the defect when removing the tumor. Huge defects can be reconstructed, and any positive margin will require the addition of radiotherapy with a resultant increase in morbidity. Similarly, segments of

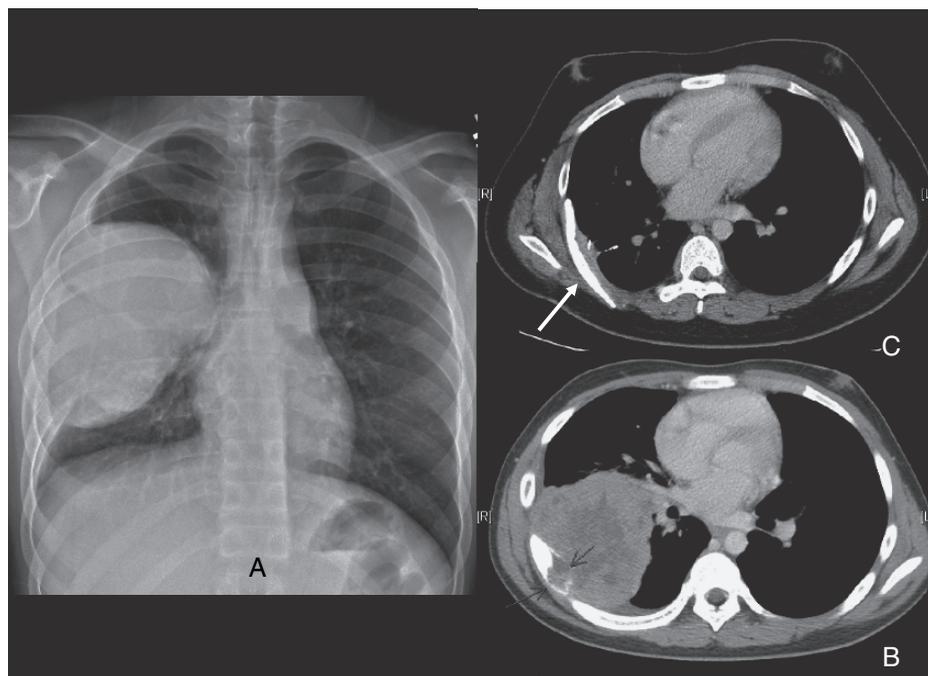


Fig. 2. (A) Plain film depicting a large chest wall PNET at diagnosis. (B) CT scan showing the same lesion with black arrows pointing to the rib of origin, which is greatly expanded and distorted. (C) The result after delayed resection and rigid reconstruction using a mesh-cement composite (white arrow).

lung, diaphragm, and pericardium should be removed in continuity with the specimen. Once the tumor has been removed, it should be carefully marked with multicolored sutures to orient the pathologist. Frozen sections should be obtained on the tumor margins intraoperatively, and positive margins should undergo further excision.

Techniques of Chest Wall Reconstruction

Adequate resection usually results in a significant chest wall defect and the possibility of flail chest. The terminology regarding reconstruction is not well defined, but can simply be divided into rigid and nonrigid techniques. Nonrigid approaches use prosthetic material to seal the pleural cavity, but do not provide chest wall stiffness. Examples include the use of expanded polytetrafluoroethylene (Gore-Tex; Gore Industries) or polypropylene mesh to close the defect. These approaches, in themselves, do not provide chest wall rigidity and will result in a flail chest and subsequent ventilator dependence when used for large defects. Usually, the chest wall will stabilize over time and patients can be weaned off the ventilator. Another disadvantage of nonrigid reconstruction is that chest contour is not preserved and a concave chest wall defect may be visible. Rigid reconstruction requires that in addition to a barrier to isolate the pleural cavity, unbending materials are incorporated. This will prevent a flail chest and restore contour to the chest wall. To accomplish this, polypropylene mesh is combined with methylmethacrylate cement to achieve the required rigidity. Two techniques are used: (1) A sandwich of mesh and cement is created and sutured into the defect; (2) Mesh is first sutured into the defect and then strengthened with rib-like struts of

cement created by hand. More recently, there has been interest in the use of expandable vertically oriented titanium ribs in reconstructing the chest wall, although reports focus on chest wall repair in conjunction with spinal surgery and scoliosis repair (12,13). Finally, latissimus dorsi rotation flaps or free tissue transfer has been used (14).

Rigid repair is generally preferable for large defects, because it prevents flail chest and preserves chest wall contour (15–17). The simplest approach to closure of a large chest wall defect is a rigid repair done with polypropylene mesh and methylmethacrylate cement. Such a composite repair is easy to do and can be made from materials readily available in the operating room. The sandwich technique requires that a template of the defect be taken using a gauze pad. The outline of the defect is then drawn out on a layer of polypropylene mesh. Methylmethacrylate cement is then layered onto the mesh and kept approximately 1 cm from the edges to allow a space for suture placement. A second layer of mesh is then used to “sandwich” the cement. The excess mesh is trimmed and the surgeon molds the composite into an outwardly convex shape to mimic the chest wall contour. The cement will get warm and set. This can be hastened by placing in warm saline. After the composite sandwich has cooled it is securely sutured into the defect with nonabsorbable material. This sometimes requires ingenuity in suture placement, and often bites are taken around the ribs for security. Tobramycin-containing cement is available, although data on infection rates with or without antibiotic-containing cement are not reported. An example of an anterior chest wall repair using a mesh-cement sandwich is illustrated in Fig. 3.

A simplification of the composite sandwich technique requires the creation of cement strips adherent to a single layer of mesh. In this variant, the mesh is securely sutured into the defect to close the pleural cavity. Two to three pairs of ribs (usually two) are selected, and the marrow cavities hollowed out for a few centimeters with a curette. Cement is then injected into the marrow cavities while a strip of cement is created by filling a .75-inch Penrose drain with cement as well. As the cement in the Penrose drain begins to harden, the ends are placed into proximity with the filled ribs and connection made by filling the intervening space with fresh cement. The drain can

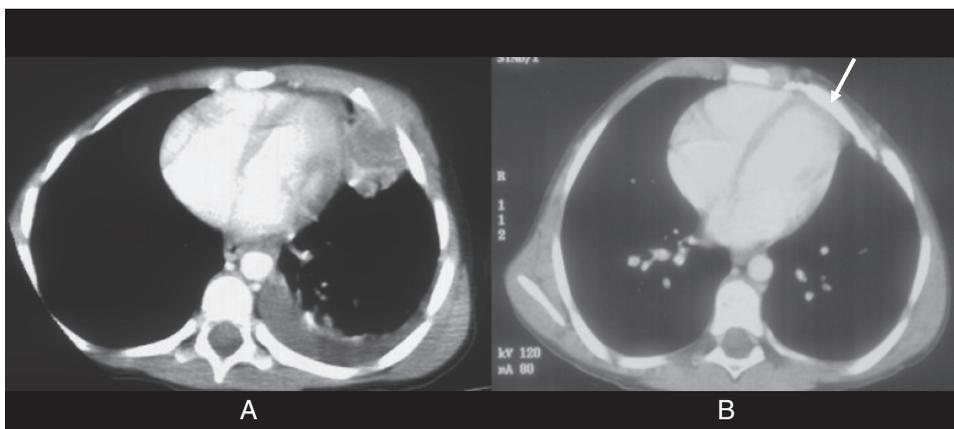


Fig. 3. (A) CT scan showing an anterior chest wall PNET that abutted the diaphragm and pericardium. Resection and rigid reconstruction using a mesh-cement sandwich was done after neoadjuvant chemotherapy at 5 years of age. (B) The status of the reconstruction 3 years later. The arrow points to the chest wall prosthesis.

easily be cut longitudinally and peeled away, taking care that it remains as one piece. Fresh cement is also placed between the cement ribs, which are kept convex outward to restore contour and the underlying mesh. As the cement hardens, it will adhere to the mesh and form an amazingly rigid and stable repair. The advantage of this technique is that smaller amounts of cement are used while rigidity and contour remain. It is also easier to place the mesh and it is more forgiving if the mesh size is not estimated correctly. Fig. 4 shows the result of this technique when applied to sternal resection after chemotherapy for the patient depicted in Fig. 1.

Complications and Late Effects

Early surgical complications of chest wall resection are the same for those of any major thoracotomy and include bleeding, pleural effusion (chylous, reactive), pneumonia, wound infections, and thrombophlebitis. Early mobilization and chest physiotherapy to promote lung expansion are mandatory. The major late effect of chest wall resection is scoliosis, with the convexity in the direction of the chest wall defect (18,19). Although there is no published data, this seems much more pronounced when nonrigid repairs are done. Bracing or formal scoliosis repair with spinal fusion may be required.

Recurrent Chest Wall PNET

Recurrence can be either local or in distant sites, or both, and is a very ominous prognostic finding. Local recurrence is associated with a positive surgical margin and may be heralded by increased local pain, or a mass appreciated clinically or on follow-up imaging studies. If a large mass is observed, the recurrence is obvious; smaller

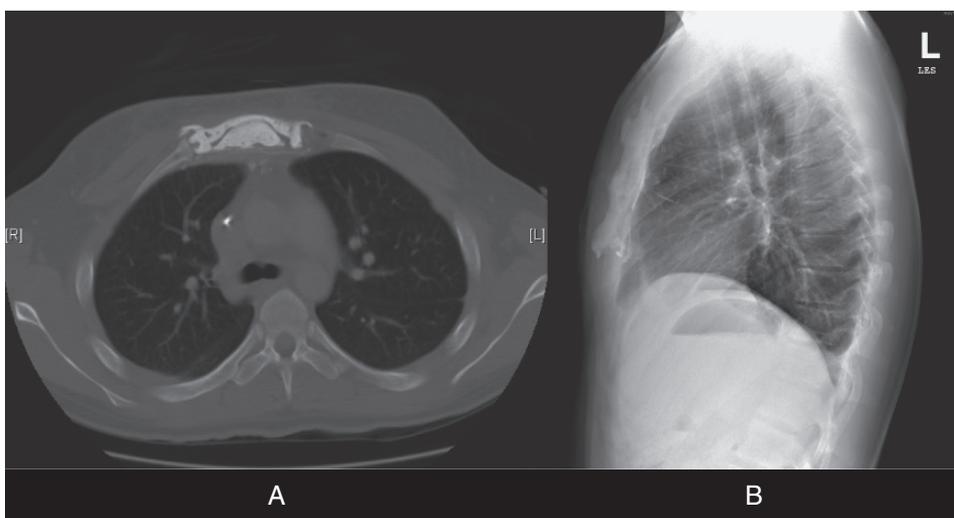


Fig. 4. The result after upper sternal resection of the patient whose initial and postchemotherapy MRI is shown in Fig. 1. First, a polypropylene mesh patch was sewn to ribs, intercostals, and periosteum to provide a backing and close the sternal and chest wall defect. Then, a “neosternum” of methylmethacrylate cement was created in a .75-inch Penrose drain and attached to cement inserted into each cut end of the sternum. These ends were first hollowed out with a curette for approximately 2 cm from the end. Further cement was added behind the sternum to secure it to the mesh and to create attachments of two ribs.

masses may require biopsy. Pleurocentesis of a recurrent effusion can show malignant cells and occasionally a more formal biopsy is required. Because the diagnosis and molecular genetics have already been established, a fine needle aspiration can be used and is less invasive.

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Osteosarcoma Metastases

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OVERVIEW OF OSTEOSARCOMA

Osteosarcoma is a high-grade malignant bone tumor arising from mesenchymal tissue. Approximately 400 children and adolescents are affected annually, making it the most common primary malignant bone tumor among children and adolescents less than 20, and the third most common malignancy among adolescents and young adults (exceeded only by leukemia and lymphoma) (1). Although osteosarcoma can afflict patients of all ages, the peak incidence is seen during the second decade of life, consistent with the time of most rapid bone growth (Fig. 1).

Basic Science

Osteosarcoma consists of malignant sarcomatous stroma producing tumor osteoid and bone. The cellular origin is likely a mesenchymal stem cell capable of differentiating toward fibrous tissue, cartilage, or bone (2). The most common pathologic variants of osteosarcoma are osteoblastic, chondroblastic, and fibroblastic, although numerous histologic subtypes exist (3). At the molecular level, osteosarcomas are characterized by an array of sequential and well-orchestrated genetic changes, including numerous tumor-suppressor genes and oncogenes (3). Two recessive oncogenes, p53 and RB, are thought to play major roles at the molecular level in the malignant transformation seen in osteosarcoma. Even still, the exact molecular pathogenesis remains unclear and many other genes seem to be involved (3).

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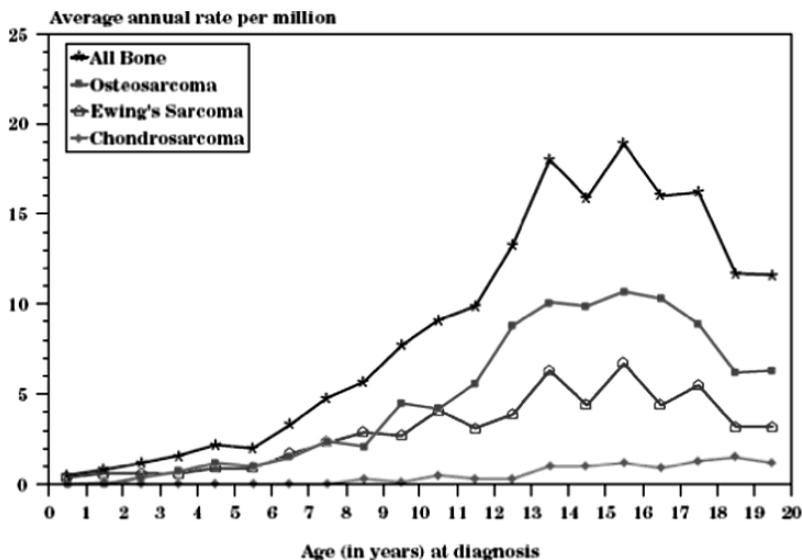


Fig. 1. Bone cancer age-specific incidence rates by histology (Osteosarcoma in solid box) (1).

PRIMARY DIAGNOSIS AND TREATMENT OF OSTEOSARCOMA

Diagnosis and Evaluation

Osteosarcoma is usually diagnosed after a plain radiograph reveals an osseous lesion extending into the soft tissues with destruction of cancellous and cortical bone. Initial evaluation includes a magnetic resonance imaging (MRI) of the affected extremity to assess the extent of local disease, a computed tomography (CT) scan of the chest to identify possible pulmonary metastases (most likely site of metastatic disease) (Fig. 2), and a radionuclide bone scan to identify osseous metastases (second most common site of disease). Chest CT scans are an important part of the initial evaluation, because 15–20% of patients present with pulmonary metastatic disease (4,5).

Treatment

Even when synchronous metastases are identified, management of the primary tumor is the initial priority. The treatment of the primary tumor is surgical, combined with neoadjuvant and adjuvant chemotherapy. Various chemotherapeutic agents, including cisplatin (administered intraarterially or intravenously), doxorubicin, high-dose methotrexate, cyclophosphamide, etoposide, and ifosfamide have been shown to be effective. Neoadjuvant chemotherapy is critical in the management of osteosarcoma for many reasons. Neoadjuvant chemotherapy provides early therapy against occult micrometastases, because up to 80% of patients presenting with osteosarcoma have occult disease (2,6). In addition, the primary tumor response to chemotherapy is an important prognostic factor. Percentage of tumor necrosis seen in the primary tumor explant accurately predicts prognosis. Patients with less than 99% necrosis in the primary osteosarcoma site have a much higher incidence of bony and pulmonary metastasis. Adjuvant chemotherapy, first introduced in the early 1970s, is largely responsible for improved overall survival. Randomized, clinical trials have confirmed the favorable impact of chemotherapy on the treatment of osteosarcoma (2,7,8).



Fig. 2. CT scan showing multiple pulmonary metastases.

Surgical Management

The goal of surgical management of the primary tumor is resection of all gross and microscopic disease, along with surrounding normal tissue to ensure complete removal. En bloc resection of the primary tumor is accomplished by an amputation or a limb salvage procedure. Limb salvage surgery has become the standard of care and can usually be accomplished, with overall survival and local recurrence rates similar to that of amputation (2,9). However, if complete excision is in doubt, secondary to tumor location, size, or extent, amputation is indicated to minimize the likelihood of local recurrence.

Patients with synchronous metastatic disease should undergo resection of the metastases after control of the primary tumor. The most common sites for metastatic disease are the lungs and bone, with pulmonary metastases being far more common. More than 80% of synchronous or metachronous metastases are located in the lung, and the lung is likely the only site of metastasis (10,11).

The fundamental principal guiding the management of metastatic pulmonary osteosarcoma is the importance of complete resection of all disease. Surgical approaches to pulmonary metastasis vary and are based on location, size, and extent of metastatic disease, as well as personal preference (Table 1). Because the majority of pulmonary metastases are located peripherally in the lung, they are amenable to wedge resection, allowing complete removal of tumor with maximal preservation of

Table 1
Surgical Approaches to Resection of Bilateral Pulmonary Metastatic Disease

<i>Surgical Approach</i>	<i>Advantages</i>	<i>Disadvantages</i>
Staged bilateral thoracotomies	<ul style="list-style-type: none"> • Optimal exposure • Short operative time • Best approach for repeat operations • Less pain • Minimal morbidity 	<ul style="list-style-type: none"> • Multiple trips to OR • Potential for long delay in between surgeries • Can delay chemotherapy
Simultaneous bilateral thoracotomies	<ul style="list-style-type: none"> • Optimal exposure • Single trip to OR • Best approach for repeat operations and bilateral disease • Rapid initiation of adjuvant chemotherapy 	<ul style="list-style-type: none"> • Lengthy operative time • More painful • Potential for postoperative pulmonary complications
Median sternotomy	<ul style="list-style-type: none"> • Single incision to expose both thoracic cavities • Single trip to OR • Good cosmesis 	<ul style="list-style-type: none"> • Poor exposure to posterior lesions/LLL • Increased morbidity with repeat operations • Intra-op cardiac compromise

OR, operating room; LLL, left lower lobe.

pulmonary parenchyma. Occasionally, segmentectomy, lobectomy, or pneumonectomy may be required to completely resect extensive, large, or centrally located disease.

Given the importance of complete resection, recent reports of thoracoscopic resection, and the current radiographic gold standard for pulmonary imaging, helical CT scanning, it is important to understand the limitations of detecting metastatic pulmonary disease. Although the speed and quality of resolution continue to improve, current CT scanning may miss small metastases (particularly those ≤ 5 mm in diameter) (12,13). The sensitivity of helical CT scanning is approximately 80% for all types of pulmonary metastases and approximately 60% for those less than 6 mm (13). Between 15 and 25% of metastases will not be identified on CT scan (13,14). A recent study found that that CT scanning underestimated the number of pathologically proven metastases in 35% of thoracotomies (14). Given these data, minimally invasive procedures should not be used in resection of pulmonary metastasis in osteosarcoma when the goal is complete resection.

Unilateral Metastases

The most common approach to the patient with osteosarcoma and unilateral metastases is unilateral thoracotomy with resection, followed by close surveillance (15). There is considerable evidence that a significant proportion of patients with presumed unilateral pulmonary disease diagnosed using current imaging techniques actually have occult bilateral disease (16). There is also ample evidence that complete resection of all disease (image detected and undetected) improves prognosis and overall survival (11,17,18). Kempf-Bielack et al. studied 576 patients with osteosarcoma relapse (469 included lung metastases) and found that patients with a second complete remission (defined as macroscopically complete surgery) had a 2-year and 5-year overall survival of 60% and 38%, respectively (10). Those without a second complete remission had

2- and 5-year overall survivals of 4% and 0%, respectively ($p < 0.0001$ for both). Given the importance of complete resection and the possibility of occult metastases after imaging, repeat unilateral thoracotomies are warranted.

Bilateral Metastases

There are multiple generally accepted strategies of surgically treating patients with bilateral lung involvement from osteosarcoma (15). The surgical options for these patients include staged bilateral thoracotomies, simultaneous bilateral thoracotomies, and median sternotomy. Probably the most common approach is staged thoracotomies, which provides optimal exposure of each hemithorax, avoids the longer operative and anesthetic time associated with simultaneous thoracotomies, and potentially minimizes postoperative pulmonary complications. A disadvantage of this approach is the potential for delay between the first and second thoracotomies owing to slow recovery or other postoperative complications. This delay could allow progression of disease, making eventual resection more difficult and postponing adjuvant therapy.

Simultaneous bilateral thoracotomies may be a safe and effective alternative to traditional staged bilateral thoracotomies. Rodeberg and colleagues recently compared staged and simultaneous bilateral thoracotomies in the pediatric population (19). They found that simultaneous bilateral thoracotomies lead to a shorter intensive care unit (ICU) stay, shorter hospital stay, decreased number of days with tube thoracostomy, and decreased time to initiation of adjuvant chemotherapy (19). They also found less postoperative complications, although not statistically significant, refuting the belief that the longer single operation, which exposed both thoracic cavities, would lead to more postoperative complications. They did note that patients with poor preoperative pulmonary function or patients who are anticipated to have marginal or inadequate postoperative pulmonary function, secondary to major resections, may benefit from the staged procedure. In these cases, pulmonary rehabilitation may improve residual lung function after initial resection (19).

Median sternotomy is an effective approach for many patients with bilateral osteosarcoma pulmonary metastases, and is the preferred approach for some surgeons (20–22). As with any operation, careful patient selection is important to ensure optimal outcome. Patient characteristics that make median sternotomy less ideal are: lung lesions that are located posteriorly; large or multiple metastases within the left lower lobe; and prior thoracic resections. The advantages include one operation for bilateral exposure and resection and probably decreased pain as compared to thoracotomy. However, because of the limited exposure of the posterior lung fields, the authors' preferred approach is bilateral thoracotomies when bilateral disease is present.

Follow-Up

Given the high rate of disease recurrence, close follow-up should be maintained for more than 1 decade after initial therapy completion. Patients should undergo a physical examination and radiographic (chest X-ray or CT scan) evaluation at 3-month intervals for 3 years, 6-month intervals through 5 years, and 8–12 month intervals through 10 years (23). Because of the risk of long-term toxicity and possibility of late relapses (10,24,25), follow-up should continue into the second decade of disease-free survival.

OUTCOMES AND PROGNOSIS AFTER TREATMENT

The first three-quarters of the twentieth century saw little change in the dismal prognosis facing patients with osteosarcoma. Over the subsequent three decades, however, greater understanding of the biology of the disease led to remarkable progress in the treatment of this malignant bone tumor. Multimodality therapeutic strategy (discussed previously) has been critical in the evolving treatment of this aggressive disease.

Until the early 1970s, the vast majority of patients with osteosarcoma developed pulmonary metastases and few patients survived beyond 1 year. The 5-year overall survival was less than 20% (2,26,27). Current therapeutic strategy, including early, multiagent chemotherapy, primary tumor resection via limb salvage procedures, and aggressive pulmonary metastatectomy, has significantly altered the prognosis of this previously lethal disease. Current overall survival for all patients at 2, 5, and 10 years is approximately 80%, 65%, and 60%, respectively (28). Only 10–15% of patients present with synchronous metastases (18), and only 30–40% of the remainder ever develop metastatic disease (24). Even those that develop metastatic disease have a 5-year survival of 30–40% (18,24).

The identification of demographic, tumor, and therapy factors that affect the prognosis of patients with osteosarcoma has helped focus therapeutic goals, clarified individual probability of survival, and stimulated basic science and clinical research. It is important to keep these prognostic factors in mind when treating and counseling patients with osteosarcoma. Some prognostic factors such as patient age, tumor site, tumor size, and synchronous metastases are assessable at diagnosis, whereas others such as chemotherapeutic response, extent of resection, or disease-free interval are assessable after primary treatment or during extended management.

Tumor response to preoperative chemotherapy has emerged as one of the most important prognostic factors in osteosarcoma (28,29). Patients that have a good response to neoadjuvant chemotherapy (>90% necrosis) have a 10-year overall survival up to 70–80% (28). Synchronous metastatic disease is a poor prognostic indicator (28). Patients with axial tumors fare worse than patients with extremity tumors (28). Extent

Table 2
10-Year Overall Survival by Prognostic Factor (28)

<i>Prognostic Factor</i>		<i>10-Year Overall Survival</i>	<i>p Value</i>
Age	<40 years	60%	0.012
	≥40 years	42%	
Tumor site	Extremity	62%	<0.0001
	Axial	29%	
Tumor size	<1/3 of bone	67%	<0.0001
	≥1/3 of bone	53%	
Synchronous metastases	No	64%	<0.0001
	Yes	27%	
Tumor necrosis	>90%	73%	<0.0001
	≤90%	47%	
Surgical remission	Complete	65%	<0.0001
	Incomplete	15%	

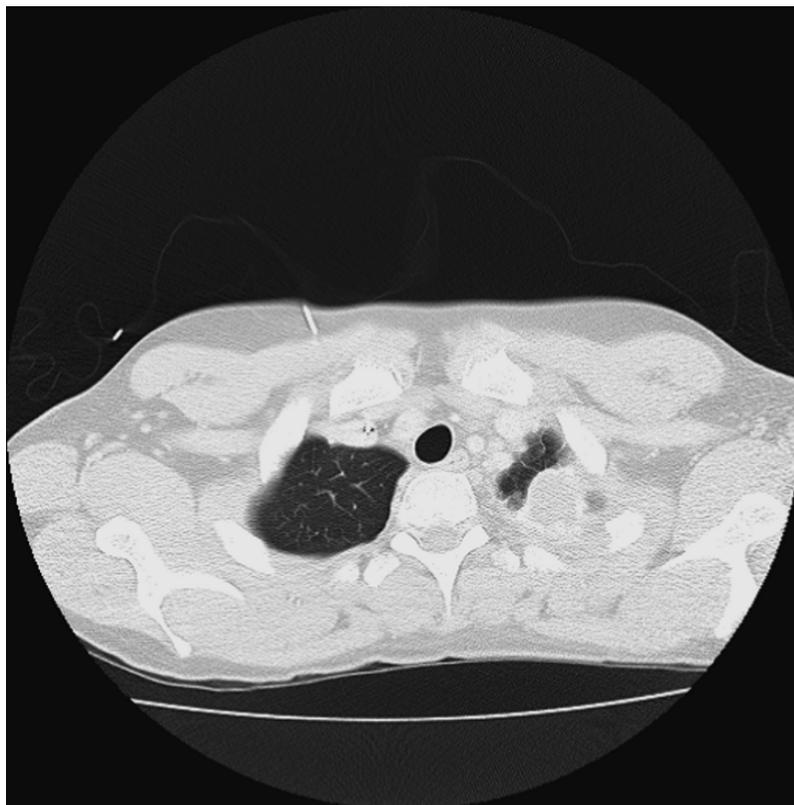


Fig. 3. CT scan of an apical metastasis that was only resectable after radiation therapy.

of surgery (macroscopically complete versus incomplete) is also a very important prognostic factor (28). Rate of overall survival at 10 years is highly variable, depending on certain key prognostic factors (*see* Table 2, extracted from the analysis of 1702 patients with osteosarcoma by Bielack et al. (28)).

A thorough understanding of the possible prognostic factors for patients undergoing pulmonary metastatectomy is particularly important for pediatric surgeons performing these resections. Most studies agree that primary tumor response to chemotherapy and disease-free interval (DFI) are the most significant prognostic indicators of long-term survival (28,30). Total tumor burden, as measured by number of metastases, location of metastases, and size of metastases, has been shown to be a significant prognostic factor (28). We recently found, however, that the measures of tumor burden (number, laterality, size) were not as significant as measures of tumor biology (response to chemotherapy and DFI) with respect to long-term survival (30). If all gross disease can be eliminated via operative resection, irrespective of the volume of disease, then long-term survival is possible through chemotherapeutic control of the microscopic disease. Often, unresectable disease is encountered (Fig. 3), and other approaches to pulmonary metastatic disease are warranted, such as radiation therapy, radiofrequency ablation, or high-dose chemotherapy.

Table 3
Sites of Relapse and Frequency of Site (10)

<i>Site</i>	<i>%</i>
Lung	81.4
Lung only	64.8
Lung + other site	16.6
Distant bones	15.6
Bones only	7.8
Bones + other site	7.8
Other ^a	9.4
Other only	2.1

^a includes lymph nodes, central nervous system, soft tissues, liver, kidney, and others

RECURRENT DISEASE

Between one-third and one-half of patients will suffer a relapse (metastatic or local) after initial multimodality management of the primary disease (10). A study of 1702 patients with osteosarcoma by Kempf-Bielack et al. included 576 relapsed patients. Recurrence was identified in the lung in 81% of the patients, with other sites (metastatic or local) much less common (Table 3). Median time from diagnosis to first relapse is 18 months to 2 years (10,24). Patients with certain risk factors, including axial site, large primary tumor, and synchronous metastases can be identified as more likely to develop osteosarcoma recurrence (11,28). After completion of treatment, initial relapse patients have an overall survival between 15–20% at 10 years (10,11).

Relapse Management

The multidisciplinary approach is maintained in the management of recurrent disease. Complete surgical resection is still key to prolonging life and maximizing the opportunity for long-term survival. Timely surgical resection of all gross pulmonary metastasis is the mainstay of therapy. There is no consensus regarding the optimal particular agent(s) or the overall effectiveness of the chemotherapeutic approach to recurrent disease. Several studies have shown a positive correlation between “second-line” chemotherapy (alternative agents to those used during the initial treatment) and survival (10,24,31). Radiotherapy is reserved for patients unable or unwilling to undergo surgery, patients with pleural-based disease, patients found to have positive or close margins after resection (10), or patients with unresectable disease.

Summary: Surgical Therapy for Recurrent Disease

Surgical remission is critical in the management of these challenging patients. Failure to resect all visible disease, irrespective of total tumor burden has been shown to be a strong negative prognostic factor, and long-term survival is only possible after complete resection (10,11,32,33). Studies support surgical resection and close follow-up (without chemotherapy) for patients that develop a solitary lung nodule after a lengthy DFI (>2 years) (17,24,33).

The specific surgical approach can be via thoracotomy or via median sternotomy (as discussed previously). With repeat resection, additional considerations increase the

complexity of the operation. Repeat thoracotomy (after a previous thoracotomy or median sternotomy) is probably the most common procedure performed. The dissection poses minimal risk and the exposure allows maximal exploration of the entire lung so that all metastatic disease can be identified and resected. After satisfactory single-lung ventilation, the collapsed lung should be very thoroughly palpated by the operating surgeon, because a preoperative CT scan with 5-mm cuts can miss 15–25% of lesions. Numerous repeated thoracotomies can be performed relatively safely. At the authors' institution, we have performed more than five sequential thoracotomies (on the same side) for several children with repeated recurrent disease and have prolonged their survival. However, recurrent thoracotomies are often complicated by increased blood loss and increased operative time. The prospect of repeating a thoracotomy for the fourth or fifth time is not itself prohibitive, but patients should undergo preoperative pulmonary function tests and echocardiography to determine the feasibility of surgical resection when a repeat thoracotomy is being considered.

Although median sternotomy offers several advantages, it is not recommended for recurrent resection (irrespective of the initial operation). Even though it exposes the thoracic cavities bilaterally, it offers poor exposure to posterior pulmonary segments. Adhesions from previous thoracic operations can make entry via the sternum a difficult task, and the approach has a higher morbidity with repeated surgeries.

Wedge resection is the preferred surgical procedure, even in reoperative thoracotomies, because complete resection can be combined with preservation of maximal respiratory capacity. As discussed previously, lobectomy and pneumonectomy are usually reserved for very extensive disease or disease located very centrally (i.e., at the hilum) (32).

Given the importance of complete surgical resection (10,11) and the possibility of long-term, disease-free survival, even after multiple recurrences (10,11,17,30,34), aggressive pulmonary resection and repeat pulmonary resection should be attempted if all gross disease is resectable and if adequate postoperative pulmonary reserve can be anticipated (30,32). Our recent research revealed that patients with a good response to chemotherapy and a DFI greater than 1 year had a 50–60% 5-year survival after aggressive, complete metastasectomy (30). However, even those with a suboptimal response to chemotherapy (<98% necrosis) and a short DFI had a 12% 5-year survival after surgical remission. Briccoli and coworkers found that 5-year event-free actuarial survival from first metastectomy (38%) to be similar to the 5-year event-free actuarial survival after second metastectomy (32%) (32). Even after the fourth and fifth relapses, patients undergoing resection were experiencing long-term, disease-free survival.

PROGNOSIS AFTER RECURRENCE/MANAGEMENT

The prognosis after osteosarcoma disease recurrence and subsequent management is not as promising as the prognosis after initial management. Ensuing relapses tend to occur after a shorter disease-free interval (10,32). Only one-quarter of patients are expected to survive 5 years after recurrence and 10-year survival is approximately 20% (10,35). Among the prognostic factors associated with survival after recurrence, the only significant factor from the *initial* disease presentation/management appears to be primary tumor response to chemotherapy (10,30). Other prognostic factors associated with prolonged survival include late relapse (i.e., DFI or recurrence-free interval [RFI]), solitary lesions, unilateral lesions, and absence of pleural involvement (10,33).

Various studies defined late relapse at different time points, including 18 months (10), 24 months (33), 36 months (31), although they all found a correlation between long RFI and long-term survival.

A recent single institution study at MD Anderson Cancer Center found that the number of lesions, size of lesions, and location of lesions were factors that were not as important as the primary tumor response to chemotherapy or the DFI (30). These results led to the theory that tumor burden was not as critical as tumor biology with respect to long-term survival.

FUTURE/ONGOING RESEARCH

Although the prognosis for patients with osteosarcoma has improved significantly over the last three decades, a disturbing trend is the more recent plateau in survival (36). Although the advances in chemotherapeutic and surgical strategy developed over the last 30 years produced enormous strides in the management of osteosarcoma, innovative discoveries must continue at the molecular, cellular, and clinical levels.

The basic science, translational, and clinical research discoveries continue to broaden our understanding and therapeutic potential for this disease. As we learn more about the biology of osteosarcoma, genetic markers of the host and/or the tumor could predict disease progression and aid the selection and development of effective therapy (11,37). This type of “molecular risk-based treatment” could help focus therapy so that high-risk patients receive the most aggressive therapy and lower risk patients are perhaps spared the side effects of this more aggressive treatment.

It may also be possible to subgroup patients at the cellular level, based on the detection of micrometastatic osteosarcoma cells (38). Immunomagnetic detection assays have been used to assess the presence of tumor cells in bone marrow and peripheral blood. Such an assay has the clinical potential to evaluate the effectiveness of a particular chemotherapeutic approach or to group patients based on cellular risk.

In addition, multicenter trials and clinical studies are leading to larger cohorts and more powerful conclusions. New chemotherapeutic and immunotherapeutic agents, used alone or in combination, are being critically assessed and more standardized recommendations are being generated. More patients are being enrolled in well-designed clinical trials, which yield valuable data and offer patients and their families some hope of prolonged survival.

Osteosarcoma remains a challenging disease to manage and treat, particularly when disease recurrences lead to multiple surgeries and consecutive rounds of intense medical treatment. The current state of the multimodality approach leads to long-term survival for many patients, even after metastatic recurrences.

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Neurofibromatosis

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INTRODUCTION

Neurofibromatosis (NF) a common genetic disorder with an autosomal dominant pattern of inheritance with variable penetrance. NF is a complex disease with many different systemic manifestations and complications. Based on symptoms and the region of chromosomal abnormality, NF has been classified into two types: neurofibromatosis I (NF 1) and neurofibromatosis II (NF 2). NF 1, or von Recklinghausen's disease, has been localized to the long arm of chromosome 17 and occurs in nearly one in 3000 live births (1,2). Because NF 1 has many different manifestations, the National Institutes of Health (NIH) developed a standardized method to diagnose NF 1, which includes two of the following criteria: six or more café-au-lait spots; two or more neurofibromas or one plexiform neurofibroma; axillary or inguinal freckling; optic gliomas; osseous lesions; lesions of the iris known as Lisch's nodules; or a first-degree relative with NF 1 (3). In contrast, NF 2 has been localized to the long arm of chromosome 22 and has a much lower incidence of only one in 55,000 live births (4). It has been defined as either bilateral acoustic neuromas visualized with appropriate imaging or a first-degree relative with NF 2, in addition to either a unilateral eighth nerve palsy or two other lesions: neurofibroma, meningioma, glioma, schwannoma, or juvenile posterior subcapsular lenticular opacities (3). Despite the autosomal dominant pattern of inheritance, it is important to note that nearly 50% of all cases of NF 1 and a greater proportion of NF 2 cases are sporadic mutations (2). NF is a progressive, systemic disease that can affect many different areas of the body. Depending on the location,

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size, and growth of lesions, patients may require multiple trips to the operating room, which may vary from simple excision of cutaneous lesions to aneurysm repair to large resections of malignant peripheral nerve sheath tumors (MPNST).

CUTANEOUS MANIFESTATIONS

The most common manifestations of NF are cutaneous neurofibromas. In the dermis, neurofibromas often extend outward and become pedunculated, but may not be visible when in the subcutaneous or deeper tissues. During periods of high hormonal activity such as puberty or pregnancy, neurofibromas may appear, grow, and increase in number (2,5,6). The lesions may be tender to palpation or trauma, but they are rarely painful. These lesions are described based on their distribution as discrete, segmental, or plexiform (2,7). Discrete neurofibromas are well-defined, rubbery masses with distinct margins that develop at any point along peripheral nerves. These lesions range in size from a few millimeters to several centimeters and can be found at multiple sites throughout the body. Segmental NF is defined as café-au-lait macules or neurofibromas in a single, unilateral segment of the body, with no crossing of the midline, no familial history, and no systemic involvement (7). In general, segmental NF arises in a dermatomal distribution, typically in the cervical, followed by the thoracic, lumbar, and sacral dermatomes (8). With discrete and segmental NF, the morbidity is usually cosmetic, because of the number visibly apparent lesions. However, when neurofibromas extend to major nerve roots, radiculopathy and weakness secondary to compression is possible (2,9). In both cases, malignant transformation is rare, and surgical management is typically reserved for resection for cosmesis, functional reasons, or pain control (10).

Plexiform neurofibromas do not refer to the involvement of a specific plexus of nerves, but rather have a network-like growth of a neurofibroma involving multiple fascicles of a nerve and may include multiple branches of a large nerve, leading to a diffuse mass of thickened nerves (11). Plexiform neurofibromas can be debilitating in patients with NF, and are one of the most common sources of morbidity. Plexiform neurofibromas can be found in nearly 30% of patients with NF by physical examination alone (6). However, to determine the true incidence of plexiform neurofibromas in NF patients, Tonsgard et al. conducted thorough radiological evaluation of 91 patients with NF. They determined that thoracic lesions were identified in 20% and abdominal lesions in 44% of patients, which were largely asymptomatic (12). Plexiform neurofibromas typically have a “bag of worms” feel on palpation and can grow very large. They can extend for variable distances along a nerve or nerve plexus and can lead to hypertrophy of the tissue around the lesion (10,11). In addition, the overlying skin may also be thickened or discolored (11). Plexiform neurofibromas tend to grow in two distinct periods: early childhood and periods of high hormonal activity, such as puberty or pregnancy (10,11,13). Plexiform neurofibromas may be tender to palpation but are rarely painful. In addition, plexiform neurofibromas may degenerate into MPNSTs, so rapid growth or constant pain should alert the clinician to the possibility of malignancy (10,11). Facial plexiform neurofibromas and pruritus were associated with increased mortality in children (14).

Although no uniform guidelines exist, surgical excision is the mainstay of management for plexiform neurofibromas. The decision to excise lesions is focused on both the timing and the extent of the operation; however, some controversy exists.

Some believe that it may be difficult to justify resection of a small, asymptomatic plexiform neurofibroma on the assumption that it may grow at a later date. In addition, others state that complete resection of the lesion is difficult with recurrence likely (11). Others have proposed that early resection of lesions may prevent functional damage at a later date. Jackson et al. have offered that early resection of plexiform neurofibromas may lead to better long-term functional outcome (15). In addition, a retrospective review of 121 patients with plexiform neurofibromatosis showed that overall tumor progression after surgical excision was 46% and that age less than 10 years at operation and incomplete resection were independent prognostic for tumor progression. It is interesting to note that the authors did show that more extensive resections did slow tumor progression (16). Although early resection may avoid functional or cosmetic impairments, it is important to consider the risk of recurrence of plexiform neurofibromas when considering surgical excision.

GASTROINTESTINAL (GI) MANIFESTATIONS

Although GI manifestations of NF 1 are well described, the true incidence and proportion that become symptomatic are unknown. Based on retrospective reviews of patients with NF 1, 10–25% of patients are estimated to have GI tumors, with less than 5% having symptoms from these tumors (6,17,18). There are reports of a subtype of NF that has isolated GI involvement without other systemic features of NF (19,20). However, it is unclear whether this represents a distinct variant of NF or incomplete penetrance of the NF 1 gene.

GI manifestations of NF are divided into three groups: hyperplasia of the gut neural tissue, stromal tumors, and duodenal or periampullary endocrine tumors (21). The hyperplastic lesions are characterized by increase in Meissner's and Auerbach's plexuses along the GI tract. Based on its patchy distribution, the lesions may appear as discrete or plexiform neurofibromas. These lesions typically cause constipation, which is found in nearly 10% of all patients with NF and may be confused for Hirschsprung's disease (22,23). If conservative management fails to control symptoms, colectomy may be necessary to relieve the symptoms (10).

GI stromal tumors (GIST) are the most common nonepithelial tumor of the GI tract (24). In patients with NF 1, there is a 45-fold increase in the incidence of GIST, suggesting that inactivation of the NF 1 tumor suppressor pathway is also involved in the pathogenesis of GIST (25). This association with NF 1 and GIST has prompted examination into the mutations seen in GIST tumors. Tazakawa et al. have recently demonstrated that 92% of sporadic GIST tumors have mutations of the gene for the type III tyrosine kinase receptor (KIT). In contrast, only 8% of NF 1-associated GIST tumors have the mutation of the KIT gene (24). These lesions are typically found in the stomach and jejunum, and present with bleeding, obstruction, perforation, intussusception, or volvulus (10,17). Treatment of GISTs involves resection with grossly negative margins and an intact pseudocapsule. Because lymph node involvement is rare with GISTs, lymph node dissection is not necessary. Typically, adjuvant chemotherapy is not necessary; however, clinical trials are examining the efficacy of these therapies.

An association has been observed between NF and duodenal or periampullary carcinoid tumors (10,26–28). These tumors are usually located at or near the ampulla of Vater and are small, tan nodules. On histologic examination, these lesions are

composed of glandular elements that involve surrounding tissue planes and are easily confused with adenocarcinoma (21). However, it is the presence of psammoma bodies that helps differentiate carcinoid tumors from adenocarcinoma (10,21).

VASCULAR MANIFESTATIONS

Hypertension is frequently seen in children with NF, and is typically caused by renovascular abnormalities or aortic coarctations (29–32). Renal artery stenosis is typically the leading cause of secondary hypertension in patients with NF, with as many as 25% also having associated coarctation of the abdominal aorta. As a result, it is typically amenable to surgical correction (33,34). However, these vasculopathies are progressive and often require repeat angioplasty, stenting, or surgical reconstruction. Restenosis is very common after angioplasty and can be especially challenging in young patients, in whom grafts or stents may not allow for sufficient growth of the vessels (35,36).

NF vasculopathy was first described in 1945 by Reubi, was subsequently revised by Salyer and Salyer in 1974, and is now the standard (10,37). They described pure intimal type, with focal nodules of spindle cells and media thinning; advanced intimal type, with eccentric fibrous thickening in the intima and fibrosis of the media; intimal-aneurysmal type, with marked fibrous thickening of the intima and elastic fragmentation in the media; and nodular/epithelial type, with cluster of spindle cells throughout all three layers. In addition to this description, Greene and coworkers have classified two primary types of vascular lesions associated with NF based on vessel diameter. Large vessels are affected by perivascular neurofibromas or ganglioneuromas that degenerate the adjacent vascular walls, and small vessels are composed of nodules of smooth muscle representing mesodermal dysplasia (38).

Peripheral vascular lesions are more common than central lesions in NF and may lead to significant functional problems. One young girl, in whom femoral and popliteal aneurysm reconstruction had failed, ultimately required bilateral below-the-knee amputations (39). Arterial aneurysms are frequently seen in the NF population and may also be the result of trauma from vascular interventions (35). Another young girl with lower limb hypoplasia and gangrene had chronic ischemia not amenable to reconstruction and required an above-the-knee amputation (40). Aneurysms have also been reported in combination with renal artery stenosis and hypertension, underscoring the importance of thorough examination of patients with NF for vascular abnormalities (41).

The vascular changes associated with NF often produce challenging hemostasis problems when resection or debulking of complex lesions is attempted. Arteriovenous malformations are common in large plexiform neurofibromas, often with multiple “feeding” vessels present, and aneurysms may actually be complex, multilobulated masses. Massive hemorrhage has been noted during many resections, frequently requiring blood transfusions of several units (42–45). Based on their successful experiences, some authors routinely request angiography for patients with plexiform neurofibromas, and if indicated, preoperative embolization (43). Some choose to perform staged excisions with hypotensive anesthesia, preliminary sutures around the lesion, ligation of all feeding vessels, and autotransfusion as necessary, though significant blood loss is still possible (42). Although not routinely required, these modalities should be considered during preoperative evaluation.

THORACIC MANIFESTATIONS

The association of cardiovascular malformations (CVM) and NF has been implied for years. Numerous case reports have described patients with NF and congenital heart defects (2,46,47). A recent sonographic study demonstrated a significant increase in cardiac abnormalities in children with NF 1 versus other adolescents (27% versus 3.6%). In addition, there was a significant decrease in the isovolemic relaxation time (τ) in patients with NF 1, suggesting that NF changes the intrinsic properties of the heart as well (48). It has been noted that pulmonic stenosis does make up a larger proportion of the CVMs of patients with NF than those of patients without NF. Based on a review of the National Neurofibromatosis Foundation International Database, 25 of the 54 CVMs documented were pulmonic stenosis (46). The association of this defect with features of NF 1 has been well described in both Watson's syndrome and NF 1-Noonan's syndrome; but in the study mentioned previously, only four of the 54 patients were diagnosed with either of these conditions (2,46). For these reasons, it may be reasonable to assume that there is a genetic predisposition for pulmonic stenosis in patients with NF and CVMs.

Though rare, NF can involve the heart, with plexiform neurofibromas invading this and other surrounding structures. A 10-year-old girl in whom a mediastinal plexiform neurofibroma was causing a mass effect on the left mainstream bronchus and both atria also had echocardiographic evidence of right atrial and arteriovenous septal invasion (49). In addition, a 29-year-old man was reported with clinical biventricular failure; his open biopsy confirmed plexiform neurofibroma encasing both atria, resulting in abnormal function. Review of this patient's history revealed that partial resection of a mediastinal mass had been attempted 15 years earlier but was limited at that time secondary to extension into the interatrial septa (50).

Intrathoracic noncardiac tumors are the most common manifestation of NF in the chest. One pediatric-based review of 260 patients over 10 years reported an incidence of thoracic tumors of 3.5%, but this study was severely limited. The true incidence of thoracic tumors is not known, but is probably closer to 10% (13). Six of the nine children with tumors were asymptomatic, so the long-term implications of minor or incidental tumors are not known. Thoracic tumors can be very aggressive, and have major functional implications. These tumors typically extend from spinal, phrenic, vagus, or intercostal nerves and progressively invest surrounding structures. They can produce dyspnea, dysphagia, invasion of the lung tissue, erosion of the ribs, arrhythmias, or diaphragm paralysis (13,51–55). In addition, tumors that develop from the structures of the chest wall can produce restrictive pulmonary disease, if they enlarge sufficiently, and pose complex reconstructive problems (56).

MPNSTS AND PHEOCHROMOCYTOMAS

Long-term studies have revealed that patients with NF have a two- to 5-fold increase in relative risk of neoplasm than the general population (57). As a result, routine examination and vigilance is necessary throughout the patient's life in order to detect malignancies early and improve patient outcome. Although patients with NF have been known to develop numerous different malignancies, the most common extracranial malignancies are Malignant Peripheral Nerve Sheath Tumors (MPNSTs).

MPNSTs have been classified by the World Health Organization to include malignant schwannoma, neurogenic sarcoma, neurofibrosarcoma, and malignant neurilemmoma

(58). Overall, MPNST is a rare form of cancer in the general population, representing only 10% of all soft tissue sarcomas with an incidence of only 0.0001% (58,59). However, more than 50% of these tumors occur in patients with NF, with an estimated lifetime risk of approximately 4%, or a 4600-fold increase in risk compared to the general population (58,60). In NF, these tumors typically arise from a portion of a plexiform neurofibroma and are very aggressive, frequently recurring locally or distantly, with hematogenous spread as the route of metastasis. Doorn et al. examined MPNST occurring in patients with and without NF and found that the median overall survival was only 24 months (61). Other authors have found similar results with survival ranging from 8–44 months, indicating the truly aggressive nature of the disease (59,62–64). Although some studies have suggested that the presence of NF has no effect on the overall survival of patients with MPNSTs, the largest study of its kind from the Italian and German Soft Tissue Sarcoma Cooperative Group has suggested that MPNSTs in patients with NF are more aggressive, with a significantly lower 5-year overall survival (32% versus 55%) and a significantly lower progression-free survival (19% versus 42%) than their counterparts without NF (59,61,64,65). These findings support prior findings from other smaller studies (58,62,66).

Prognosis for patients with MPNST is dependent on several factors. Size of tumors greater than 5 cm, local invasion, presence of NF, and higher histologic grade have been identified as factors indicating a poor prognosis (58,59,67,68). Improved prognosis is associated with duration of symptoms less than 6 months and clear surgical margins (64).

Overall, the natural history of MPNST is such that the majority of patients will relapse. A retrospective review of 57 years of experience with MPNST in children at MD Anderson Cancer Center revealed 38 documented cases (60). A group of 25 of the 32 patients who initially had a complete response relapsed after therapy. Of these, 14 occurred as distant disease, with the lung as the most common location (86%). Of the 22 children with negative surgical margins, survival was 33% as compared with 23% in those with positive margins. Gender, site, margins, adjuvant therapy, or presence of NF did not notably affect long-term outcome (60). In contrast, a recent study has shown that primary site, age, and presence of NF are prognostic factors in addition to size and invasiveness (65). Because of the rarity of MPNSTs in children, it may be difficult to resolve the controversy in the literature. However, the current data do demonstrate the aggressive nature and poor overall prognosis of MPNSTs.

The most important goal for successful management of MPNST is curative resection with negative margins. When attainable, this provides the best hope for long-term survival (58–61,63,64,68). Over the past several decades, a trend toward limb-sparing resection over amputation has developed, in part because of earlier diagnosis of smaller tumors and the increased use of perioperative radiation therapy (58,59,69,70). A prospective randomized trial has shown a decrease in local recurrence when adjuvant brachytherapy radiation was used in conjunction with limb-sparing operation, provided there was no evidence of gross residual disease (59). Although no significant difference in overall survival was found, it was felt that more long-term data were needed before this conclusion could be made. Others have also found an advantage in prolonged disease-free survival when radiation was used postoperatively (58). The use of adjuvant chemotherapy continues to be controversial. Given the rare nature of the disease and the changes in chemotherapy used over the years, it has been difficult, if not impossible, to conduct prospective randomized trials or systematic retrospective reviews. Although

there is no consensus, the largest study to date has recommended that ifosfamide and doxorubicin be given to patients with unresectable tumors greater than 5 cm (71,72). In addition, new medications that target the epidermal growth factor receptor, which is overexpressed in NF 1, may provide future modalities for treatment of MPNSTs (65).

In addition, patients with NF are at increased risk for developing pheochromocytomas. Because these tumors are derived from neural crest cells in the adrenal medulla or other portions of the sympathetic nervous system, the cause of the association with NF is clear. In several reports of patients with NF, the incidence is 0.1–6%, as compared with 0.4 per 100,000 in the general population (6,57,73,74). These tumors are notable for their ability to secrete norepinephrine or other catecholamines, which results in intermittent symptoms that mimic a sympathetic response: tachycardia, palpitations, paroxysmal hypertension, headaches, perspiration, or unexplained anxiety (10). Though rare in children, pheochromocytomas typically present in female patients with NF in their early forties (75). However, with the strong association between pheochromocytomas and NF, it is important to consider in children with NF and hypertension.

Identification and localization of pheochromocytomas in patients with NF are the same as those of a patient with similar symptoms who does not have NF. Approximately 85–95% of pheochromocytomas are located in the medulla of either adrenal gland, with 10% located bilaterally and a 10% incidence of malignancy (2). The evaluation for diagnosis of pheochromocytoma usually includes a 24-hour urine collection for quantification of vanillylmandelic acid and catecholamines, although this is not 100% sensitive because of the episodic nature of the tumor's secretion. Localization may be obtained through various imaging techniques, focused initially at the adrenal glands. Magnetic resonance imaging (MRI), which is recognized as the most sensitive method, has been shown to have up to 100% detection; computed tomography (CT) scans (94%) and metaiodobenzylguanidine (MIBG) nuclear scans (88%) are less sensitive (75,76).

Preoperative preparation for patients with NF is essential to minimize the risks associated with resection of pheochromocytomas. Well-known causes of adrenergic crisis associated with pheochromocytomas include anesthesia, surgery, labor, and trauma, which can precipitate cerebral vascular accidents, cardiac arrest, arrhythmias, or shock (77). Management typically begins with alpha blockade to reduce peripheral vasoconstriction followed by beta blockade to control tachycardia. Patients with pheochromocytoma often exhibit plasma volume depletion, decreased hematocrit, or both once the alpha blockade is established, which has led many authors to also recommend preoperative volume expansion (77,78). Both the surgeon and anesthesiologist should be aware that once the tumor is removed, the patient is highly susceptible to hypotension and shock and should be treated accordingly (79).

Surgical resection is the mainstay of therapy for all patients with pheochromocytomas, and as progress in techniques is made, the approach is changing. With adequate imaging for tumor localization, the need to examine the contralateral adrenal gland or to search blindly for ectopic foci and metastases does not justify the risk of significant injury to the patient in the majority of cases (10). Typically, a transabdominal approach is employed to approach the tumor. Although laparoscopic adrenalectomy has been studied in adults (80–82), there is only anecdotal evidence at this point to recommend its use in children (81,83). Though further studies will need to be done to verify its benefits in children, laparoscopic resection does show promise as a less morbid treatment modality. In patients with NF, care must be taken in considering the

individual patient's associated manifestations when planning the approach. Previous resections of intraabdominal disease or overlying cutaneous lesions may preclude certain techniques, so a thorough knowledge of the patient's past surgical history is a requirement.

CONCLUSION

NF is a complex, systematic disease with a wide variety of sequelae and complications. Because of its systemic sequelae, a surgeon may be called on to operate multiple times on the same patient. These operations may range from simple excisions of cutaneous neurofibromas for cosmesis to resections of a life-threatening malignancy. As a result, it is important that the surgeon approach these patients with a thorough understanding of the progressive and ubiquitous nature of this disease in order to adequately manage the disease over the life of the patient.

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Tumors of the Liver

Rebecka L. Meyers, MD

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BENIGN LIVER TUMORS

The differential diagnosis of benign liver tumors in children varies tremendously with age. In infancy, the most common benign hepatic tumor is hepatic hemangioma. Infantile hepatic teratoma is exceedingly rare. Commonly seen benign tumors of the toddler age group are mesenchymal hamartoma and focal nodular hyperplasia. Hepatic adenoma is almost exclusively a disease of older children and adolescents. There are several distinguishing characteristics of these benign tumors on radiographic evaluation; however, imaging techniques such as ultrasound scan, computed tomography (CT) angiography, and magnetic resonance imaging (MRI) are not always reliable in differentiating benign from malignant tumors.

MALIGNANT LIVER TUMORS

The differential diagnosis of malignant liver tumors in children also varies with age. In infants and children, most malignant liver tumors are hepatoblastoma and present with an asymptomatic right upper quadrant mass. Symptoms of inanition are more common in older children with hepatocellular carcinoma. Obstructive jaundice may be the presenting symptom in biliary rhabdomyosarcoma. Other rare malignant liver tumors of childhood include a spectrum of sarcomas (angiosarcoma, rhabdomyosarcoma, rhabdoid, undifferentiated), malignant germ cell tumors, and metastatic Wilm's or neuroblastoma. Ultrasound will identify the liver as the organ of origin; additional testing is aimed at distinguishing benign from malignant disease. The serum alpha-fetoprotein (AFP) level is elevated in 90% of patients with hepatoblastoma and one-half of children with hepatocellular carcinoma. A CT scan outlines the anatomic extent of the tumor, clarifies its relationship to the central venous, arterial,

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and biliary conduits, and evaluates for multicentricity. Chest CT at diagnosis will show lung metastasis in approximately 20% of hepatoblastomas, and approximately 40% of childhood hepatocellular carcinomas. For small-localized disease, a primary resection is followed by chemotherapy. When the tumor is large, multicentric, and shows radiographic evidence of portal or hepatic venous invasion, or pulmonary metastatic lesions, the chance of curative resection may be improved by initial biopsy, neoadjuvant chemotherapy, and delayed definitive resection. Liver transplantation may be the most appropriate definitive resection technique for large tumors involving all four hepatic sectors, for diffusely multicentric tumors, or for tumors with unreconstructible major venous involvement.

Reoperative surgery in the treatment of the malignant liver tumors is directed either at remediation of a surgical complication or attempted eradication of recurrent or relapse tumor. Surgical complications that may require operative intervention encompass a wide spectrum including bleeding, impairment of blood flow or biliary drainage, liver failure, infection, and others (1,2) (Table 1). Surgery for tumor relapse may be exploratory in the setting of a positive margin or elevation in AFP, directed at extirpation or biopsy of a radiographically apparent local or distant metastatic lesion, or involve a second tumor. Surgery was reported for three second tumors in the long-term follow-up of children enrolled in the Intergroup Children's Cancer Group (CCG), Pediatric Oncology Group (POG) study INT-0098 and included scapular desmoid tumor, colon adenomatous polyp, and a salivary pleomorphic adenoma (3).

Table 1
Complications of Liver Tumor Surgery (1)

<i>Bleeding</i>	<i>Blood flow</i>	<i>Bile drainage</i>	<i>Liver failure</i>	<i>Infection</i>	<i>Other</i>
Intraoperative hemorrhage	Obstructed venous outflow	Bile leak	Coagulopathy	Sepsis	Bowel obstruction
Postoperative hemorrhage	Hepatic artery injury or thrombosis	Biliary fistula	Hypoglycemia	Cholangitis	Diaphragm injury
Intrahepatic hematoma	Portal venous injury or thrombosis	Biloma	Encephalopathy	Wound infection	Pleural effusion
Hemobilia	Hepatic necrosis	Biliary stricture	Ascites	Hepatic or perihepatic abscess	Wound dehiscence
Gastrointestinal bleeding	Bile peritonitis			Pneumonia	Gastric outlet obstruction
				Peritonitis	Second tumor

REOPERATION FOR SURGICAL COMPLICATIONS

Bleeding

Operation for bleeding after a biopsy, whether percutaneous or open, will depend on the indications for the biopsy and the extent of bleeding. Bleeding from liver tissue at the site of needle entry can almost always be stopped with pressure, combined with fulguration, or with horizontal mattress suture of the needle entrance site at the liver capsule. Bleeding from a preoperative tumor rupture often produces an intracapsular hematoma and may stop spontaneously. Occasionally hepatoblastoma or hemangioma may present with hemorrhage and free tumor rupture into the peritoneal cavity with life-threatening hypovolemic shock. In these instances the initial operation is directed at control of the hemorrhage, either with resection if it can be performed in a controlled, expeditious manner, or a nonanatomic debulking procedure in combination with fulguration, hemostatic agents, and if necessary packing followed by percutaneous tumor embolization (4). Inadvertent injury to vital structures can be minimized if heroic, uncontrolled procedures are avoided and definitive surgical resection is performed as a second-look procedure, once hemodynamic stability is fully restored and complete radiographic tumor staging has been completed. Massive blood transfusion should be avoided where possible, as the extent of blood transfusion has been correlated with postoperative morbidity and has been reported to increase the risk of tumor recurrence in hepatocellular carcinoma (5).

Postoperative intrahepatic or perihepatic hematoma may require surgical drainage, predisposes to the risk of abscess, and may delay the administration of adjuvant chemotherapy. The preferred treatment is prevention, and a postoperative hematoma is best avoided by meticulous attention to hemostatic surgical technique. Resection of large tumors requires extensive preoperative planning by an experienced surgical team, wide exposure, the appropriate use of inflow and/or outflow occlusion, close communication with anesthesia to keep the central venous pressure (CVP) relatively low and clotting factors appropriately replaced, and the avoidance of unplanned, inappropriately aggressive attempts at tumor resection in proximity to major vessels (6). In the event of a failed initial resection attempt, reoperative resection may be associated with increased perihepatic blood loss because of the density of liver adhesions to the diaphragm, retroperitoneum, and right adrenal gland. There is difficulty staying out of the liver and out of the diaphragm and adrenal gland when attempting to dissect those structures one from the other. Judicious use of electrocautery and good exposure are invaluable. Special care should be exercised when approaching the liver hilum and the regions of the vena cava and the termini of the hepatic veins.

Vascular Injury

A normal liver can occasionally survive permanent interruption of arterial inflow or portal venous inflow, but not both. In the rare instance that inflow of the portal vein, hepatic artery, or both are disrupted at surgery, salvage requires immediate revascularization. Fulminant hepatic failure will progress rapidly with total devascularization of a normal liver and occurs with less than total devascularization of a cirrhotic or otherwise damaged liver. Alternatively, if these vital vessels are not gently handled with care throughout the operative procedure, postoperative thrombosis can occur. Early diagnosis and prompt revascularization with some combination of percutaneous catheter infusion of thrombolytic agents, thrombectomy, and/or operative vascular

repair may be life-saving. When there is irretrievable liver devascularization, the only option may be emergent liver transplantation.

Maintenance of venous drainage from the residual liver remnant is of paramount importance, as liver tissue without adequate venous drainage will become congested and not remain viable. Protection of venous drainage includes judicious planning of anticipated surgical resection margins, and avoidance of inadvertent hepatic venous occlusion with ill-placed sutures into the hepatic parenchyma in an attempt to control bleeding. In extreme cases, salvage transplantation or venous outflow reconstruction with either autologous or synthetic grafts may be required (7).

Biliary Injury

The bile ducts, particularly at the hilar plate, are more easily disrupted than the vessels and are usually more difficult to dissect individually at that level. If injury occurs to an extrahepatic bile duct and the injury is minor, it can usually be directly repaired. If there is major injury, with loss of more than 50% of the wall, complete division, or loss of length, the remaining ductal system should be debrided back to healthy well-perfused ducts and drained with a Roux-en-Y jejunal limb.

Bile leak occurs in approximately 5–10% of patients after major liver resection and can be lessened by meticulous dissection and closure of all sites of bile drainage on the raw surface of the liver. Although placing drains at the time of operation does not lessen the rate of bile leakage, it may facilitate postoperative management (8). Bile peritonitis results from bile accessing the general peritoneal cavity rather than being walled off into a discrete collection (biloma) or external drain (fistula). Treatment of large-volume bile peritonitis often requires a combination of peritoneal drainage and decompression of the biliary tree with a transhepatic or endobiliary drain. Bile leaks that do not respond to appropriate drainage are often associated with a distal obstruction. Etiologies of distal bile duct obstruction include a retained section of viable liver excluded from the remaining biliary system, iatrogenic occlusion (clip, ligature, thermal injury), hematoma, stone, residual obstructing tumor, or an ischemic stricture. If the fistula is confined to a drain, the drain should be left in place for several weeks for formation of a fibrous tract and resolution of self-limiting forms of distal obstruction (e.g., hematoma). If leakage persists and no distal obstruction is identified with magnetic resonance cholangiopancreatography (MRCP), hepatobiliary radiotracer 99mTc dimethyliminodiacetic acid (HIDA), or endoscopic retrograde cholangiopancreatography (ERCP), the drain should be advanced slightly away from the liver edge to encourage fibrous obliteration of the leak. If the leak or radiographically evident site of distal biliary obstruction persists, operative biliary reconstruction with Roux-en-Y jejunal limb is indicated (Fig. 1). When the treatment goal is curative rather than palliative, biliary stricture treatment with percutaneously placed stents provide a temporary (not long-term) solution, especially in children. Stents have a limited lifetime (usually <16 weeks) because of build-up of bile salts and proteinaceous material with occlusion, repeat obstruction, and cholangitis. Most critical in treating and avoiding cholangitis is the operative establishment of adequate biliary drainage.

Liver Failure

The most common cause of postoperative liver failure following liver resection is a small functioning liver remnant, generally a much more common concern in

- Drainage is primarily through the external drain site
- Only a small amount of activity in duodenum on delayed images suggests partial obstruction of the extrahepatic biliary tree

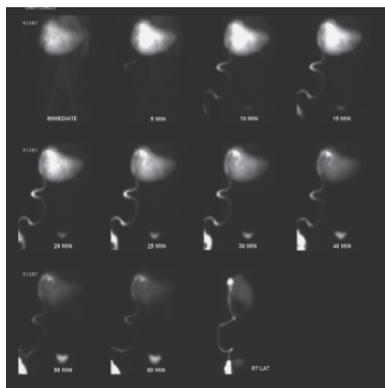


Fig. 1. Biliary fistula. HIDA scan shows all tracer exiting the liver through the external drain site with minimal activity in the extrahepatic biliary tree.

the adult population, where hepatocellular carcinoma often occurs in patients with preexisting liver disease. The decision about how much liver may be removed when doing liver resection or ablation is based on the anatomic extent of the tumor and the associated liver anatomy. A guideline has been that long-term survival and appropriate liver regeneration can be anticipated with a remnant volume of 25–30% of normal liver, or 60–70% of cirrhotic liver. Other etiologies of postoperative liver failure discussed previously include liver devascularization, interruption of venous drainage, excessive liver warm ischemia owing to prolonged operative vascular occlusion and massive bleeding, major bile duct obstruction, halogenated anesthetic agents, viral infections, and reactions to certain drugs. If vascular occlusion is used intraoperatively, there has been an increasing trend toward “ischemic preconditioning,” using an initial 10-minute period of warm ischemia, followed a 10-minute period of reperfusion, after which a continuous longer stretch of warm ischemia seems to cause less liver cell injury (9). Although warm ischemia up to 60 minutes may be tolerated, most experienced liver surgeons will limit inflow occlusion to periods of 15–20 minutes, interspersed with 5–10 minute intervals of reperfusion. If postoperative liver failure does not show definitive signs of improvement in the first few days, and is not amenable to revascularization or biliary decompression, the potential need for liver transplantation should be considered.

OPERATIVE PROCEDURES FOR TUMOR RELAPSE

Local Tumor Persistence or Relapse

In all patients, but especially in those whose initial operative resection results in gross or microscopic tumor residual, careful follow-up with radiographic imaging and serial AFP levels is mandatory. Atypical or wedge resections—as opposed to anatomic lobectomy, trisegmentectomy, or mesohepatectomy—have been associated with an increased risk of local relapse (10). Second-look laparotomy with biopsy and possibly extension of the liver resection is indicated in the setting of a radiographically identifiable small focal mass lesion, or a persistent elevation in the AFP after completing the prescribed courses of postoperative chemotherapy. In SIOPEL-1, a multicenter trial from the International Childhood Liver Tumour Strategy Group (SIOPEL), there were

five of 128 patients who underwent surgery for a local recurrence after having had a definitive resection (11). Of these five patients, two showed no evidence of disease at long-term follow-up, one died during surgery because of uncontrollable bleeding. One patient developed a second recurrence and then a third, underwent “rescue” transplantation and died shortly thereafter. The fifth survived after four recurrences, three local recurrences were treated with resection, followed by prolonged chemotherapy for a lung recurrence, followed by liver transplantation.

The potential role of positron emission tomographic (PET) imaging in screening for tumor recurrence when serum AFP is elevated, but CT or MRI imaging is normal, remains controversial. There are case reports of PET identification and subsequent successful surgical resection of small occult lesions occurring locally, within the abdominal cavity, and in the lung (12). Caution must be exercised because of similar reports describing operative exploration and biopsy of histologically negative regenerative liver nodules in patients with a positive PET scan (13) (Fig. 2).

Heroic resections with a high probability of leaving residual tumor should be avoided whenever possible (14). If large or extensive residual or recurrent tumor is present in the remaining liver, a “rescue” liver transplant may be required (15,16). A recent analysis of the SIOPEL 1 data and a review of the world experience on transplantation for hepatoblastoma clearly showed that a primary transplant after good response to chemotherapy is now associated with a prognosis similar to a conventional complete resection, with a long-term survival of more than 80%. In contrast, “rescue” transplant for recurrent tumor after resection had only a 30% chance of cure (15). Transplantation for hepatocellular carcinoma and other malignant tumors is more problematic, and as a routine should be strictly reserved to tumors without extrahepatic extension (17). Recently, the SIOPEL study group, in collaboration with the COG, SPLIT pediatric liver transplant registry, and German cooperative liver tumor study group (GPOH), have established a worldwide electronic registry for transplantation in the treatment of childhood hepatoblastoma and hepatocellular carcinoma. This international cooperative registry is coined PLUTO (Pediatric Liver Unresectable Tumor Observatory), is administered by the Italian group CINECA, and can be reached at www.PLUTO.org (18).

Whether to treat primary or relapse tumor that focally involves the hepatic venous outflow tract with resection and extensive venous reconstruction, transplant, or both

- PET Scan shows increase in activity at the cut surface of the regenerating liver 2 months after lobectomy

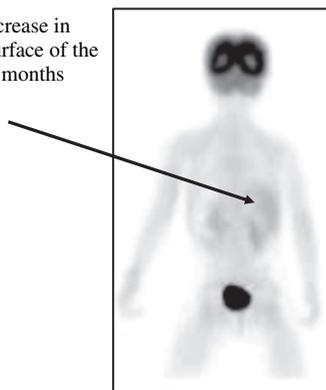


Fig. 2. False-positive PET Scan. PET scan shows increase in activity at the cut surface of the regenerating liver.

is a matter of much debate. Certainly, the surgical techniques necessary to perform complex venous outflow reconstruction have improved dramatically in recent years. Outflow reconstruction usually involves total vascular occlusion to provide adequate visualization and prevent life-threatening hemorrhage. Other techniques described in the adult literature include veno-venous bypass, *in situ* cooling, or *ex vivo* resection (19–21). Several autologous and synthetic graft materials have been used for the venous reconstruction including saphenous vein, iliac vein, cryopreserved iliac vein or inferior vena cava (IVC), polytetrafluoroethylene (PTFE) or Gore-Tex grafts (WL Gore & Assoc), and bovine pericardium (No-React, Shellhigh Inc.) (20,22–28). Synthetic grafts may carry an increased risk of infection and thrombosis, although harvesting of autogenous tissue requires additional operative procedures with the risk of added morbidity.

Metastatic Tumor Relapse

In numerous large multicenter trials, the incidence of pulmonary metastatic disease at diagnosis is approximately 20% for hepatoblastoma, and approximately 40% for childhood hepatocellular carcinoma. The rate is less clearly defined for the rare hepatic sarcomas. Lung metastasis from hepatoblastoma, but not hepatocellular carcinoma, may resolve with platinum based chemotherapy. If focal pulmonary metastatic disease remains after chemotherapy, thoracotomy for wedge resection has been recommended (3). Nine of the 22 (41%) patients with lung metastasis at diagnosis in SIOPEL–1 were long-term survivors, and four of the survivors underwent pulmonary metastectomy as well as chemotherapy (29). Thoracotomy and biopsy of a lung nodule or focal area of pulmonary atelectasis may be necessary in a patient with elevated screening AFP, or in the setting of impending liver transplantation. Transplantation should never be performed in the face of potential persistent pulmonary metastatic disease. Suspicious lung lesions must disappear completely on chemotherapy, be amenable to complete surgical resection, or be biopsied to prove that they are benign.

Two of five patients in SIOPEL–1, without metastatic disease at diagnosis, developed tumor relapse in the lungs after initial therapy and underwent pulmonary metastectomy. Both were long-term survivors (11). Although the role of pulmonary metastectomy remains unclear, surgical resection of pulmonary metastases seems to be a good treatment option if local control of the liver has been accomplished. The current COG multicenter trial of hepatoblastoma will prospectively investigate the potential role of thoracotomy in these patients.

Rarely, surgery may be required for metastatic disease at a site other than the lung. There is a case report in the literature of a long-term survivor who underwent surgical resection of metastatic hepatoblastoma to the brain (30). The CCG/POG Intergroup-0098 study included five patients who underwent surgical resection of extrapulmonary metastatic disease; three had resection of a mediastinal tumor recurrence, two had resection of a spinal or paraspinous tumor recurrence—none were long-term survivors (3).

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Sacrococcygeal Teratoma

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INTRODUCTION

With an incidence of one in 30,000–40,000 live births, sacrococcygeal teratoma (SCT), one of the type I germ cell tumors (GCT) (1), is truly a very rare tumor. Yet it is the most frequent tumor in newborns. SCT is also the most frequent extragonadal GCT, far ahead of tumors in other extragonadal sites such as the brain, mediastinum, retroperitoneum, and neck.

Diagnosis

SCT occurs most commonly in girls (75–80%). In approximately 70% of the patients, the diagnosis is made at latest at birth, seeing that an increasing number of patients are being diagnosed antenatally. It follows that a still-important subset (around 30%) is being diagnosed at a later age. The Altman classification (2) divides tumors in function of their anatomical extension into four groups (Fig. 1). Roughly one-third of patients can be classified as Altman I (predominantly external tumor with minimal extension into pelvis), one-quarter as Altman II (external tumor with significant intrapelvic extension), 15–20% as Altman III (external tumor with intra-abdominal extension), and finally, some 20% as Altman IV (intrapelvic tumor only, not visible externally) (3,4). A correct clinical diagnosis is usually made on the presence of a sacrococcygeal tumor that displaces the anus ventrally. However, the diagnosis of Altman IV tumors is often delayed until the second or third year of life, following investigation for intractable constipation or urinary symptoms. Imaging studies (computed tomography [CT] or magnetic resonance imaging [MRI]) are crucial for determining the intrapelvic/intraabdominal extension of

the tumor, allowing for correct operative planning. Plasma levels of alpha-fetoprotein (α -FP) and, to a lesser extent, beta-human chorionic gonadotropin (β -HCG), have proven to be very reliable markers for malignancy (yolk sac tumor and choriocarcinoma, respectively). Unfortunately, as α -FP is high at birth and decreases steadily over the first year of life (5,6), it has hardly any value in discriminating between benign and malignant tumor at birth and during the first months of life. The vast majority (i.e., 80–85%) of SCTs are histologically benign (mature and immature teratoma) (3,4,7). Malignancy (in some 15% of cases) is most frequently encountered in yolk sac tumor—embryonic carcinomas and mixed malignant tumors are really exceptional diagnoses. Only 5% of patients show metastases at presentation (4).

Treatment

Treatment has been well standardized for many years already. For SCT diagnosed at birth, complete surgical excision en bloc with the os coccyx is the golden standard and sole treatment. For large tumors, this is best achieved through an inverted V incision; smaller tumors can be managed by a posterior sagittal incision. When the intraabdominal tumoral component is important (Altman III), the abdominal component is first dissected using a Pfannenstiell incision, followed by the perineal approach, allowing for en-bloc removal of the entire tumor together with the coccyx. Newborns will rarely have malignant tumors, and therefore seldom need adjuvant (chemo)therapy. Older children, especially in Altman IV SCT, have a greater likelihood of malignant histology. Tumor resection may also be undertaken as the first step in the face of malignancy (elevated tumor markers, metastases), but if judged inoperable, biopsy or debulking as well as up-front chemotherapy are valid options. In all such cases, the residual tumor has to be removed after several courses of chemotherapy. Since the mid-1980s, very efficacious multiagent chemotherapy regimens have been made available for GCTs; these regimens all include platinum compounds (cisplatin or carboplatin). The PEB regimen of the Pediatric Oncology Group (POG)/Children's Cancer Group (CCG) combines cisplatin (P) with etoposide (E) and bleomycin (B). The PEI regimen according to the MAKEI protocol of the German Association of Pediatric Oncology prescribes cisplatin (P) in combination with etoposide (E) and ifosfamide (I). The JEB protocol of the United Kingdom Children's Cancer Study Group (UKCCSG) blends carboplatin (J) with etoposide (E) and bleomycin (B). These are all very efficacious protocols that yield comparable results but differ as to number, type and severity of complications.

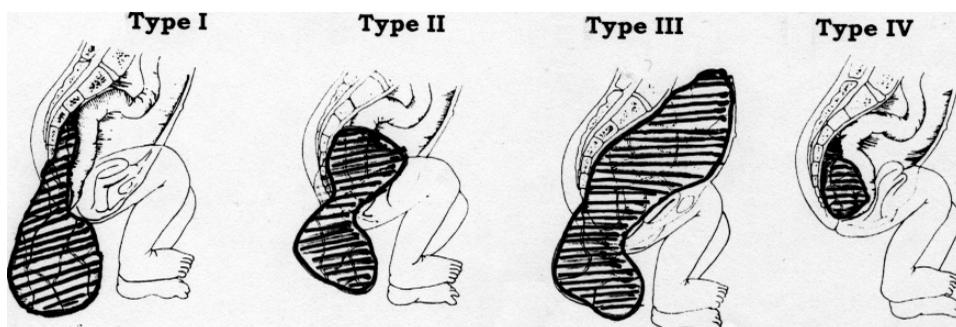


Fig. 1. Altman classification of SCTs.

Results

Overall, with a survival rate of more than 95%, the prognosis for newborns and children with SCT is excellent. However, a significant proportion will suffer from functional sequelae (fecal soiling, constipation, fecal incontinence, urinary incontinence, and aesthetic sequelae) that impair quality of life (8). A small proportion will show tumor recurrence, either histologically benign or malignant. Patients with malignant SCT are facing a less favorable prognosis than those with benign histology, but substantial improvements have been noted over the past 20 years. Survival rates from 80–90% are now achieved in subgroups with malignant tumors (9,10).

SOME EPIDEMIOLOGICAL DATA ON RECURRENCE

One of the most feared complications is recurrent tumor, as it spells impaired prognosis. Table 1 summarizes the percentages of recurrences according to histology as reported in the most recent series (3,4,7,11–20). Of course, these series show important variations. Overall, recurrence is reported in 3–35% of the patients (mean: 12.9%). Initial histological diagnosis of mature teratoma is associated with a mean 10.2% recurrence rate (range 0–26%). Recurrence rates double for immature teratoma (mean: 20.2%, range 4–55%) and initial malignant histology (yolk sac tumor or embryonic carcinoma) (mean: 19.2%, range 0–100%). Mean time between primary surgery and diagnosis of recurrence is 10 months; however, extremes range from 1–35 months. More than 50% of recurrent SCTs show comparable histology after resection of the primary SCT and after resection of the recurrent tumor, but a shift towards either a more undifferentiated tumor or towards a more differentiated tumor has also been observed (4). Malignancy after resection of a benign teratoma could result from a sampling error missing small foci of malignancy, or could develop from a small malignant focus left in place after surgical excision. In a study of the CCG-POG, four of the six children with malignant recurrence after neonatal resection of mature or immature teratomas were noted to have microscopic foci of yolk sac tumor in the original specimen at reexamination (21). Most recurrences in children with SCT occur at the primary site of the tumor, but distant metastases are seen as well. As mentioned earlier, recurrent tumor is associated with serious impairment of prognosis. Overall mortality in patients with recurrent SCT (all histologies) is reported to equal 35% (3) (Fig. 2). However, malignant recurrence is associated with mortality rates as high as 55% (22). It is therefore of extreme importance to prevent recurrence whenever possible. This would require full knowledge of the factors that are possibly associated with recurrence.

FACTORS ASSOCIATED WITH RECURRENCE

Insufficient local therapy has been documented to represent an important risk factor of recurrence (9,22). For quite a while already, surgeons have accepted the axiom that complete en bloc resection of the primary sacrococcygeal tumor and the coccyx without tumor spillage offers the best perspective of tumor control. This cannot always be (easily) achieved. Histologically benign tumors (teratomas) are mostly well encapsulated, but tumor borders may be difficult to identify in some regions. In particular anatomical sites, it is impossible to remove the tumor with a sheet of healthy tissue all around. Also, if a substantial intrapelvic tumoral component is present, as in Altman

Table 1
Survey of mortality and recurrence rates reported in 825 patients with SCT

Author (ref)	Year of publication	Period	No. of patients	Mortality (%)	Recurrence				Comment
					MT	IT	“Malignant” GCT	Total recurrences (%)	
Gonzalez-Crussi et al ¹¹	1978	1946–1976	40	22%	4/22 (18%)	10/18 (55%)	0	14/40 (35%)	Exclusively benign SCT
Ein et al ¹²	1980	1951–1976	33	3%	1/33 (3%)	0	0	1/33 (3%)	Exclusively benign SCT
Engelskirchen et al ¹³	1987	1960–1984	87	16%	9/76 (12%)	0	4/11 (36%)	13/87 (15%)	
Schropp et al ¹⁴	1992	1950–1990	73	16%	5/57 (9%)	0/16 (0%)	5/73 (7%)		
Havranek et al ¹⁵	1992	1980–1989	32	6%	4/23 (17%)	2/4 (50%)	1/5 (20%)	7/32 (22%)	
Biilik et al ¹⁶	1993	1972–1990	28	?	6/28 (21%)	0	0	6/28 (21%)	Exclusively benign SCT
Rescorla et al ⁷	1998	1972–1994	126	7%	9/80 (11%)	1/24 (4%)	2/13 (15%)	12/117 (10.2%)	Multicentric study (15 centers)
Schmidt et al ¹⁷	1999	1976–1995	23	22%	5/19 (26%)	3/3 (60%)	8/22 (35%)		
Wakhlou et al ¹⁸	2002	1983–2000	72	28%	0/47 (0%)	0	5/25 (20%)	5/72 (7%)	
Perelli et al ¹⁹	2002	1985–1998	17	29%	2/11 (18%)	1/3 (33%)	0/3 (0%)	3/15 (20%)	
Huddart et al ²⁰	2003	1989–1997	51	4%	5/29 (17%)	2/16 (12.5%)	0/6 (0%)	7/51 (14%)	Exclusively newborns < 4 wk
De Backer et al ³	2005	1960–2003	70	8.5%	3/43 (7%)	2/11 (18%)	0/16 (0%)	5/70 (7%)	
Derix et al ⁴	2006	1970–2003	173	9.8%	6/110 (5%)	4/33 (12%)	8/22 (36%)	19/173 (11%)	
Total			825		59/578 (10.2%)	22/109 (20.2%)	23/120 (19.2%)	105/813 (12.9%)	

MT mature teratoma

IT immature teratoma

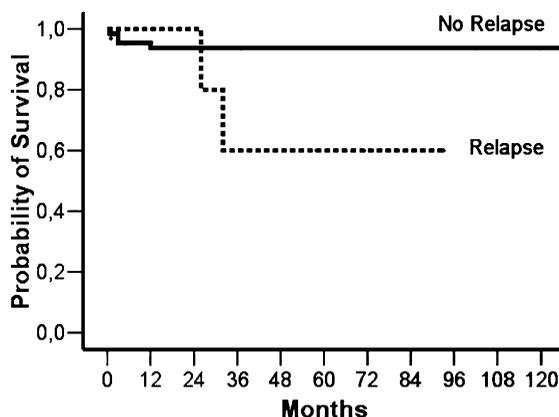


Fig. 2. The 10-year overall survival (OS) according to occurrence of relapse in a series of 70 patients with sacrococcygeal teratoma, treated between 1960 and 2003, showing that OS in patients who experienced relapse was significantly lower than in those patients without relapse. Reprinted from ref. 3, with permission.

types II–IV, dissection may be difficult because of the narrow space to operate in. In histologically malignant SCT, there is often local bone involvement and infiltration into the spinal canal or the nerve sheets of the sacral plexus. In two recent publications, we attempted to identify surgical and nonsurgical risk factors associated with recurrence. One described the courses of 70 patients with SCT treated in two institutions between 1960 and 2003 (3). Tumor recurrence was observed in five patients (7% of total), leading to death in two of them. The second paper analyzed the fates of 173 patients with SCT treated in the six centers for pediatric surgery in the Netherlands between 1970 and 2003 (4). A total of 19 patients suffered recurrence (11% of total), leading to death in seven. From the findings of these studies, which have some patient overlap, we concluded that age at diagnosis, year of diagnosis, Altman stage, and tumor size are not associated with recurrence. Results regarding the influence of complete coccygectomy were somewhat conflicted: in the first study, this factor had no impact on overall survival, but the event-free survival was lower in patients whose coccyx was left *in situ*. In the second study, coccygectomy did not appear to be a risk factor for recurrence. Rupture of tumoral cysts during the operation with spill of cyst fluid did not influence the outcome. Rupture of solid tumor, on the other hand, impaired the outcome in a statistically significant manner. In the first study, we could demonstrate that microscopic involvement of the resection margins of mature or immature SCT (if macroscopically completely excised) was not associated with recurrence, provided there were no foci of yolk sac tumor in the resection margins. The second study showed that patients with pathologically proved incomplete resection had higher risk of recurrence. With regard to this aspect, the study did not distinguish between macroscopic and microscopic resection, or between benign and malignant histology. Moreover, immature histology and malignant histology were also found to be risk factors for recurrence (4).

The conclusions from these studies are not really surprising: complete resection of tumor without spill offers patients the best guarantee for recurrence-free survival. Contradictory to what we found regarding the necessity for en bloc coccygectomy, we still would recommend performing coccygectomy, as the small sample size in that particular study does not really allow for a solid conclusion.

DIAGNOSIS OF RECURRENT SCT

The necessity of following the patients after discharge is widely recognized. A strict follow-up protocol for all the patients with SCT is in place in our department. For patients with benign SCT, this protocol includes regular control visits with clinical examination, determinations of serum α -FP, β -HCG, and pelvic ultrasound; patients are seen every 3 months during the first year, every 4 months during the second year, every 6 months during the third year, and finally at 4 and 5 years after treatment. For those with malignant histology, the protocol is even more intense. As a consequence, most recurrent tumors will be diagnosed during these follow-up visits, either through clinical rectal examination, elevated tumor markers, abnormal ultrasound scan, and/or other imaging. The usual symptoms in local recurrence are constipation, tenesmus, and globus of the urinary bladder. Symptoms associated with distant metastases (usually lungs, liver, lymph nodes) are anorexia, weight loss, unexplained fever, respiratory insufficiency, and others. Relapse calls for a complete clinical and radiological restaging, including imaging by either CT scan or MRI.

PRINCIPLES OF REOPERATIVE SURGERY

Presumably Benign Recurrence

These are the patients showing a well-circumscribed tumor that does not infiltrate into the sacrum, who have no distant metastases, and whose tumor markers are not raised. Complete surgical excision of the tumor is the only treatment. Rectal preparation is carried out preoperatively. The patient is positioned in the prone, jack-knife position. The rectum is filled with a polividone iodine gauze. The approach depends on the precise location and extent of the recurrent tumor and of the former scar. A posterior sagittal incision usually provides adequate exposure for centrally located tumors. The operation should not be mutilating. Sometimes, the rectum appears to be invaded by tumor, but there is no place for rectum amputation. Rarely, a combined abdominoperineal approach may be necessary.

Presumably Malignant Recurrence

These patients usually show raised tumor markers; tumors are very irregular, invade the sacrum, and some show distant metastases. Surgery alone may be adequate therapy for stage I lesions (7). In all other cases, surgery must be combined with chemotherapy and occasionally radiotherapy, autologous bone marrow transplantation, and/or locoregional hyperthermia (22). If complete surgical excision is judged feasible, this may be undertaken. Otherwise, up-front chemotherapy (after biopsy, if requested) followed by surgical resection of the residual tumoral mass is another valid option.

LONG-TERM RESULTS

Benign Recurrence

In our experience, patients with recurrent mature teratoma all do well after complete excision (3,4). However, we have observed some deaths among those with immature recurrent teratoma.

Malignant Recurrence

There are two reports detailing the outcomes of children with recurrent malignant SCT. One describes six patients from the POG and CCG study with malignant recurrence of mature or immature teratoma (21). Four responded to therapy, one died, and one was lost to follow-up. The second is a large series of 22 patients with recurrent malignant sacrococcygeal GCTs, registered in the MAKEI protocols (22). Of these, 17 presented with an isolated local recurrence, two showed a distant relapse, and three patients suffered from combined local and distant recurrences. Those 22 patients presented with 22 first, 14 second, five third, and two fourth relapses. A total of 12 patients achieved complete remission, seven of whom remained in continuous complete remission. Five of these patients experienced a second relapse. All patients who achieved only a partial remission after the first relapse developed a second relapse. Only three of 14 patients who developed a second relapse could be cured. Altogether, 10 of 22 patients survived disease-free (45%), and 12 patients died as a result of tumor progression or therapy-related complications.

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Recurrent Hernia, Hydrocele, and Varicocele

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CONTENTS

RECURRENT INGUINAL HERNIA
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RECURRENT INGUINAL HERNIA

Technical Variations in Pediatric Inguinal Hernia Repair

The technique of pediatric inguinal hernia repair varies among pediatric surgeons according to training and personal habits developed over time (1). Variations in technique include the degree to which the inguinal canal is exposed (stretching of the external ring or long incision in the external oblique), the extent of the retroperitoneal dissection of the indirect sac (i.e., how high the high ligation is performed), the frequency of addition of a floor repair to indirect hernia repair (never, frequently in premature infants with “weak floors”), handling of the distal hernia/processus vaginalis/tunica vaginalis testis (ignore it every time, explore it every time, selectively explore), the decision to create surgical fixation of a mobile testicle, type of suture material, open versus laparoscopic technique, and the use of prosthetic material in adult-size teenagers. Common to every pediatric hernia is the “high” ligation of the indirect hernia sac. The contribution of the technical variation to the risk of recurrence is unknown. It is unknown whether laparoscopic or laparoscopic-assisted ligation of indirect inguinal hernias in infants and children will result in higher (or lower) recurrence rates with long-term follow-up.

Factors Affecting Inguinal Hernia Recurrence

Recurrence rates for inguinal hernia repair in infants and children are lower than in adults, and vary from less than 0.5% to approximately 4% (2–6). The recurrence rate

for inguinal hernia repair may be lower when performed by trained pediatric surgeons compared to adult general surgeons (7). Patient characteristics and biologic factors may predispose to much higher rates of recurrence. Premature neonates with repair of symptomatic hernias are at increased risk compared to other neonates and children. Chronic lung disease and the presence of ascites increase the complication rate for hernia repair in all ages. In the pediatric population, cystic fibrosis patients and those with ventriculoperitoneal shunts or peritoneal dialysis catheters are at higher risk for recurrence. Patients with connective tissue disorders such as mucopolysaccharidoses and Ehlers-Danlos or Marfan syndrome are also at higher risk for recurrence. Patients with irreducible incarcerated hernias requiring emergent surgery are at increased risk of recurrence. Difficult dissection, damage to surrounding structures, and tissue edema are implicated. The use of rapidly absorbing suture material such as “catgut” may also predispose to recurrence. Abnormalities of the collagen matrix in patients with recurrent inguinal hernias may account for recurrences in some patients (8).

Clinical Presentation and Diagnosis of Recurrent Inguinal Hernias

SWELLING AFTER INDIRECT INGUINAL HERNIA REPAIR

Inguinoscrotal swelling that occurs after pediatric inguinal hernia repair may or may not be a result of recurrence of the hernia. Recurrent/persistent hydrocele after hernia/hydrocele repair is discussed later in this chapter. All questions of recurrence of an inguinal hernia in child should be addressed thoughtfully. One should not overdiagnose the problem in patients operated on by others or deny or underdiagnose in patients operated on by oneself. Pediatric surgeons may be asked to assess patients postoperatively with impressive inguinoscrotal swelling and concerns for early hernia recurrence. Significant edema may occur after the repair of giant inguinal hernias in premature infants. Careful physical exam, possibly supplemented by ultrasound evaluation, is needed to distinguish tissue edema/swelling from early technical failure.

DIRECT AND FEMORAL HERNIAS RECOGNIZED AFTER INDIRECT INGUINAL HERNIA REPAIR

A direct inguinal hernia may occur after pediatric hernia repair for several possible reasons. Excessive widening of the internal ring may predispose to direct hernia recurrence. It is not known whether maneuvers that tighten the internal ring or the addition of an inguinal canal floor repair reduce this risk. The floor of the inguinal canal can theoretically be injured by excessive dissection at the time of indirect repair. Minimizing the posterior dissection of the cremasteric fibers maintains the integrity of the floor of the inguinal canal. Weakness of the floor of the inguinal canal is likely a predisposing factor for direct recurrence in premature infants and may account for their much higher recurrence rate. It is unknown whether floor repair at the time of the initial repair of the premature infant with very large indirect hernia (large enough to encroach on the direct space) increases or decreases recurrence rate. Experienced pediatric surgeons are aware of the clinical scenario of a patient with a persistent hernia after indirect hernia repair that proved to be a femoral hernia or a direct hernia. Inguinofemoral masses that are present both before and after indirect inguinal hernia repair likely represent missed femoral or direct hernias (9). These are not true recurrent hernias, but rather represent “misdiagnosed” hernias. Pure direct and femoral hernias are relatively rare, and indirect inguinal hernias are very common in children. The

importance of a careful history and physical exam is obvious. One advantage of a laparoscopic hernia repair may be the ability to differentiate more accurately among the different defects.

TECHNICAL FAILURES AND RECURRENCE

If the hernia sac is not dissected far enough and the “high ligation” of the hernia sac retains enough of the proximal portion of the hernia sac, an indirect hernia may persist. This condition is not technically a recurrent inguinal hernia, but rather should be termed a “retained” hernia. The presence of visible bulging along the inguinal canal, near the level of the internal ring in slender patients without other symptoms, should be observed. The natural history of this finding is unknown, and does not warrant reoperative surgery with its attendant risks. Other technical failures may result in a “recurrent” inguinal hernia. Immediate or early “recurrence” of an inguinal mass after a pediatric hernia repair results from failure to control the hernia sac circumferentially in the ligation sutures or failure to recognize a tear in the sac. Viscera escape from the peritoneal cavity into the inguinal region after the procedure. Twisting and double ligation of the sac with permanent or long-term absorbable suture minimizes this problem. Reoperation with control of the peritoneal opening is required. Excessive dissection has been implicated in damage to the floor of the canal and may increase the direct recurrence rate; however, the sac must be dissected adequately to assure its integrity and the completeness of the high ligation. Inspection of the distal sac below the site of division of the sac helps confirm that the entire sac has been completely circumferentially isolated. Incomplete isolation of the sac usually occurs with a portion of the sac overlying the cord left behind. Rents in the hernia sac, which usually occur laterally, may extend deep into the retroperitoneum. All defects and rents in a hernia sac can be fully assessed and repaired with adequate mobilization if recognized at the time of initial repair.

TRUE RECURRENT INGUINAL HERNIAS

Any child who has had repair of an indirect inguinal hernia is at risk for recurrence. Progressive inguinal bulging may be either at the indirect space (probably related to technical failure as noted previously) or in the direct space, representing a true weakness of the floor of the inguinal canal. The presence of a reducible mass that contains visceral contents is the most definitive indication for reoperative surgery. More subtle masses should be observed for progression. A progressively enlarging inguinal bulge is an indication for reoperation. Photographic documentation may assist in the objective assessment of progression. Unresolved questions of recurrence often persist, especially in patients with other symptoms such as pain and in those with higher body mass index. Diagnostic imaging is rarely helpful or indicated, but computed tomography and ultrasound may be considered. Diagnostic laparoscopy may be needed to definitively answer the question of recurrence. Indirect, direct, and femoral hernias are visualized definitively and repaired either laparoscopically or by open technique. This has been particularly helpful to rule out a recurrent hernia in patients with unremitting symptoms. When the definitive diagnosis of a recurrent hernia has been made, repair is recommended.

Surgical Correction of Recurrent Inguinal Hernias

Risks of reoperation include greater likelihood of nerve injury, damage to the vas deferens and/or testicular blood supply, and subsequent testicular atrophy. There is no data regarding the risks of second recurrence in pediatric hernia repair. It seems likely that weakness of the floor of the inguinal canal will predispose these patients to additional recurrence later in life. As the cadre of survivors of prematurity ages, the long-term results of our pediatric surgical efforts will become apparent.

The standard principles of all reoperative surgery should be considered in recurrent hernia repair (10). An alternative anatomic approach to the condition should be considered. If the original inguinal approach is chosen, wider exposure of the reoperative field is used to allow clear anatomic visualization. Laparoscopic repair and preperitoneal repair represent alternative anatomic approaches that allow the problem to be visualized from a different orientation (11). These approaches may allow dissection and repair to be performed with less risk of injury to the cord structures. The laparoscopic approach is particularly appealing in the case a recurrent indirect hernia. A high ligation of the indirect sac may be performed under direct vision. There are no outcome data comparing the reoperative inguinal approach with either of these techniques.

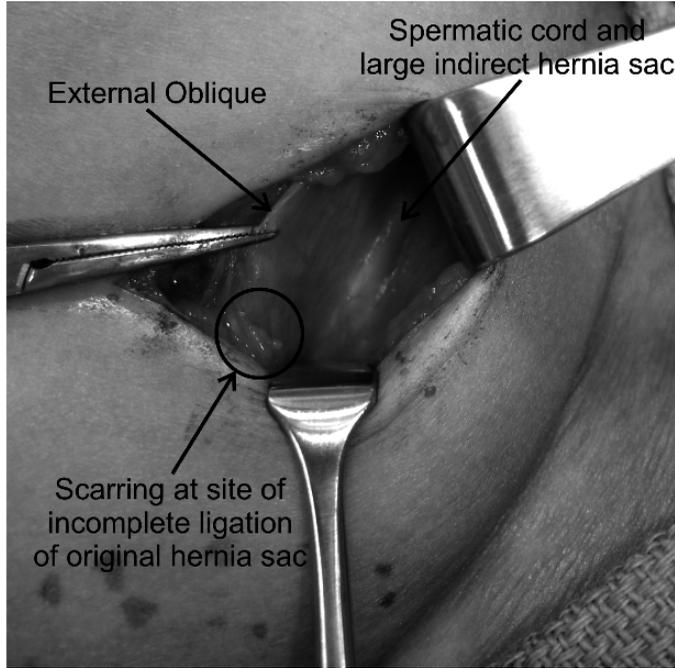
An inguinal operative approach is used most often in cases of recurrent inguinal hernia. As noted previously, laparoscopy may be used as a diagnostic adjunct to confirm the presence and type of hernia. An inguinal approach to a recurrent inguinal hernia is illustrated in Fig. 1. The original incision is expanded and initial dissection is begun in unscarred tissue planes, which are usually found lateral and superior to the original operative site. The cord structures may be identified inferior to the scarred external ring. The inguinal canal is opened where the least scarring is encountered. This

A



Fig. 1. (Continued).

B



C

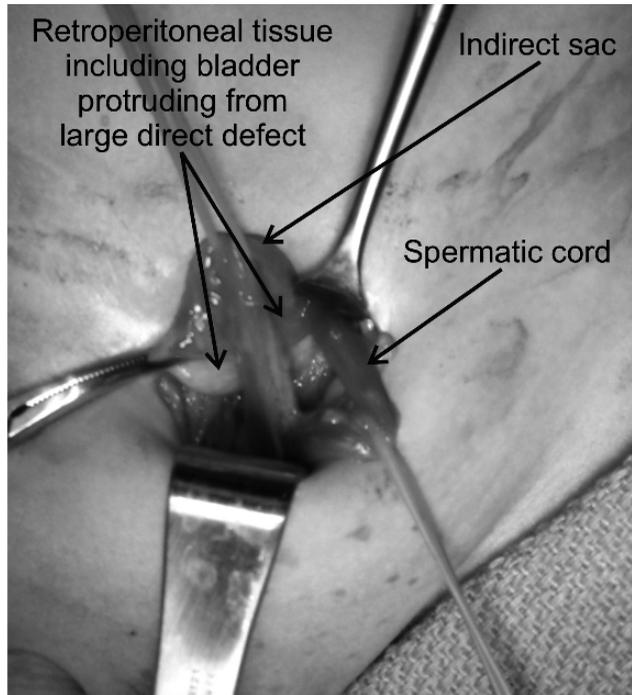


Fig. 1. (Continued).

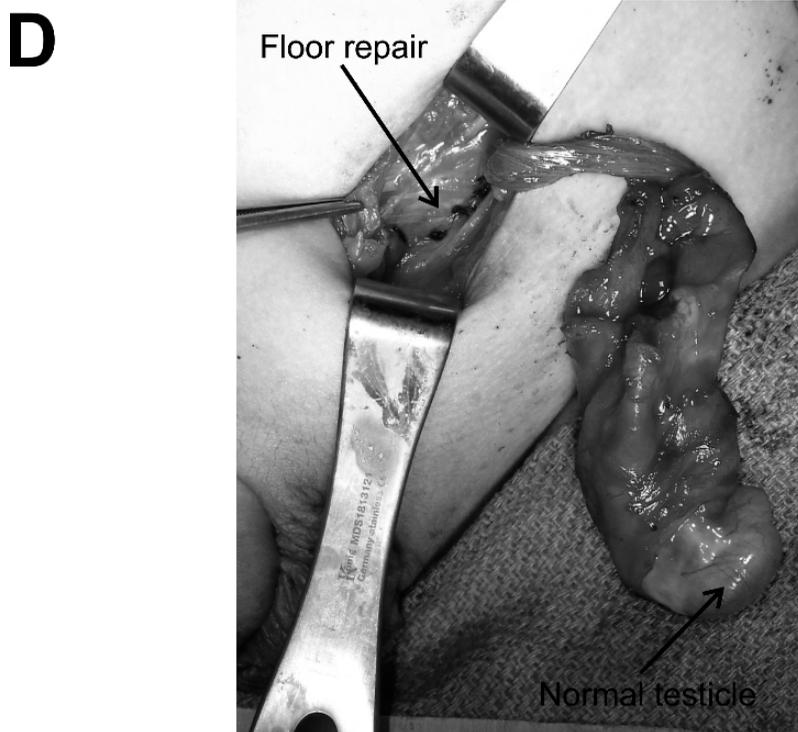


Fig. 1. Operative photographs of recurrent inguinal hernia repair. The patient is an 18-month-old male who underwent left inguinal hernia repair at age 1 month. After 3 months he developed a similar inguinal bulge, which progressively enlarged. He was referred for recurrent left inguinal hernia and reoperation was recommended. The recurrence resulted from inadequate circumferential control of the original indirect hernia sac. The original sac had been partially ligated on its anterior surface. It is uncertain if the large direct defect was caused by damage to the floor of the inguinal canal at the original procedure. (A) The original incision is marked. The extended left inguinal incision and area of visible bulging is indicated. (B) Exposure of the inguinal anatomy. (C) Residual indirect inguinal hernia sac and large direct defect/absent floor of the inguinal canal. (D) The anatomy of the distal sac and cord structures was clarified by disarticulating the testicle. The reconstruction of the floor of the inguinal canal was performed as a Cooper's ligament repair with a relaxing incision. A standard orchiopexy completed the procedure.

may be at the external ring, medial, or lateral to the original incision in the external oblique aponeurosis or superior to the original dissection of the canal. If possible, nerve branches are identified and spared. If a remnant of an indirect sac is present, it is dissected from the vas and vessels superior to the original "high ligation" and ligated again. The integrity of the sac must be maintained. It may be particularly difficult to assess the integrity of the floor of the inguinal canal in a reoperative procedure. Formal reconstruction of the floor is recommended if there is any doubt about its integrity or the type of recurrence that is present. Disarticulation of the testicle while maintaining its blood supply is useful to visualize the floor anatomy in a small reoperative surgical field and may facilitate precise anatomic repair. After floor reconstruction, the testicle may be relocated as in an orchiopexy. Prophylactic antibiotics should be considered in reoperative surgery.

RECURRENT HYDROCELE

A true recurrent hydrocele is defined as the accumulation of scrotal fluid surrounding an otherwise normal testicle that persists beyond the immediate postoperative period after primary hydrocele repair or repair of inguinal hernia where the tunica vaginalis testis was manipulated at the time of initial surgery. The appearance of scrotal fluid after hernia repair that involved only high ligation of the sac and no distal manipulation is not technically a recurrence. There is little information relating the type of hydrocele procedure originally performed (puncture/aspiration, fenestration, partial excision, eversion) and the risk of recurrence.

Postoperative Hydrocele Following Inguinal Hernia Repair and Hydrocelectomy

Inguinal hernia repair and hydrocele surgery are typically included together when describing postoperative hydrocele formation. Postoperative hydroceles typically develop within several months of surgery. Morecroft et. al. reported a rate of 1.4% in their series of 556 boys (12). Most of these resolved without treatment within a few months. Davies et. al., in their series of inguinal hernias in boys less than 3 kg, reported a high incidence of postoperative hydrocele formation of 11% (five of 46) (13). Two hydroceles resolved spontaneously and one resolved after a single aspiration. Two patients underwent exploration and fenestration of the sack, with one hydrocele persisting. Moss and Hatch reported two hydroceles requiring surgical treatment out of 328 boys less than 2 months of age requiring hernia repair (14). In a series of 5343 male hernia repairs, Ein et. al. reported that only two boys required surgery for this complication (15). The incidence of postoperative hydroceles and the use of aspiration were not mentioned. The overall consensus seems to be that hydrocele formation is relatively uncommon after inguinal hernia repair or hydrocele surgery and it should usually be treated by observation, with aspiration being therapeutic for the few that do not resolve spontaneously; reoperation is rarely required (16–18).

Hydroceles That Occur After Varicocele Repair

The occurrence of hydrocele after surgical treatment of varicocele is common. These are not truly recurrent hydroceles, because hydrocele was not recognized before the varicocele procedure. Postoperative rates of hydrocele formation range from 1–40% (19–24). The median time to hydrocele formation is 2–22 months, with some hydroceles appearing as late as 6 years after surgery (21,23). The large variability in the reports is partly explained by the differences in follow-up periods between the studies, with short-term studies likely missing a number of delayed presentations. Hydrocele formation after varicocele treatment presumably results from disruption of lymphatic channels that follow the spermatic vessels (25). Disrupted lymphatics may regenerate or collaterals may develop, resulting in spontaneous resolution of some of these hydroceles. Spontaneous resolution has been reported in 14–60% of postoperative hydroceles (21–23). The management of postoperative hydroceles that do not resolve spontaneously includes aspiration and hydrocelectomy. There is no clear consensus regarding treatment. When hydrocelectomy is performed after varicocele repair, true recurrence is much higher than after simple hydrocele repair. Esposito et al. (23) reported true recurrence in two of six children who underwent hydrocelectomy after varicocelectomy.

Management Strategies for Postoperative Hydroceles

The management of hydrocele formation after inguinal hernia repair, hydrocelectomy, and varicocele surgery has neither clear consensus nor defined algorithm, and to date has been dictated by surgeon experience and preference. Because many such hydroceles will resolve spontaneously, expectant observation should be the initial approach. Appropriate follow-up is necessary to assess for resolution. For hydroceles that do not resolve after months (six) of observation, are very large, or cause discomfort, percutaneous aspiration with local anesthetic should be the first treatment modality. Aspiration can be repeated if fluid reaccumulates. For those hydroceles that do not respond to aspiration after several (up to five) attempts, hydrocelectomy should be considered. Inguinal or scrotal approach for hydrocelectomy is acceptable, and the decision should depend on the surgeon's experience and preference. An inguinal approach is recommended if hydrocele recurs and is unresponsive to aspiration after a transscrotal procedure. The inguinal approach confirms that a hernia/communicating hydrocele is not responsible for the recurrence. If there is any concern for recurrent hernia or communication with the peritoneal cavity, aspirations should not be attempted and an inguinal approach should be taken. These considerations argue for an inguinal approach in all primary pediatric hydrocele repairs.

RECURRENT VARICOCELE

Background on Varicocele (26)

A varicocele is defined as swelling of the veins of the pampiniform plexus that provide venous drainage of the testicle via the testicular veins into the renal vein or vena cava. Varicocele occurs much more commonly on the left and may be bilateral. Small varicoceles are visible only with valsalva maneuver, medium varicoceles are palpable masses, and large varicoceles are visible masses within the scrotum. These severities of varicocele are also referred to as grade I, II, and III. Typically, the varicocele fills with blood when standing and becomes less prominent in the supine position. Untreated varicocele is associated with testicular atrophy and infertility. Patients may experience a sense of fullness, heaviness, or discomfort. Varicocele is thought to result from absence or abnormality of the valves in the gonadal vein, which results in high venous pressure. Elevated venous pressure in the left renal vein has also been implicated. Obstructing retroperitoneal lesions such as tumors must be considered in unusual presentations such as young children. The testicle has additional blood supply besides the testicular vessels, including arterial and venous channels that accompany the vas deferens and cremasteric vessels. These collateral vessels provide adequate arterial and venous circulation to allow division of part or all of the testicular vessels in the treatment of varicocele. Adolescent males with symptomatic varicocele or evidence of testicular atrophy are treated.

Treatment Options for Adolescent Varicocele

Each of the treatment options includes the division or occlusion of the gonadal veins on the affected side. The different procedural options are indicated in Table 1 (20,27–31). There is no consensus on the primary treatment of varicocele in children and adolescents; however, laparoscopic division/ligation of the spermatic vessels (both arteries and veins) has a low recurrence rate as well as low risk of testicular atrophy,

Table 1
Treatment Options for Varicocele

<i>Procedure</i>	<i>Variations</i>	<i>Comments</i>
“Palomo” procedure (mass ligation of testicular artery and vein in retroperitoneum above internal ring)	May be performed using open or laparoscopic techniques	Technically straight-forward, amenable to laparoscopy, collaterals result in low atrophy rate. Conflicting data on incidence of postoperative hydrocele.
Ligation of testicular veins in the inguinal canal	Magnification, microsurgical techniques, Doppler ultrasound and intraoperative venography have been described to allow more accurate identification of veins, arteries and lymphatics	Division of lymphatic channels at this level may account for reports of frequent postoperative hydrocele. Microsurgical technique results in low complication rate.
Ligation of testicular veins below the inguinal canal		Referred to as “subinguinal” approach. May be performed with sedation and local anesthesia.
Retrograde venous injection sclerotherapy (coils and/or sclerosants injected into the spermatic veins via a central venous approach)	Many technical variations described (type of vascular occlusion, exact level of occlusion, inclusion of contralateral vessels) Usually performed by interventional radiologists.	Pampiniform phlebitis and thromboembolic problems are potential complications not present with surgical treatments. May be performed with sedation and local anesthesia. Anatomic variations may prevent use.
Antegrade sclerotherapy (injection of sclerosants into pampiniform plexus vessels below the inguinal canal)		May be good choice for recurrent varicocele. Requires operative exposure of pampiniform vessels in upper scrotum. May be performed with sedation and local anesthesia.

Techniques and procedures used to treat varicocele.

presumable owing to preservation of the collateral testicular blood supply. Previous reports of low risk of post-operative hydrocele have been questioned (24). I have utilized the technique of ligation of the testicular veins within the inguinal canal using magnification and Doppler ultrasound (A theoretical advantage of this technique is that it allows the processus vaginalis to be identified and ligated, eliminating a “missed communicating hydrocele” as a cause for postoperative hydrocele.).

Postoperative Care and Recurrence

After any procedure for varicocele, patients should be monitored for testicular growth and size, recurrence of the varicocele, and hydrocele formation. Hydrocele formation may be delayed and patients should be instructed of this. Recurrence and postoperative hydrocele are the most important complications that may require reoperative treatment. Postprocedural hydrocele may respond to aspiration or require operative hydrocelectomy. Recurrent hydrocele, even after operative treatment, is common as discussed previously. If varicocele recurs after primary treatment, it must be assumed that some of the testicular veins were not identified and divided with the original procedure or that other collaterals exist. Although reoperation through the same anatomic approach is feasible, reoperation via an inguinal approach, if this was used for the first procedure, puts the remaining testicular blood supply at risk. Antegrade sclerotherapy has been applied to treat failed surgical ligation and failures after retrograde sclerotherapy (32). Recurrence after failed retrograde sclerotherapy has been successfully treated with repeat retrograde sclerotherapy (33). The Palomo procedure has been used for sclerotherapy failures as well. Venography performed in conjunction with sclerotherapy can be used to identify the presence of persistent vessels and to identify any vascular anomalies that might preclude success. Because the patterns of recurrence and the anatomic reasons for recurrence may differ with each technique, it seems reasonable to recommend an alternate anatomic approach for symptomatic recurrence.

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Failed Orchiopexy and Complications of Circumcision

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and Venkata R. Jayanthi, MD*

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FAILED ORCHIOPEXY

Orchiopexy for the correction of the undescended testicle is a commonly performed surgical procedure and is generally very successful, implying that a normal-sized testis is located dependently in the scrotum. The overall success of the procedure is dependent on the initial location of the testis and the age of the patient (1). The incidence of persistent cryptorchidism after orchiopexy is estimated to be 7.5–13% (2–4). This can occur because of either residual or recurrent cryptorchidism after the initial orchiopexy attempt. Residual cryptorchidism is recognized immediately at the time of the first surgery as an inability to bring the testis into a dependent scrotal location and requires that a planned second-stage procedure be undertaken at least 6 months later. Recurrent cryptorchidism noted on later follow-up after the initial procedure can be repaired electively at that time. In several studies, the mean time between the initial surgery and reoperation for failed orchiopexy ranged from 3–4.7 years (3,5). After orchiopexy, the size of the testis should be assessed to determine that testicular growth has occurred. Although most testes will grow normally after orchiopexy, some may have growth retardation, atrophy, or may involute altogether. This phenomenon may be caused by compromise of the vascular supply to the testis during the initial orchiopexy attempt or may reflect intrinsic abnormality of a high intraabdominal testis as, for example, in the prune belly syndrome.

Factors Predisposing to Unsuccessful Orchiopexy

The causes for failure of an orchiopexy can include technical factors, the proximal nature of the original location, intrinsic testicular or spermatic cord abnormalities, or prior inguinal surgery. Most frequently, technical factors during the original surgery

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are responsible for orchiopexy failure. Inadequate mobilization of the spermatic cord far enough proximally is the chief factor predisposing to recurrent cryptorchidism (2,4,6,7). High ligation and division of the persistent patent processus vaginalis is a necessary step in most orchiopexies, and when this is not properly completed, a high-riding testis can result. Residual cremasteric or other fascial attachments create upward traction on the cord and may prevent successful scrotal placement of the testis. One study found that 60% of cord length was attributable to division of the patent processus and the remaining 40% was caused by separation of the vessels from the peritoneum and abdominal wall (division of the lateral bands) (8).

After corrective surgery for a hydrocele or hernia, a high-riding testicle may be created if the surgeon fails to replace the testis in a low intrascrotal position (7). This may occur when the testis is pushed back into the hemiscrotum from above, instead of pulling it back down from below by its scrotal and gubernacular attachments. Kaplan reviewed a series of cases of iatrogenic cryptorchidism that occurred after hernia or hydrocele repair; he postulated that excessive retractility of the involved testis with subsequent formation of scar tissue may trap the testis in an elevated position (9). He also noted inappropriate replacement of the testis in the wrong tissue plane as another cause for this outcome.

Injury to the vascular supply of the testis may occur during the dissection to separate the spermatic cord from the processus vaginalis, especially in a very young or premature infant or child (7). Furthermore, excessive tension on the spermatic vessels, which may result from an inadequately mobilized cord, may contribute to poor vascular flow to the testis and subsequent atrophy and/or involution.

The distance that a testicle must be brought down into the scrotum has a bearing on the success of orchiopexy. In a metaanalysis of more than 8000 undescended testes performed by Docimo, the preoperative location of testis position was directly related to the success rate of orchiopexy. The success rate increased as the initial location of the testis was more distal. When all studies were included, the success rates for abdominal, peeping (at the internal ring), canilicular, and beyond the external ring testes were 74, 82, 87, and 92%, respectively. When data before 1985 were excluded, the success rates were 77, 100, 96, and 100%, respectively (1). In the same study, one-stage Fowler-Stephens orchiopexies were successful in 68.5%, whereas two-stage procedures were successful 76.8% of the time.

Diagnosis

In most cases, the diagnosis of a failed orchiopexy can be made on a thorough physical examination by an experienced clinician to determine the position and size of the testis. A failed orchiopexy must be differentiated from a retractile, but otherwise normal, testis, which may require an extended or repeat examination. Rarely, an imaging study such as an ultrasound, computed tomography (CT) scan, or magnetic resonance imaging (MRI) may be helpful to localize a small undescended testis, especially if the patient is obese (7). Measurement of testis size before and after orchiopexy will allow testis atrophy to be distinguished from growth retardation; the former reflects a testis that has become smaller than its preoperative size and the latter represents a testis that has grown but has not kept pace with its mate.

Preoperative Counseling

Prior to undertaking the reoperation for a failed orchidopexy, several issues must be considered. The procedure will be more labor-intensive and challenging than the initial procedure because of fibrosis and scarring present in the surgical field. There is an increased risk of injury to the vas deferens or spermatic vessels for the same reason. Lastly, if the testis is found to be dysplastic or atrophic, then biopsy or orchiectomy may be required. It is of utmost importance that the parents understand these inherent issues prior to surgery so that complete informed consent is possible. It is usually recommended that the reoperation be delayed at least 6 months from the time of the initial procedure in order to allow postoperative edema and fibrosis to stabilize.

Techniques for Redo Orchidopexy

In most cases, although the prior inguinal incision can be used for the redo orchidopexy procedure, adequate exposure is essential and one should not hesitate to extend the length of the incision as needed or make a new incision if the original scar is too low. As mentioned previously, the most common reason for failure of initial orchidopexy is inadequate division of the patent processus vaginalis and/or incomplete proximal dissection of the spermatic cord. The first step is to rectify these deficiencies with careful dissection of the spermatic cord proximally and high ligation and division of a persistent patent processus if present. This alone may provide adequate length to successfully bring the testis down into a dependent scrotal location. The cord structures may be virgin higher up in the inguinal canal if no dissection was performed here previously. In addition, aggressive retroperitoneal dissection and creation of an adequate subdartos pouch has been required in a large number of redo orchidopexies (2).

Opening the floor of the inguinal canal, dividing the inferior epigastric vessels to reroute the spermatic cord medially and straighten its course (the Prentiss maneuver) can all provide additional cord length and occasionally may be required to permit the testis to reach a dependent scrotal location (10). Redman described reoperation for persistent cryptorchidism or hernia via an inguinal approach to the spermatic cord through the cremasteric fascia (11). In this method, the external oblique fascia is incised laterally to medially through the external ring. The spermatic cord structures are then approached caudally, where they may be less scarred than from a standard cranioventral approach. Further proximal dissection is then undertaken, up to and into the retroperitoneum, if necessary. Using this technique, Redman reported 15 successful repeat orchidopexies in 13 patients with no evidence of testicular atrophy.

Cartwright et al. described a method for en-bloc mobilization of the spermatic cord in reoperative orchidopexy whereby a 1- to 2-cm-wide strip of external oblique fascia is left intact on top of the cord structures (12). This maneuver minimizes the risk of injury to cord structures by dissecting lateral and posterior to the spermatic vessels and vas deferens instead of anteriorly beneath the fascia when these structures are the most scarred and adherent. Standard techniques for completing an orchidopexy are then used once en-bloc cord mobilization is completed, including further retroperitoneal dissection and higher ligation of the patent processus. A total of 25 reoperative orchidopexies were accomplished in this manner; one patient needed another procedure to bring the testis down completely and one patient had slight testis atrophy.

Palacio et al. built upon the en-bloc spermatic cord and external oblique fascia dissection technique by suturing a strip of the aponeurosis on the cord to the pubic

bone with nonabsorbable suture. They refer to this as “inguinal cordopexy,” because the proximal cord structures are fixed above the scrotum so that tension on the distal portion of the cord is minimized (13). This technique is not always feasible because the fascial adherence to the cord is sometimes too lax to permit proper fixation or the testis is proximal to the external ring and the fascial strip would descend into the scrotum with the testis. In their study reporting this technique, these authors stated that inguinal cordopexy was possible in 11 testes of 29 patients referred to their institution with malpositioned testes after initial orchiopexy or hernia repair. All testes after this procedure were present in the scrotum almost 2 years later.

The Jones technique for high undescended testicles is another option for reoperative orchiopexy. Jones and Bagley described an abdominal extraperitoneal approach to orchiopexy that allows extensive retroperitoneal dissection of the spermatic vessels and was originally advocated as a single-stage alternative to a staged Fowler-Stephens procedure (14–16). The Jones technique starts with an incision medial to the anterior superior iliac spine with splitting of the anterior abdominal wall muscles and medial reflection of the peritoneum. Once the hernia sac is located and opened, the testis is located within the abdomen. The hernia sac is then ligated and the peritoneum is closed. The spermatic vessels are then dissected proximally in the retroperitoneum until enough length is obtained. A subcutaneous tunnel is then created from the inferior edge of the incision down to the pubic tubercle and the testis is delivered through an incision in the external oblique fascia here. Then the testis is brought down into a subdartos pouch in the scrotum through the standard method.

Multiple reoperative techniques have been described to deal with the failed orchiopexy. To avoid injury to the testis and spermatic cord, we have found that the testis can be palpated and identified early in the procedure by initiating dissection inferior to the external inguinal ring. En-bloc dissection of the testis and spermatic cord complex in a cephalad direction allows scar tissue and fibrosis to be separated from the spermatic vascular supply and vas until the region of the internal ring has been reached.

Testicular autotransplantation has been described as an alternative for Fowler-Stephens orchiopexy in high intraabdominal testes. Using microvascular surgical techniques, the spermatic vessels of the testis are anastomosed to the inferior epigastric vessels and the testis is placed into the scrotum. Silber and Kelly(17) first described this procedure in 1976, but it has not gained widespread popularity owing to the microsurgical skill and instrumentation required.

Testis atrophy is a known complication of repeat orchiopexy and is reported in approximately 4% of cases. Pesce et al. looked at the long-term follow-up after reoperative orchiopexy and found that, compared to controls, the volumes of the majority of reoperated testes were smaller but fertility was only markedly affected in 15% of 20 patients assessed (18).

COMPLICATIONS OF CIRCUMCISION

Circumcision is the most commonly performed surgical procedure in the United States and is probably one of the oldest of all surgical procedures (19–22). Data is not available to estimate the total number of circumcisions done presently because many performed are ritual circumcisions outside of a healthcare setting. Therefore, the true rate of complications after circumcision cannot be precisely calculated. Major

complications from circumcision are very rare and minor complications are estimated to be as low as 0.2% to as high as 6% (19–21,23,24).

Methods

Circumcision can be performed in multiple different ways, either with various specially designed clamp devices, a Plastibell ring, or by open surgery. Neonatal circumcisions are performed with a clamp device or with the Plastibell (Fig. 1), usually with a local penile block as the method of anesthesia. The commonly used clamp devices are the GOMCO clamp, the Mogen clamp, and less commonly, the Sheldon clamp (Figs. 2 and 3). Each technique has been associated with various types of complications, which will be discussed in the next section.

Complications

Death after circumcision has been reported in the literature (25) in isolated case report, but this is extremely uncommon. Because circumcision is a surgical procedure, it is not surprising that hemorrhage is the most common complication and is estimated to occur in approximately 1% of cases (26). When present, bleeding is frequently seen in the area of the frenulum or at the cut skin edges. Treatment consists of first applying local pressure, which in the newborn is often all that is required, but more definitive treatment may be necessary, such as silver nitrate application, thrombin foam, electrocautery, or suture ligation.

Infection is the second most common complication seen and is estimated to occur in 0.2–0.4% of cases (26). Prophylactic antibiotics for neonatal circumcision are rarely used, although use of a topical antibiotic for a short time after the procedure is not uncommon. If a local skin infection does occur, routine wound care and oral antibiotics will usually provide resolution in a short period of time. Less commonly, intravenous antibiotics may be necessary. Bliss et al. reported two cases of necrotizing fasciitis in newborns after circumcision with Plastibell (27). These cases required broad-spectrum antibiotics, wide surgical debridement of involved tissues, and skin grafting.

Redundant foreskin or even secondary phimosis after circumcision is a frequent reason for referral to a surgical specialist (Fig. 4). These complications are often caused by inadequate removal of sufficient skin at the time of the initial procedure. Often, the redundant skin is mostly a cosmetic issue, but a secondary phimosis owing to a tight cicatrix can predispose to infection caused by entrapped urine or can cause pain with erections (28) (Fig. 5). If the child has a prominent suprapubic fat pad or poor fixation of the penile shaft at the penopubic junction, a hidden/buried/concealed penis may result after circumcision (Fig. 6). This entity may resolve spontaneously as the

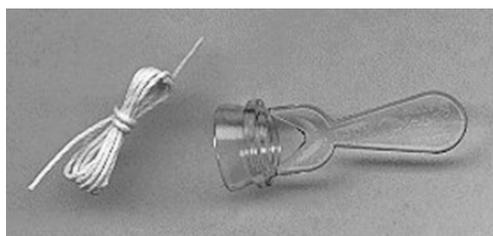


Fig. 1. Plastibell circumcision device.



Fig. 2. GOMCO clamp.



Fig. 3. Mogen clamp.



Fig. 4. Redundant foreskin in a child who has previously undergone neonatal circumcision.



Fig. 5. Secondary phimosis caused by a tight cicatrix, which formed subsequent to neonatal circumcision.



Fig. 6. Buried (hidden) penis after circumcision.

child ages and body habitus changes, but persistent penile burying may require some form of penoplasty to correct poor fixation of the penile shaft to the penopubic fascia (29) (Fig. 7).

Adhesions of the shaft skin to the glans are fairly common after circumcision and are usually caused by inadequate genital hygiene. Mild adhesions caused by entrapped smegma can often be separated manually in the office or can be treated medically with a several-week course of a steroid ointment (30). When penile adhesions epithialize and become more dense, these skin bridges do not respond to topical steroids and often require surgical lysis (Fig. 8). One study suggests that skin bridges are more likely to form when the adhesions involve the circumcision line (31).



Fig. 7. Appearance of penis immediately after penoplasty to correct buried penis.

Inclusion cysts at the circumcision line have been described (19,20,26). These are epidermoid cysts and are caused by entrapment of smegma in the circumcision wound or inadvertent rolling in of the epidermis during the initial procedure. These cysts can grow to a large size and/or become infected (Fig. 9). Treatment consists of surgical excision.

Inadvertent injury to the urethra at the time of circumcision is a rare complication and has been reported with a resulting fistula occurring either early or late after the procedure (19,20,32). Urethral injury may result from overzealous treatment of

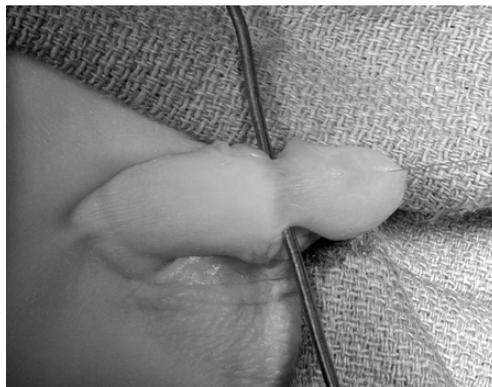


Fig. 8. Penile skin bridge on the dorsal aspect of the penis: a probe can be seen being passed beneath the shaft skin, which is adherent to the glans.



Fig. 9. Penile inclusion cyst.

bleeding at the frenulum with either suturing or cauterization (26). These fistulas tend to become apparent early on after circumcision because of ischemic injury of the urethra; however, one case report noted a urethral fistula in association with a penile skin bridge 13 years after neonatal circumcision (33). This fistula was felt to be subclinical until puberty, when penile growth and more frequent erections may have caused its unmasking. The urethra can be accidentally incised during a dorsal slit if the penis has a 180° torsion and the glans is not appropriately visualized prior to circumcision (34). Surgical repair of urethral fistula must be individualized to each patient, but similar principles to that of hypospadias fistula repair apply. Formal urethroplasty should be delayed at least 6–9 months after initial fistula formation to allow tissue inflammation and fibrosis to subside.

Partial or complete glans amputation is a very serious complication and is usually seen when a clamp is used for circumcision. This has been reported with a freehand procedure as well as with Sheldon, Mogen, and GOMCO clamps (23,35–38) (Fig. 10). One case report in an older child noted an entire glans penis amputated during a guillotine-type procedure (39). If minimal partial glans amputation has occurred without urethral involvement, local wound care and hemostasis may be adequate because this may granulate and heal with a satisfactory cosmetic result. If more serious glans amputation with urethral involvement is found, immediate reanastomosis of the amputated glans and urethra after debridement of the wound base is required. Good results have been reported with glans reattachment surgery, although tissue loss, fistula, and/or meatal stenosis can result. Penile injury and glans necrosis have also occurred when diathermy was used to remove skin during metal clamp circumcision and the current flows to the glans.

Meatitis or meatal stenosis occurs in 8–31% of circumcised boys, and is rarely seen in the uncircumcised (26). Inflammation and potential stenosis of the meatus are felt to occur because of inflammation from exposure to urine in the diaper. Meatitis can be treated conservatively with observation or a brief course of topical antibiotics or steroids. Symptomatic meatal stenosis (Fig. 11) is corrected in an office setting with a meatotomy after a local anesthetic block or with a meatoplasty under general anesthesia in older children or those in whom adequate local anesthesia or immobility cannot be achieved.

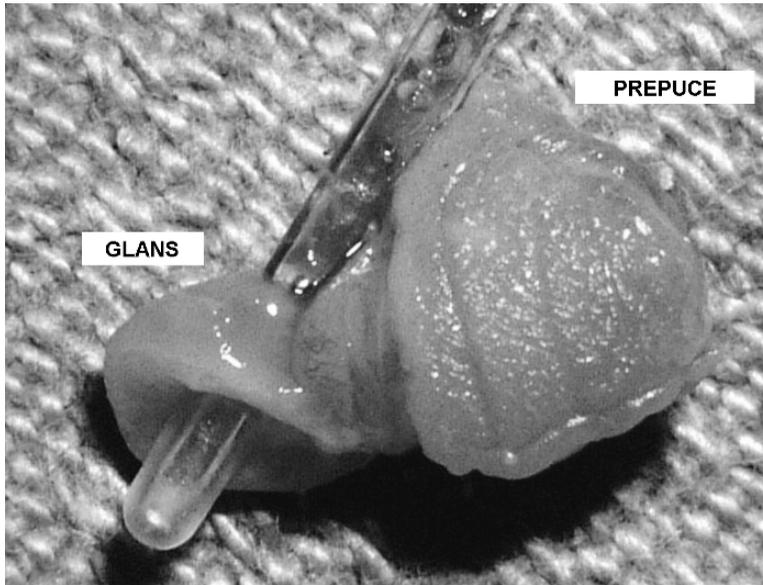


Fig. 10. Glans amputation after circumcision.

One complication specific to the Plastibell ring is dislocation onto the penile shaft proximal to the glans such that the glans becomes entrapped and compressed (40). This is believed to occur when too large a ring is used, so that the device can slide back over the glans. This situation causes compression of the penile shaft proximal to the glans and subsequent glans edema similar to that seen in paraphimosis. The ring requires removal by cutting the ring away from the underlying skin with a ring- or wire-cutting instrument, followed by local wound care (Fig. 12). Another rare complication caused by proximal Plastibell ring migration is urinary retention owing to edema and compression of the urethra (41).

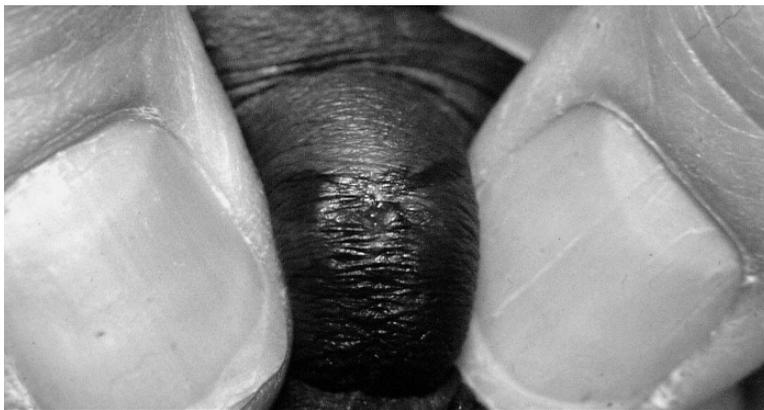


Fig. 11. Meatal stenosis: the urethral meatus is pinpoint in size and can cause irritative or obstructive voiding symptoms.

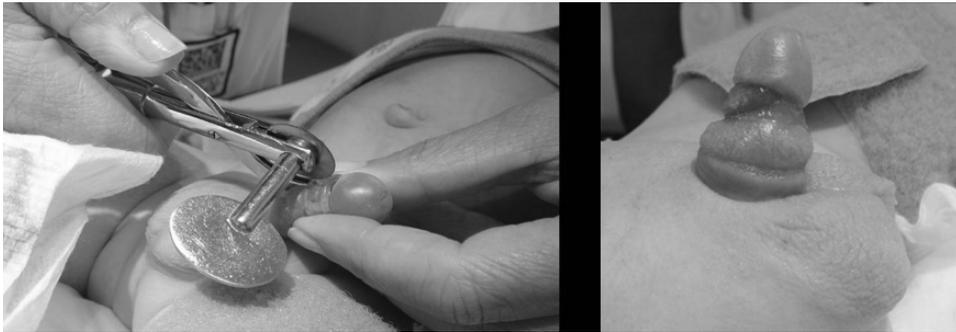


Fig. 12. Proximal migration of Plastibell circumcision device before (A) and after removal (B).

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Complications of Vaginoplasty and Clitoroplasty

Lesley L. Breech, MD

CONTENTS

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Pelvic reconstructive surgery in a pediatric population is primarily undertaken in the treatment of congenital anomalies or congenital conditions. Pelvic reconstruction is performed in patients with conditions such as intersex conditions, complex urogenital anomalies, or isolated reproductive anomalies. Preoperative recognition of potential surgical challenges is crucial. This chapter describes potential complications associated with genital reconstructive surgery and consideration of possible causes.

Surgical goals in pelvic reconstructive surgery are the establishment of an anatomically appropriate, physiologically sensitive clitoris; an adequate, appropriately placed vagina for comfortable sexual relations in adulthood; and normal-appearing female external genitalia. The major reconstructive procedures include feminizing genitoplasty and vaginoplasty. Both procedures may be performed during the same operative procedure; however, because the postoperative complications are significantly different, each will be addressed independently. As will be emphasized, it is also important to consider the indication for surgical intervention. Certainly optimal function and normalization of anatomic appearance are of primary importance, but the underlying medical condition and the possible surgical and postoperative ramifications are necessary to consider. In patients with the genital effects of androgen excess, intraoperative dissection and postoperative response may be more challenging because of the role of hormonal stimulation. Surgical manipulations in conditions without hormonal influence are more likely to proceed more smoothly, with less risk of postoperative bleeding complications.

FEMINIZING GENITOPLASTY

Feminizing genitoplasty as a surgical intervention has evolved significantly over recent years. Much discussion has occurred relating to the appropriate timing and indications, in addition to the appropriate procedure. Surgical intervention for

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clitoromegaly, as a component of genital ambiguity generally associated with intersex conditions, has been discussed and reviewed owing to concerns regarding long-term patient outcome. Unfortunately, much of the potential controversy relates to the more vocal experiences of some patients. Public concern has influenced medical standards such that some providers and families contemplate deferring surgical manipulations until the patient is able to make her own operative decision. This premise, while desirous in theory, is likely not realistic for most families. In the majority of cases, it is the child's parents who request surgical management of an enlarged clitoral structure. Often parents are less cognizant of the possible existence of a persistent urogenital sinus, but are acutely aware of clitoromegaly. Discomfort with genital appearance inconsistent with chromosomal sex is the primary reason for early intervention in girls with female pseudohermaphroditism. In cases of extreme virilization, it is understandable why families or surgeons push for early surgical management. However, consideration of possible variation in outcome should also factor in operative decision-making as well as in discussions with family.

Postoperative issues after surgery at the clitoris include nerve damage, sensation, pain, recurrent clitoromegaly caused by persistence of erectile tissue, body image concerns, and sexual function. Late outcomes following feminizing genitoplasty have not been systematically reviewed to allow obvious pursuit or abandonment of individual procedures. Consideration of the type of procedure performed and the combination of intervention at the vagina for a single-stage procedure is essential.

The evolution of clitoral reconstruction began with clitorectomy, followed by clitoral recession, and now most commonly, clitoral reduction. Clitorectomy results in the obvious loss of sensation and may jeopardize the ability of an orgasmic response. Clitoral recession maintains sensation; however, pain secondary to clitoral engorgement is a definite risk. Without adequate removal of erectile tissue, adolescent young women may experience pain, discomfort, and extreme enlargement of the clitoral area owing to stimulation and engorgement of residual corporal tissue (*see Fig. 1*) (1). Recognition of potential sensory consequences that may affect pubertal adolescents or young adult women are necessary to consider, despite the reality that most girls will undergo clitoral surgery as an infant or young child. The most appropriate procedure for feminizing genitoplasty is based on the concept of maintaining the clitoral glans and sensory input, which facilitates orgasm (2–4). The technique involves resection of the corpora at the crura, with careful preservation of dorsal nerves and vessels and ventral mucosa that supplies the glans (5). Management of a persistent urogenital sinus and possible vaginoplasty are the next consideration in the management plan of girls with genital ambiguity. Several techniques have been described, primarily predicated on the level of confluence between the vagina and the urethra (6–8). Historically, a low vaginal insertion has been managed by a simple cut back incision or a posterior flap vaginoplasty (9,10). The use of this technique when the confluence is higher is inappropriate because it leaves the urethral meatus on the anterior vaginal wall, resulting in vaginal voiding and difficult access to the meatus (*see Fig. 2*). A high vaginal insertion requires more elaborate operative intervention, likely performed as a two-stage procedure. A pull-through vaginoplasty (11), intestinal neovaginal segment (12), perineal skin flaps (7), total urogenital sinus mobilization (TUM) (13), or posterior sagittal transanorectal approach (14) have all been proposed to provide adequate urinary and reproductive function. The intimate relationship between the vagina and the urogenital sinus mandates meticulous dissection of the vagina from the urethra

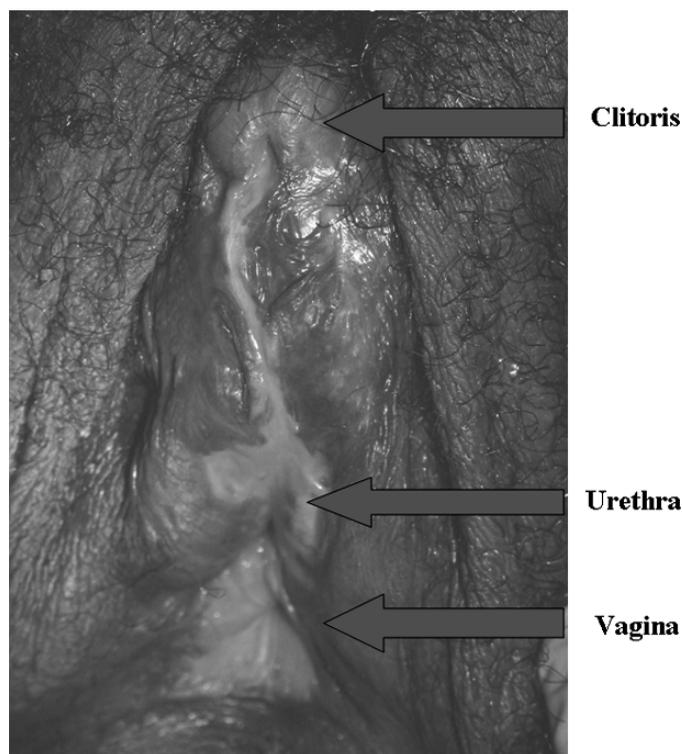


Fig. 1. A 15-year-old female with a history of CAH who underwent a single-stage feminizing genitoplasty-reduction clitoroplasty and a pull-through vaginoplasty at 2 years of age. She presented with pain and enlargement of the clitoral region.

proximal to the confluence of the sinus. Some authors postulate that fibrosis in this field of dissection may play a pivotal role in the development of vaginal stenosis (*see* Fig. 3) (7,15). Procedures that rely upon the use of the distal (fibrotic) vaginal portion of the urogenital sinus may increase the risk of the development of stenosis or fistula formation.

Several reviews have provided insight into both short- and long-term outcomes in this population. Eroglu and colleagues in Turkey reported on the results of 55 intersex patients who underwent feminizing surgical management (16). They reported the outcomes of patients managed over a 16-year period with multiple procedures including single-stage clitorovaginoplasty (29 patients), staging clitoral surgery and vaginoplasty (seven patients), and TUM (three patients). The mean age at surgery was 3.5 years and the follow-up averaged 4.1 years (2 months to 17 years). The most frequently encountered complication was vaginal stenosis; however, less vaginal stenosis was found in patients who underwent an early single-stage procedure (3.4%, 1/29) than in those with a staged procedure (42.8%, 3/7). Comparison with TUM in this study was impossible, as all patients were too young with a short postprocedure interval. Bocciardi and colleagues reported a large series of Passerini-Glazel clitorovaginoplasty procedures in 66 children, of whom 46 were available for extended follow-up (17). Outcome regarding anatomical results of the vaginoplasty and the cosmetic effect of the entire procedure were described. All of the patients had good cosmetic outcomes. None of the patients had introital stenosis. The patients were divided relative to age at

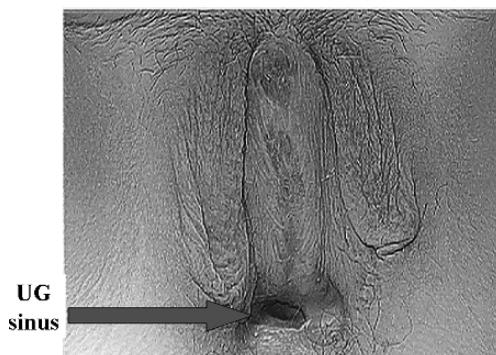


Fig. 2. A 12-year-old female with CAH s/p reduction clitoroplasty at age 8 months followed by a high pull-through vaginoplasty at age 2 years. Of note, a persistent urogenital is present. Because the patient is experiencing pain and discomfort at the clitoris, she will undergo a repeat clitoroplasty and TUM.

surgical intervention. Of the prepubertal group, 45.5%, and of the postpubertal group, 25% had vaginal stenosis at the anastomosis of the mucocutaneous cylinder and the true vagina. All stenoses were corrected by Y-V plasties. The authors proposed that the vaginal stenosis was related to persistence of dysplastic or ischemic tissue at the distal edge of the vagina. They recommend aggressive removal of the distal vagina until soft, thick, highly vascularized native vagina is identified to avoid such stenosis.



Fig. 3. Photo of a 21.5-year-old female with CAH treated with a single-stage procedure at age 2 with a reduction clitoroplasty and vaginal pull-through. The confluence of the urethra and vagina was 2 cm into the urogenital sinus. The patient currently complains of the inability to engage in vaginal intercourse because of vaginal stenosis. She did not previously perform vaginal dilatation.

Other reviewers dismiss this process and instead emphasize the detrimental role of a circular anastomosis in constrictive healing and the development of stenosis (18). Krege and colleagues reported 27 females who underwent reconstructive surgery owing to congenital adrenal hyperplasia (CAH), with a special emphasis on the long-term results of vaginoplasty (19). Vaginoplasty was performed in 25 girls, of whom 20 underwent a single-stage approach and five had a two-stage procedure. Vaginoplasty was performed using the technique described by Fortunoff and others (9), using a perineal flap sutured to the margin of the vaginal introitus; one patient required a pull-through vaginoplasty, described by Hendren and Crawford (10). During long-term follow-up, nine of the 25 (36%) had secondary vaginal stenosis. The mean age of diagnosis of the stenosis was 14 years (10–19 years). Stenosis was treated with laser surgery, Z-plasty, or a pedicled skin-fat island flap. Lack of flap advancement far enough into the vagina, lack of precise coaptation of the mucosa to the perineal skin flap, inadequate blood supply to the flap, inadequate marsupialization of the vaginal floor, and imprecise suture placement have all been reported causes of vaginal stenosis (20,21). TUM was initially described by Peña to repair cloacas (22). This procedure has been applied to children with urogenital sinus anomalies, including CAH (22,23). The most remarkable advantage of this procedure is the avoidance of the difficult dissection required for separation of the vagina from the urinary tract by treating the urethra and vagina as a single unit. Additionally, the mobilized portion of the urogenital sinus may be split and placed anteriorly to create a mucosal surface or an additional flap. Other advantages are reported to be a decrease in operating time, decreased risk of vaginal stenosis, no risk of fistula formation because there is no need for separation of the structures within the urogenital sinus, less urine pooling within the vagina, the absence of postvoid dribbling, and the lack of skin flaps to reach the vagina (22,23). The main concern of TUM is the theoretical risk of stress urinary incontinence caused by the short and distally placed urethra (24). Jenak and colleagues report on six patients who were treated with a modification of the TUM and did not find any changes in voiding habits postoperatively in patients who were toilet-trained before surgery; however, the follow-up period was less than 1 year. Farkas and colleagues reported a follow-up period of 4.7 ± 2.6 years for a group of 49 patients who underwent a single-stage feminizing genitoplasty (25). Of the 49 patients, 46 were treated with an en bloc mobilization of the vagina and urethra via the perineal approach. All patients were reported to have successful cosmetic and early functional results. In patients who reached puberty, the authors reported normal menstruation, a wet and wide introitus, and no evidence of fibrosis or scarring of the perineum. No urinary tract infections were reported and all patients were continent. No patients with CAH had initiated vaginal intercourse. Several reports confirm the technical ease of the TUM, especially in cases of a urogenital sinus less than 3 cm in length and improved cosmetic results (13,16,25). Longer-term follow-up will be necessary to confirm positive early results, because most patients will now be entering puberty and adolescence.

Possible complications after feminizing genital reconstruction include vaginal stenosis, meatal stenosis, vaginourethral fistula, female hypospadias, urinary tract injuries, and recurrent clitoromegaly (especially in patients with difficulties in compliance with hormonal regimens). Limited surgical and long-term outcome data are currently available. Al-Bassam and Gado reported the development of vaginal stenosis in 16.2% patients who underwent either flap vaginoplasty or vaginal pull-through (26). Krege and colleagues reported 27 females who underwent reconstructive surgery owing

to CAH, with a special emphasis on the long-term results of vaginoplasty (19). Vaginoplasty was performed in 25 girls, of whom 20 underwent a single-stage approach and five patients had a two-stage procedure. During follow-up, all patients were asked to complete a questionnaire. Questions regarded appearance of the external genitalia, overall body image, frequency of heterosexual intercourse or other forms of sexual activity, dyspareunia, and ability to achieve orgasm. Nine patients were noted to have vaginal stenosis and of those nine patients, six completed the questionnaire. Only one patient reported regular sexual intercourse. Of the remaining 18 patients, ten completed the questionnaire. Four of nine patients 15 years and older reported sexual intercourse and seven reported having orgasm (including those having vaginal intercourse). Few studies have been performed to assess the sexual and social functions of patients in late adolescence or adulthood (19,25,27,28).

The question of timing for feminizing genital reconstruction can be a difficult one. In conditions such as CAH and other intersex conditions, clitoral surgery and vaginoplasty are generally performed in infancy and childhood. Most authors recommend timing between newborn to age 3 years. The prevailing opinion is that clitoroplasty should be performed as early as possible; however, the timing of vaginoplasty remains controversial. The primary reason described for intervention before age 2 years is the potential detrimental psychological effects for both children and parents (19,29). Other surgeons defer surgery until puberty, if possible, to better differentiate the demarcation between hair bearing and non-hair bearing skin and to potentially avoid possible vaginal dilatation in childhood (19,29); however, bleeding complications can be more challenging in the pubertal patient. Krege and colleagues argue for vaginoplasty to be performed at puberty in patients with a longer confluence of the urethra and vagina, who may potentially need vaginal dilatation (19). They also note the potential advantage of a distensible vagina provided by estrogen stimulation. Conversely, Lobe and others reported a decreasing complication rate in patients undergoing an early, single-stage procedure (30). Although successful results from neonatal one-stage surgery have been reported (31), many still prefer to perform the single stage clitorovaginoplasty procedure sometime before 3 years of age, citing better compliance with treatment and alleviating parent's concerns (26,27).

VAGINOPLASTY

Vaginoplasty is a procedure relevant to the care of female patients of any age. Physicians may care for children with complex pelvic congenital anomalies or adolescents with vaginal agenesis. Vaginal agenesis is an uncommon condition, which may result from a variety of different underlying diagnoses. Mayer-Rokitansky-Kuster-Hauser (MRKH) Syndrome, the most common clinical scenario the general provider will encounter, describes vaginal agenesis with variable Müllerian duct development and possible associated renal, skeletal, and auditory abnormalities. It is generally thought that MRKH affects 1/5000 live-born females (32).

The timing of intervention is an important consideration. Vaginal agenesis is a particularly challenging problem for genitourinary surgeons. In patients diagnosed with vaginal agenesis as a child with additional organ system involvement, life-threatening conditions should be stabilized and addressed before operative planning is undertaken for neovaginal creation. Nonsurgical and surgical interventions that require continued dilation and maintenance have historically been deferred to adolescence or adulthood.

Reconstruction with the use of a bowel graft, as it often does not require continued dilation, can be accomplished at any age. Hendren and others have advocated for intervention during childhood (33). Hendren maintains that young girls suffer significant psychological and social trauma from the knowledge of an anatomically absent vagina and from the necessary surgical intervention. He stresses that he has not seen a traumatic response to surgical revision of a previous vaginoplasty created as a child. He emphasizes the outpatient nature of a minor revision and that it is regarded by patients and families with less importance than original construction of a neovagina. This must be weighed against the psychological effects of potential daily vaginal dilation or stenting in prepubertal girls. Grafting or flap techniques that require persistent stenting or dilation may be unsuitable for younger patients because of the requisite maturity and motivation required for maintenance and the avoidance of postoperative complications. Bowel substitutions may be applied to wider ranges of patients, even infants. Long-term functional studies are needed to assess introital size and sexual satisfaction with consideration of timing of surgical intervention. An evaluation of the psychological outcomes related to technique, the age at diagnosis, and age of repair is also necessary.

Over time, many methods of surgical vaginoplasty have not only been described but also advocated. Even today, there is no consensus regarding the best surgical approach. The primary goal of a neovaginoplasty is to create a vagina adequate for satisfying sexual interactions. In patients with a uterus, adequate egress of menses and the potential to accommodate a vaginal delivery are also of paramount importance. Additional goals should also include normal amounts of secretions/lubrication, the absence of malodor, and minimal maintenance care. Unfortunately, one method has not yet been demonstrated to achieve all of these goals. Management options are described addressing procedure types, advantages, disadvantages, and outcomes. Controversy continues regarding the best technique, as little complete long-term data, including sexual identity and function data, about any procedure is truly available.

A well-established technique for vaginal reconstruction is the use of intestinal segment substitution. Baldwin initially reported the use of ileum in 1904 and 1907 followed by Wallace, who described the use of sigmoid colon in 1911 (34–36). Wesley and Coran modified Baldwin's technique for the sigmoid colon (37). Pratt further popularized the technique with sigmoid colon (38). The purported benefits of using intestinal segments include the lack of obligatory continued dilation or stenting, the potential for growth in small children, and beneficial lubrication. Rajimwale and associates published a metaanalysis of seven recent series of neovaginal creation with bowel (33,37,39–43). Patient ages ranged from 4 days to 26 years. Bowel vaginoplasty demonstrated a complication rate of 35%, with a reoperation rate of 4% in 202 cases. The authors argue that the postoperative complications after bowel vaginoplasty may be managed more simply, using dilation or minor surgical repairs, than those associated with skin grafting techniques. They also note that the use of bowel transposition is broader, as it can be employed in a wider age range and for a variety of diagnoses compared to skin grafting techniques, which are primarily described in the more isolated anomaly of MRKH.

Several segments of intestinal vaginoplasty replacement have been described: ileum, cecum, sigmoid, and rectosigmoid. Selection may relate to the availability of a suitable segment of an appropriate length; patients also undergoing bladder augmentation or other more complex anorectal or urogenital repairs may have any combination of small or large bowel for reconstruction, dependent on the needs of other reconstruction.

Ileal or cecal segments may be problematic because of a shorter mesentery, which may produce tension on the neovagina; Hensle and Reiley reported a higher incidence of stenosis attributed to this concern (39). In complex anomaly cases, the ileum may be the only bowel segment available for use for vaginal construction. The rectum and sigmoid may be more optimally used in other urinary or bowel reconstruction. It is important to be cognizant of the role of the ileocecal valve within the gastrointestinal tract and the subsequent morbidity associated with any disruption. Pratt reported a higher incidence of dyspareunia and persistent mucus secretions with ileal vaginoplasty (38).

Sigmoid vaginoplasty has been the favored bowel segment due to its potential advantages: adequate length, natural lubrication, and early coitus in older patients. The sigmoid neovagina is thought to be self-lubricating (*see* Fig. 4), without the excess mucus production associated with segments of small bowel (39). The risk of stenosis is small, without the requirement of continuous dilation or stenting. Mobilization of the vascular pedicle is usually straightforward and has been accomplished using a minimally invasive, laparoscopic approach (44,45). When replacement is performed during childhood, the potential for adequate growth into adulthood remains unclear. Koyle and colleagues describe the sigmoid neovagina as aesthetically pleasing and more compatible with sexual activity (46). The thickness of the sigmoid tissue may withstand trauma associated with sexual intercourse better than small bowel or skin grafts. Freundt and colleagues reported on psychosexual and psychosocial performance in adult patients with a sigmoid neovagina (47). The series included eight patients with vaginal agenesis and one patient with androgen insensitivity. Psychosexual profiles, as determined by patient questionnaire, indicated that most of these patients achieved acceptable outcomes with respect to sexual relations, function, and satisfaction. The validity of the evaluation is difficult to interpret as the same questionnaire was used in transsexual patients. Communal and colleagues described the sexual outcomes of 12 patients with MRKH syndrome who underwent sigmoid neocolpopoiesis using a standardized questionnaire, the Female Sexual Function Index (FSFI), a validated assessment of female quality of life and sexual function in adults (48). Six separate domains of female sexual function are included in the questionnaire: desire disorder, arousal disorder, orgasmic disorder, sexual pain disorder, quality of vaginal lubrication, and sexual/relationship satisfaction. The authors also added details on sexual inter-



Fig. 4. Perineal photograph taken intraoperatively immediately after creation of the sigmoid neovagina in a 16-year-old female with MRKH syndrome.

course, vaginal discharge, self-esteem, and anxiety. The authors reported patients had a normal sexual life. A total of 72% of patients had vaginal intercourse at least once weekly; however, 18% had severe dyspareunia. Vaginal discharge was described as almost constant, but the daily discomfort score was “moderate.” Patients who were “dry” preoperatively later reported wearing one to two pads daily. Consideration of development of colitis in the graft, which may play an integral role in the development of persistent vaginal discharge, is essential (49,50). In severe cases of colitis, pain, irritation, and bleeding may occur, necessitating vaginoscopy. Stenosis of the introitus may occur related to excessive traction on the sigmoid transposition, or limited perineal cleavage may necessitate dilation or additional surgery.

More recent publications by both Hensle and Kapoor confirm the successful outcomes in patients treated with a sigmoid vaginoplasty (51,52). Hensle and colleagues reviewed the records of 57 patients who underwent bowel replacement vaginoplasty over a 24-year period. Replacement vaginoplasty was performed using sigmoid colon in 39 patients. The authors evaluated the postoperative quality of life, with a special emphasis on sexual function. Postoperative sexual function was evaluated in 44 patients; however, only 36 completed the sexual function questionnaire. Of the 36 patients who responded, 78% reported sexual desire, 33% sexual arousal, 33% sexual confidence, and 78% sexual satisfaction. A group of 20 patients (56%) reported frequent orgasms and eight (22%) reported occasional orgasms. Only 22% denied having orgasm. A total of 32 patients (89%) reported adequate lubrication for intercourse, and two reported dyspareunia. A group of 34 patients used home douching, and 20 (56%) required pads for mucus production.

Sigmoid neocolporrhaphy has historically involved a major laparotomy and the associated risks of bowel surgery, including perforation. In patients with more isolated vaginal anomalies, sigmoid neovaginoplasty offers the potential of a laparoscopic-perineal approach to decrease operative morbidity and adverse aesthetic consequences associated with a laparotomy. Darai and others conducted a preliminary study with seven patients with MRKH syndrome, confirming the feasibility of the laparoscopic approach when performed by surgeons with experience in both gynecological and gastrointestinal laparoscopic surgery (53). The mean operating time was 312 minutes (range 220–450 minutes). No intraoperative complications were reported, and conversion to laparotomy was never necessary. The mean fall in hemoglobin was 3.6 g/dL (range 2.0–4.4 g/dL). The mean hospital stay was 7.7 days (range 6–12 days). The mean length of the neovagina was 11.5 cm without documented shrinkage over the initial 6-month follow-up. Mean follow-up was 31 months (range 5–90 months). The mean interval between surgery and first intercourse was 4 months (range 3–6 months). Despite promising results in this small population, randomized and/or multicenter trials are necessary to confirm the advantages.

Possible advantages of the use of sigmoid colon, as in other bowel neovaginoplasty, are the avoidance of long-term stenting or dilation and the rare contraction of the neovagina. Potential disadvantages include the complexity of the surgical procedure, generally requiring laparotomy or extensive laparoscopic skills, significant mucous discharge reported by patients, and postoperative complications. Periumbilical pain with vaginal intercourse, attributed to traumatic stretching of the graft vascularity, and problems with defecation (constipation) are not uncommon.

An alternative technique familiar to many gynecologic surgeons is the Abbe-McIndoe vaginoplasty (54). The procedure utilizes a split-thickness skin graft to line

a neovaginal canal, dissected in the potential space between the urethra and rectum. The three most important principles in performing this procedure are dissection of an adequate space, inlay of a proper split-thickness skin graft, and continuous, prolonged dilation during the contractile phase of healing. Many consider this technique unsuitable for children, as it requires strict adherence to dilation until regular intercourse occurs. In addition, it is unclear how well this type of neovagina will grow with the child. It has successfully been reported in patients with vaginal agenesis who undergo the procedure as late adolescents or young adults. Templeman and colleagues published a review of the procedure and included the results of multiple surgeons (55–61). Success rates vary from 80–100%, depending on the experience of the surgeon. Klingele and colleagues reported on 71 patients who responded to a questionnaire after a McIndoe procedure (62). Of the 71 patients, 10 (14%) had significant complications, including vaginal prolapse, and rectovaginal and vesicovaginal fistula. In this series, 56% of patients reported improvement in self-image, 83% reported satisfactory vaginal function during intercourse, and 75% were able to achieve orgasm. Only 28% of patients had a disfiguring graft harvest site. Relevant complications to consider for this procedure include failure of graft take, rectal perforation during dissection, and fistula formation.

The advantages of the Abbe-McIndoe procedure include the long-term experience with excellent published results, low complication rates, and the avoidance of a laparotomy and intestinal complications. The disadvantages should include prolonged postoperative hospitalization, the necessity for continued dilation, and the visible skin scar from acquisition of the skin graft. Some authors also report difficulties with lubrication and dyspareunia (39).

Several authors have described a similar approach as the Abbe-McIndoe; however, they have utilized synthetic materials to wrap around the vaginal mold and line the neovaginal space. Jackson and colleagues originally reported the use of oxidized regenerated cellulose fabric (Interceed™) after a segment of amnion was inadvertently contaminated during a surgical neovaginoplasty (63). Moyotama and associates described 10 patients who were diagnosed with vaginal agenesis and treated with oxidative regenerated cellulose fabric (Interceed) (64). Operative time (<30 minutes) and postoperative hospitalization (2 days) were significantly shorter than that for the traditional skin graft method. Vaginal depth was 8–10 cm, and no significant operative complications were reported. Patients had only been followed for 6 months at the time of publication; however, vaginal intercourse was already described as easy and successful.

Benefits of this intervention include the short operative time and hospitalization. The procedure is technically easier without acquisition of a skin graft. No scarring, as occurs in a skin graft procedure, is visible to others. Although short follow-up is reported, patients have early satisfactory sexual interactions. As in the skin graft procedure, continued postoperative dilation is necessary. It is essential to provide additional long-term follow-up data regarding this procedure to truly assess its role in patient care.

Skin grafts provide a thin lining for the neovaginal cavity, which does not alter the length and diameter of the dissected neovaginal cavity during the operation. Split-thickness skin grafts have been used most often for adolescents being treated by gynecologists, and long-term follow-up has demonstrated a patent and functional vagina. The split skin segment retains most of its original morphological and histochemical characteristics, but atrophy of sweat glands and hair follicle has been

observed (65). The main concern is possible inadequate take of the graft leading to a reduction in the size of the neovaginal cavity during the contractile phase of healing (66,67). Patients are required to wear a vaginal stent continuously for many months to prevent contracture and stenosis. Full-thickness skin grafts may have a decreased tendency to contract than split-thickness grafts during the contractile phase of healing. Additionally, full-thickness grafts may successfully grow in children (68). Reduced stenosis without consistent dilation or intercourse may be especially beneficial in patients in whom vaginal construction is performed at an earlier age (66,67). It also has some benefit even in select adolescents who may experience more significant psychological distress with dilation. Because sebaceous and sweat glands are better preserved in full-thickness grafts, secretions produce vaginal lubrication within 4–5 months postoperatively (68). Although the full-thickness graft method may be acceptable for neocolporrhaphy, most pediatric surgeons would have less expertise and require plastic surgery collaboration for optimal results.

The Vecchiatti operation was first described in 1965 by Giuseppe Vecchiatti (69). Veronikus and colleagues reviewed the use of the technique and described a laparoscopic modification, which combines cystoscopy to confirm bladder integrity (70). The Vecchiatti procedure is a surgical technique that constructs a dilatation-type neovagina in 7–9 days. The procedure utilizes specialized equipment including a traction device, a ligature carrier, and an olive-shaped acrylic vaginal dilator. The process involves two steps with essential operative and postoperative components. The operative phase involves positioning the olive at the perineum, and the traction sutures are positioned extraperitoneally. Classically performed through a Pfannenstiel incision, although may be approached laparoscopically, the ligature carrier introduces the suture into a newly dissected vesicorectal space. During the postoperative invagination phase, the neovagina is created by applying constant traction to the olive. The process reportedly occurs at a rate of 1.0–1.5 cm/day, developing a 10–12 cm vagina in 7–9 days.

The Vecchiatti procedure is an interesting procedure that applies the principles of progressive dilation against the vaginal dimple in an innovative fashion. Notable benefits are the avoidance of vesicorectal dissection and postprocedure visible scarring, in addition to the opportunity for a laparoscopic approach. Functional success rates vary from 98–100% (71,72). Disadvantages include poor lubrication, long-term dilation in patients who are not sexually active, and a significant risk of neovaginal prolapse.

Davydov described a three-stage surgical creation of a neovagina in 1969 (73). The procedure involves dissection of the rectovesical space, abdominal mobilization of the peritoneum followed by attachment of the peritoneum to the introitus. Laparoscopic modifications to perform the rectovesical dissection and the purse-string closure of the abdominal neovagina have been described. The most common postoperative problem involved the formation of granulation tissue at the vaginal vault. As with any open abdominal procedure, laparoscopic conversion has allowed shorter hospital stays, decreased morbidity, and a substantial decrease in visible scarring. However, the technically challenging components of the laparoscopic approach may limit the use of this interesting technique. Additional follow-up information regarding this technique will be helpful in determining its role in the care of young women.

Carcinoma of the neovagina is an extremely rare disease, at least 10 times less common than vaginal cancer, which represents only 1–2% of all gynecological cancers (74). The first cases of a neoplastic process in the neovagina involved the development of adenocarcinoma in small bowel grafts (75,76). Subsequently, neovaginal

squamous cell carcinoma was also described (77–80), as well as carcinoma *in situ* (81,82). The evidence supports a risk of neoplastic change in both skin and bowel grafts, with a reported average interval from vaginoplasty to diagnosis on the order of 19 years (83). The use of bowel grafts in children and teenagers could result in the possibility of invasive adenocarcinoma developing in very young women. It is unclear if this represents a risk for any particular colonic segment independently developing adenocarcinoma or whether it may be related to the transplantation of colonic tissue to a new environment. It is therefore important that patients with any type of neovagina have long-term follow-up examinations after vaginoplasty. Patients should also undergo regular cytologic screening. Epithelium transplanted to the vagina will assume the oncogenic potential of the lower reproductive tract. Consideration of a plan for surveillance and counseling regarding transmission risk for virally related dysplastic change in the lower genital tract is also extremely important for any patients undergoing neovaginoplasty. Suspicious symptoms such as bleeding or a palpable mass should necessitate prompt evaluation.

Few cases of neovaginal prolapse have been reported in the literature. Prolapse may be a concern in patients managed surgically or nonsurgically. Creation of a neovagina with dilation may be at risk, as the apex is not anchored to any supporting structures or ligaments. O'Connor and colleagues reported a patient that required conversion to a sigmoid neovagina after complete prolapse of the vagina after progressive dilatation (84). Some authors note a significant incidence of neovaginal prolapse, which may be attributed to the absence of normal supporting ligament (41). A similar phenomenon may also occur in bowel transpositions (39). Mucosal prolapse requiring trimming of the exposed mucosa or anchoring of the bowel segment to the sacrum is known to occur. Hendren and Atala described 16 cases of eversion-prolapse of the bowel segment that required trimming (11). Hendren qualified the report as a simple eversion of the distal segment, not an intussusception as in apical vaginal prolapse. He reported no significant change in the length of the neovagina with resection of the prolapsed tissue. Although the etiology of neovaginal prolapse remains unclear, retroperitoneal sacropexy may be a successful solution, especially when apical prolapse is the concern (85). Communal and colleagues incorporated anchorage of the distal end of the sigmoid neovagina to the fascia of the sacral promontory at the time of the neocolporrhaphy for prophylaxis of prolapse (48).

Regardless of the method of neocolpopoiesis performed, it is important that the operation be performed correctly the first time. If the vagina becomes constricted because of granulation tissue formation, injury to adjacent structures, or failure to comply with postoperative maintenance, then subsequent attempts to create a satisfactory vagina will be more difficult. The first operation has the best chance of success.

Clearly, multiple techniques remain available for creation of the neovagina. There is truly no ideal procedure secondary to the lack of clearly defined and compared measures of success. Commonly, the decision is based on the degree and area of expertise of the surgeon. The application of intestinal neovaginoplasty has widened to treating patients with isolated vaginal agenesis, in addition to patients with more complex anomalies. The McIndoe vaginoplasty has been the most widely performed by gynecologic surgeons. The Vecchiotti procedure, performed laparoscopically, has been performed more readily outside of the United States but has gained some popularity here in recent years. Innovative neovaginoplasty techniques, such as use of Interceed or buccal mucosa, may also have promise as more data is gained regarding long-

term outcomes. Despite all the options, an appreciation of the varying methods of vaginoplasty is essential to provide complete care and counseling to patients and families regarding outcomes and potential reproductive consequences.

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Abdominal Complications of Ventricular-Peritoneal Shunts

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OVERVIEW

The usual reoperative neurosurgical procedure that the pediatric surgeon encounters is revision of a failed ventricular drainage device, most commonly a ventriculoperitoneal shunt (VPS). Shunt failure after primary insertion occurs frequently, with incidence of 40% at 1 year and 50% after 2 years. Shunt complications include infection and dysfunction caused by obstruction, mechanical failure, or abdominal causes. This chapter will focus on the abdominal complications of VPS and the abdominal component of shunt revision. We will also discuss alternative drainage sites for those occasions when adhesions from prior laparotomy or previous shunt procedures make further peritoneal access difficult or impractical. For a comprehensive account of the pathophysiology of hydrocephalus and the different approaches to surgical drainage, the reader should consult a textbook of pediatric neurosurgery.

SHUNT INFECTION

The incidence of shunt infection is 8–10% and usually occurs within the first 6 months after placement. Younger children, and particularly neonates, are at greater

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risk for infection. The most common pathogens are skin flora such as *Staphylococcus epidermidis*.

Clinical Presentation

The onset of illness is generally insidious, and approximately one-half of patients will manifest evidence of shunt dysfunction. Other symptoms include fever, pain, swelling or redness along the shunt tract, meningeal irritation, and abdominal pain. Cellulitis at the shunt insertion site or tenderness along the shunt tract may be observed. Shunt infection is well known to masquerade as an acute surgical abdomen, therefore any child with a recent VPS surgery who presents with abdominal pain and signs of peritonitis must be thoroughly investigated for a shunt infection.

Diagnostic Evaluation

The diagnosis is made by Gram stain and culture of cerebrospinal fluid (CSF) aspirated from the shunt system. If peritonitis is suspected abdominal imaging with computed tomography (CT) or ultrasound may help to detect intraperitoneal abscesses or pseudocyst. These abscesses usually occur near the VPS catheter, although they may be sequestered in distant locations within the peritoneal cavity (Fig. 1).

Operative Management

Once the spinal fluid has been obtained to assess for shunt infection, intravenous antibiotics are initiated. The shunt is externalized, either by removal of the shunt and placement of an external ventricular drain, or simple externalization of the abdominal catheter. Small intraabdominal abscesses may respond to antibiotics alone, but most will require drainage by the percutaneous ultrasound- or CT-guided technique or by operative drainage. Revision of the shunt is performed once eradication of the infection is proved by serial negative CSF cultures. Whenever possible, old incisions should be used in order to reduce the number of abdominal scars in the child who might require multiple revisions in the future. The peritoneal incision should be limited to the minimum extent required to achieve safe placement of the catheter into a large free peritoneal space. If extensive lysis of adhesions is necessary, a larger incision may be required.

Laparoscopic-assisted revision of VPS offers the advantage that peritoneal incisions are minimized and a superior view of the peritoneal cavity is usually possible compared to open procedures. In addition, adhesiolysis can be safely performed and the tip of the catheter guided to a precise location. In laparoscopic VPS revision, it is advisable to place the initial trocar using the open Hasson technique in order to reduce the risk of iatrogenic perforation of viscus.

SHUNT OBSTRUCTION

Obstruction of the VPS can occur at any time after insertion and most frequently involves the ventricular catheter or valve, but may occur distally at any point along the course of the shunt. Shunt obstruction is the most frequent indication for surgical revision.

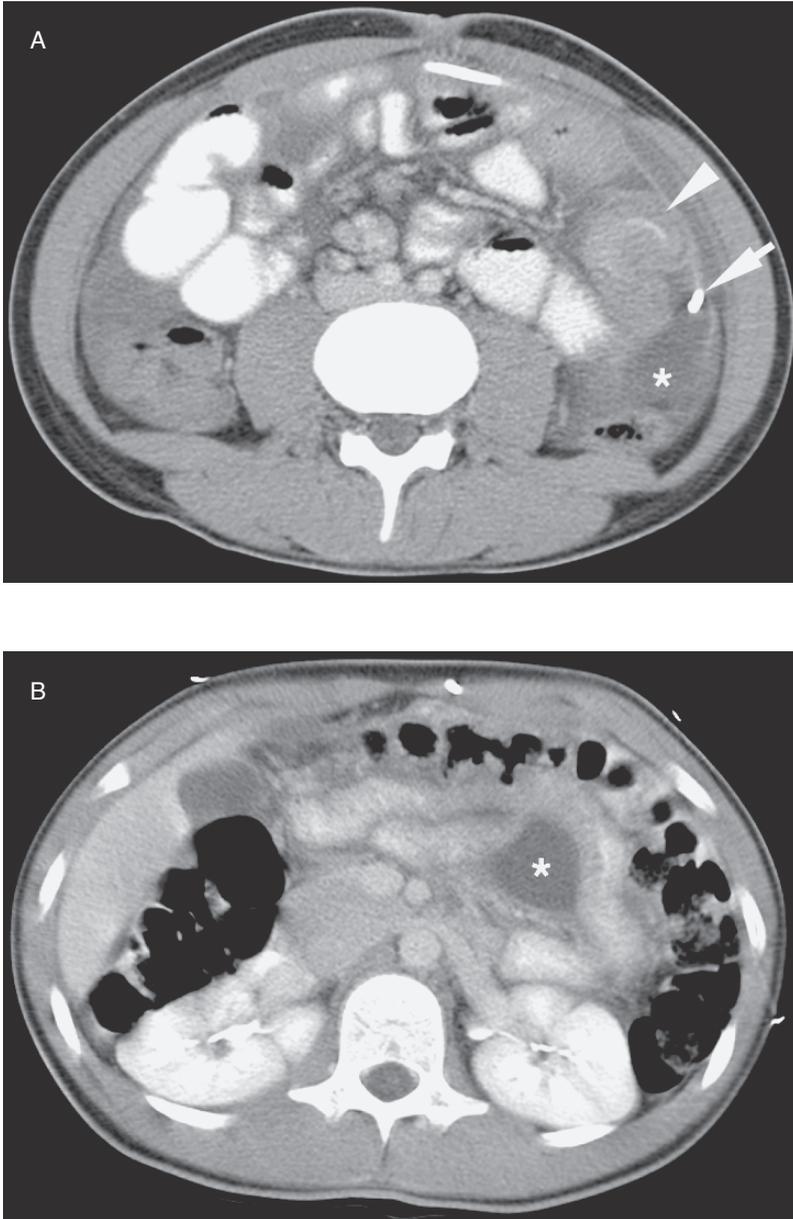


Fig. 1. Intra-peritoneal infection. (A) Contrast-enhanced CT scan shows a focal fluid collection (*) associated with the VPS (arrow). There is associated bowel wall thickening (arrowhead) and peritoneal enhancement indicating infection. (B) Contrast-enhanced CT shows an interloop abscess (*).

Clinical Presentation

The clinical presentation of an obstructed shunt is one of raised intracranial pressure similar to the untreated hydrocephalus. The onset is usually insidious, although can be rapid and life threatening. Symptoms may occur intermittently prior to frank failure. The infant usually presents with irritability, vomiting, and bulging fontanel. Older children often complain of nausea, projectile vomiting, and headaches, frequently occurring in

the morning. Physical examination may reveal fluctuance around the shunt, ataxia, cranial nerve palsies, papilledema, and mental status changes such as lethargy.

Diagnostic Evaluation

Clinical evaluation is paramount in determining the presence of shunt malfunction. Other tests, including imaging studies and shunt taps, may assist in confirming the diagnosis and identifying the site of obstruction. Imaging studies are most helpful when compared with baseline films, especially because early radiologic changes may be quite subtle. A shunt series helps to rule out mechanical failure and CT scan will usually show ventricular enlargement, although shunt failure in the absence of changes on CT is well known. Additional contrast study may help to identify the site of obstruction.

A shunt tap, provided a shunt reservoir is present, may be helpful to determine the location of obstruction. Once the needle is inserted into the reservoir, an elevated opening pressure signals distal obstruction, whereas reduced flow may point to obstruction more proximally. Relief of symptoms by shunt aspiration supports the diagnosis of obstructed CSF flow.

Operative Management

When obstruction at the ventricular end, shunt valve obstruction, and mechanical causes along the shunt tract have been ruled out, poor drainage at the abdominal end is the most likely problem. In these instances, an exploratory laparotomy or laparoscopy may be necessary to detect and relieve the obstruction. Occasionally, the tip of the catheter may be lodged in a preperitoneal pocket or there may be kinking or knotting of the tubing as it enters the peritoneal cavity, caused possibly by a tight anchoring suture. The tip of the catheter may also be obstructed by omentum, or adhesions, or it may be wedged against the surface of abdominal viscera. Finally, the catheter tip may be sequestered within an abdominal pseudocyst, preventing CSF drainage into the general peritoneal cavity (*see* the next section).

PSEUDOCYSTS

In patients with a VPS, abdominal pseudocysts are loculated pockets filled with unabsorbed CSF, usually developing around the peritoneal catheter. They may develop within a few weeks of shunt insertion, but can also present in a delayed fashion several years afterwards. The overall incidence is 1–5% of patients with VPS. Pseudocysts are lined by chronic inflammatory tissue rather than a mesothelial membrane. Although pseudocysts are commonly believed to result from low-grade chronic infection, the fluid is frequently found to be sterile upon aspiration. Predisposing factors include previous shunt infections and peritoneal adhesions from previous abdominal operations. Elevated protein level in the CSF, as may occur in children with intracranial tumors, has been associated with pseudocysts. This is presumably because protein-rich CSF is not well absorbed from the peritoneal space and is more likely to be trapped in loculations (Fig. 2).

Clinical Presentation

Pseudocysts may present as a localized abdominal mass or diffuse swelling. Patients may complain of abdominal pain or discomfort, nausea, vomiting, anorexia, or fever. Large cysts may produce frank bowel obstruction. Interestingly, the vast majority of patients show no signs of shunt dysfunction.

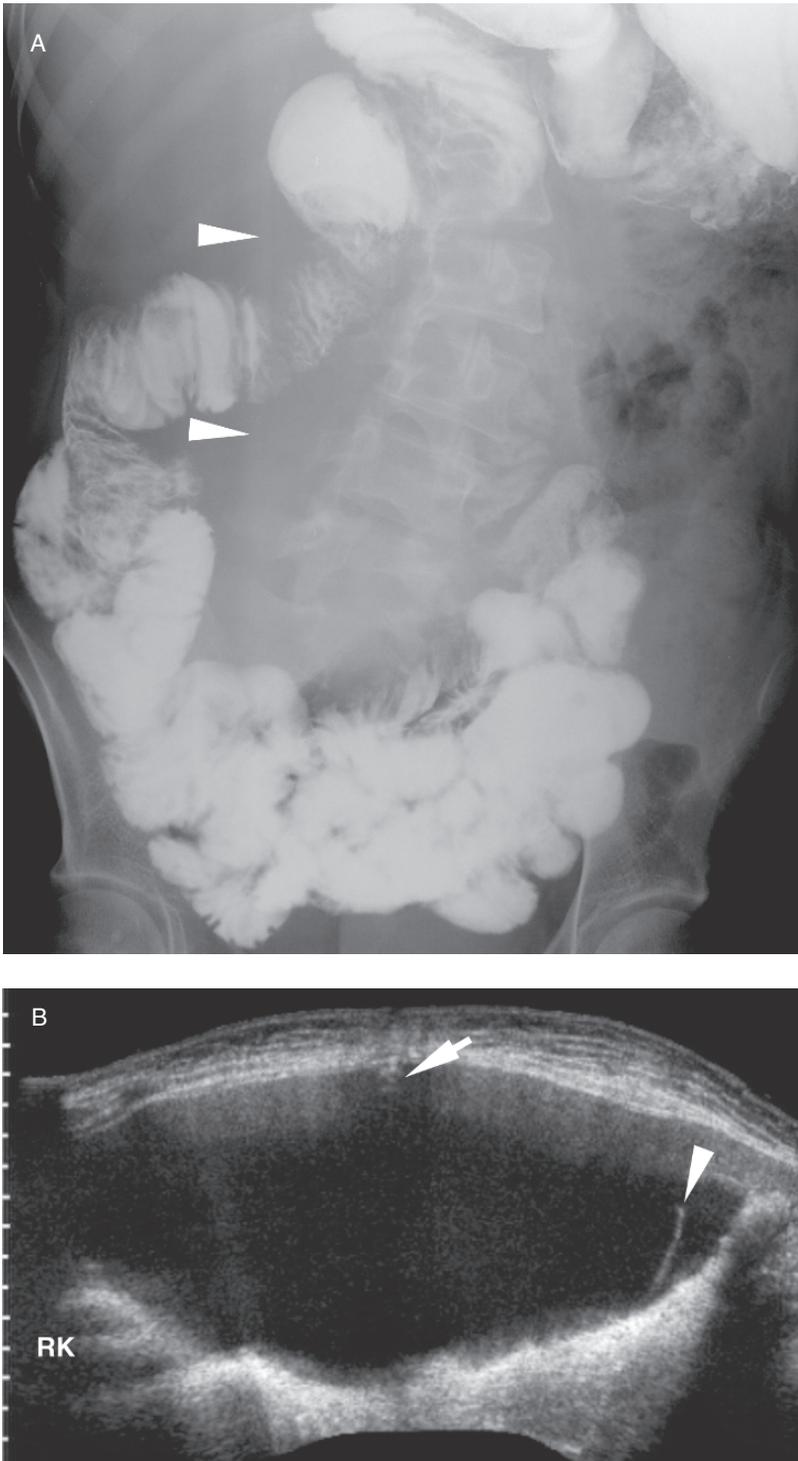


Fig. 2. CSF pseudocyst. (A) Barium study in a patient with spinal dysraphism and abdominal distension and a VPS (arrowheads) shows a mass displacing loops of small bowel. (B) Longitudinal extended field of view sonogram shows a 22-cm-long pseudocyst associated with the VPS (arrow). There is a single septations within the cyst (arrowhead). RK = right kidney.

Diagnostic Evaluation

Plain films may show a large cyst causing bowel displacement or intestinal obstruction. However, the best diagnostic modality is ultrasound, which shows the fluid collection adjacent to or surrounding the tip of the catheter, and the cyst contents completely anechoic. CT scan is essential only if distinction from an abscess is indicated; the cyst walls are thin and nonenhancing whereas an abscess wall is thick and enhances.

Operative Management

With a pseudocyst, the shunt should be removed and externalized in most cases. The cyst fluid is aspirated and submitted to Gram stain and culture. Even if the fluid is sterile, some surgeons will externalize the shunt and treat the patient with a course of antibiotics with the presumption of an underlying low-grade infection. Other authors recommend that a sterile peritoneal shunt should be converted to an atrial or pleural shunt. Usually, removal of the catheter from the peritoneal cavity allows the pseudocyst to resolve spontaneously. Once the infection is eradicated and the pseudocyst collapsed, it is reasonable to reinsert the new catheter into the peritoneum. In the treatment algorithm proposed by Mobley (Fig. 2), only the abdominal portion of the shunt is removed when no infection is detected, with immediate replacement in the peritoneal cavity.

SHUNT MIGRATION

Shunt malfunction may be caused by migration of the shunt outside of the peritoneal space. Children with shunts inserted in early life will occasionally outgrow the length of their shunts, causing the tip to retract into the abdominal wall. The catheter may also migrate through preexistent peritoneal recesses such as a patent processus vaginalis, a Bochdalek's hernia, or other abdominal wall hernia.

Clinical Presentation

The clinical finding will depend on the nature of the shunt migration. If CSF absorption is impeded, the child will develop signs of increased intracranial pressure as described previously. A large indirect hernia or hydrocele in an infant or child with a VPS is likely caused by a persistent processus vaginalis and not necessarily by shunt migration into the sac.

Diagnostic Evaluation

When shunt migration is suspected, plain radiographs in frontal and lateral projection can be very helpful, or the catheter may be localized by ultrasound or CT scan.

Operative Management

Shunt revision is required in these cases so that the appropriate length of catheter is used and optimal placement performed. Abdominal wall and diaphragmatic hernias should be repaired in order to prevent recurrent migration.

VISCERAL PERFORATION

Visceral perforation may occur surgically, with erroneous placement of a catheter into a hollow organ such as the bowel, urinary bladder, vagina, gall bladder, and through the diaphragm into the bronchial tree or into the umbilicus. There may be an increased risk of visceral perforation when the trocar technique is used for insertion. If the trocar is to be used, it is advisable to insert a nasogastric tube and Foley catheter to reduce the risk of perforation of stomach and bladder. A spontaneous mechanism for visceral entry of catheter is chronic erosion of the tubing into the viscus, which usually occurs after several months or years.

Clinical Presentation

Perforation of hollow viscera, especially bowel, may present acutely with peritonitis; but the catheter may completely plug the hole in the viscera, thus preventing further peritoneal leak and resulting in delayed presentation several days or weeks later. Depending on the organ perforated by the catheter, the child may present with watery diarrhea, pleural effusion, copious fluid drainage through the vagina or umbilicus, urinary frequency or incontinence, or shunt infections owing to Gram-negative organisms. Chronic erosion of catheter tubing into a hollow viscus usually has a more insidious and occasionally dramatic presentation, including the extrusion of the shunt catheter through the anus or urethra. In these cases, there may be no associated peritoneal reaction (Fig. 3).

Diagnostic Evaluation

Ultrasound or CT scan may also localize the catheter in a hernia or in an intraluminal site. Bowel perforation can be confirmed by a contrast shunt study, which shows filling of the bowel.

Operative Management

If bowel perforation is suspected, antibiotic therapy should be instituted. In acute cases, emergent laparotomy and repair of viscera is indicated, and the shunt is externalized. When the CSF is sterile, the shunt should be replaced into an alternative drainage site, although the abdomen may be used again when peritoneal contamination has been eliminated. With delayed presentation there is usually a well-formed fistula tract, which closes spontaneously once the catheter is externalized or removed and does not require acute surgical intervention.

LAPAROTOMY IN PATIENTS WITH VPS

The major consideration when performing laparotomy in a child with known VPS is the avoidance of injury to the shunt tubing and minimizing the risk of iatrogenic shunt infection. The surgeon should be aware of the course of the tubing so that injury can be avoided when placing the abdominal incision, including laparoscopic port sites. The risk of shunt infection is directly proportional to the degree of intraperitoneal contamination associated with the surgical procedure. A thorough antiseptic skin preparation should be performed and prophylactic antibiotic used against Gram-positive cocci. In elective cases with minimal contamination, the shunt should not be disturbed. We have performed laparoscopic procedures including Nissen fundoplication, cholecystectomy,

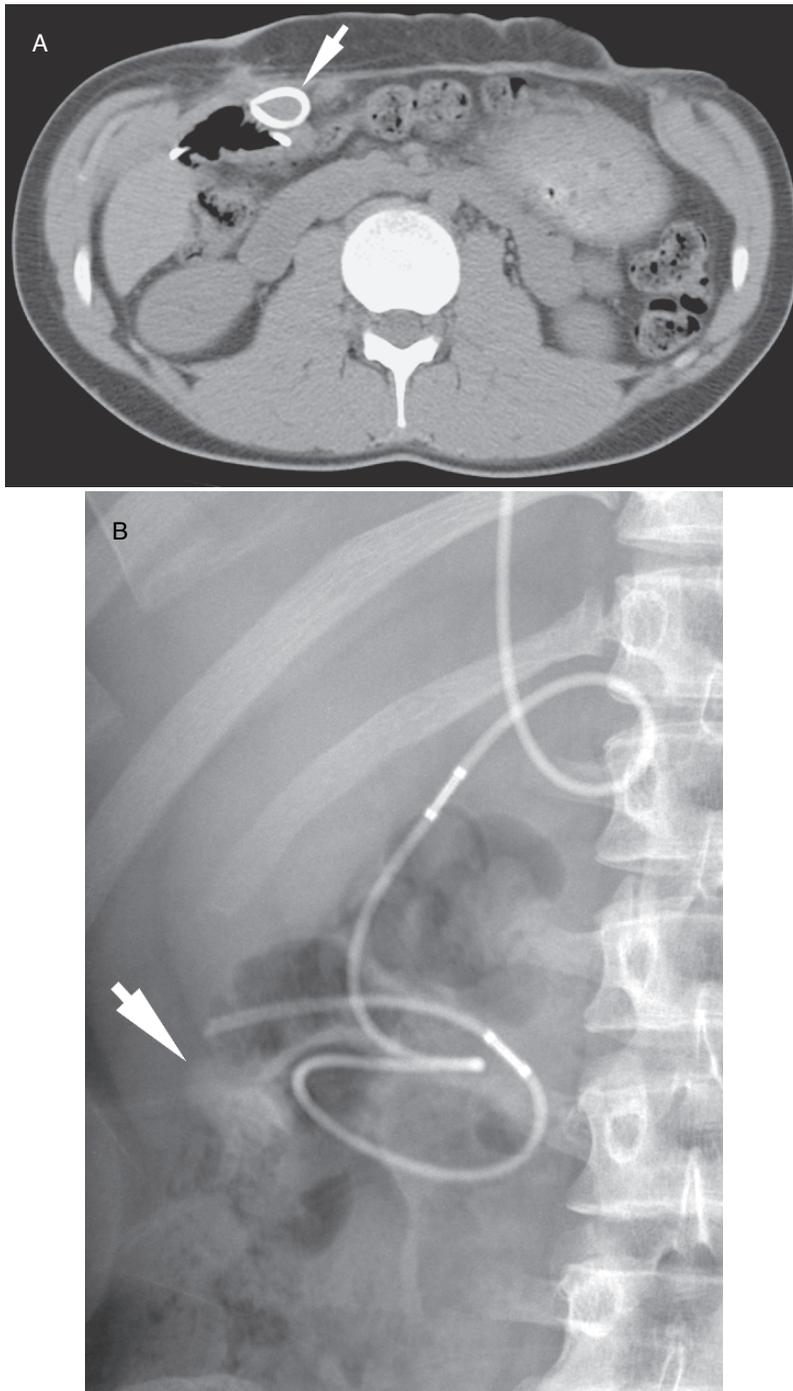


Fig. 3. Bowel perforation. (A) CT scan shows the VPS (arrow) inseparable from the small bowel with minimal surrounding inflammation. (B) Shunt contrast study shows contrast passing from the shunt into the bowel (arrow) confirming bowel perforation.

and gastrostomy in several children with VPS with no adverse sequelae. Several studies support the safety of elective percutaneous endoscopic gastrostomy (PEG) in patients with VPS, but Gassas has pointed out the risk when PEG insertion occurs within a few weeks of the VPS.

For elective surgery, in which significant contamination is anticipated, the shunt should be externalized or converted to other sites before the abdominal procedure. In addition, when peritonitis or other peritoneal contamination is encountered during emergent laparotomy, consideration should be given to externalizing the shunt. Subsequently, the shunt may be relocated to other sites or returned to the abdomen with resolution of peritonitis. Whether the shunt should be externalized in all cases of peritonitis arising from other organs is controversial. Some authors have reported low rate of shunt infection when it was left in the abdomen.

Marked intraperitoneal adhesion formation is often encountered in patients with VPS, which in severe cases could cause a “frozen” abdomen. These adhesions rarely cause bowel obstruction, but could make abdominal reentry quite difficult. Extensive adhesions will likely impair shunt drainage and necessitate relocation to other sites. When bowel obstruction occurs in the patient with VPS, potential causes include intestinal volvulus around the shunt tubing.

ALTERNATIVE TECHNIQUES FOR SHUNT DRAINAGE

As discussed previously, there are several occasions when reinsertion of a VPS is impractical or carries unacceptable risks of shunt infection. In those cases, a number of alternative sites can be used.

Ventriculoatrial shunts are typically used with failure of the VPS. These carry similar risks to VPSs, although they have the added risk of bacteremia and organ failure should they become infected. Ventriculopleural shunts have been utilized with variable success. In children lacking access to these sites, typically for various anatomic or pathologic reasons, we as well as others have successfully placed a number of ventriculogallbladder shunts. Imaging is performed to assess the gallbladder for both stones and tone through ultrasound and stimulation tests. The gallbladder is typically approached through an open laparotomy, irrigated for stones and debris, and the catheter secured into the gallbladder via purse-string suture around a metal connector that has been slid in to the distal end of the shunt catheter. Other anatomic sites have been utilized, including the sagittal sinus, the right cardiac atrium, the kidneys, ureters, and bladder with varying degrees of success.

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Revisional Bariatric Surgery in Adolescents

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INTRODUCTION

The recent surge of interest in various bariatric procedures over the past decade have led to a much better understanding of the pathophysiology of morbid obesity, as well as the complex physiologic and metabolic consequences of surgical weight-reduction procedures. With the geometric increase in the number of patients undergoing bariatric surgery both in this country and abroad, there has been an increased need to understand the role for revisional bariatric surgery in order to optimize clinical outcomes and recognize the need for potential reoperation. In addition to the expanding field of adult bariatric surgery, the surgical community is now faced with an increasing interest in the application of such techniques in the morbidly obese adolescent population.

Childhood obesity has been called “a new pandemic of the new millennium” (1). Although the overall health status of the pediatric population in the US has improved, the last quarter century has witnessed a dramatic rise in the prevalence of childhood obesity (15%) and its associated comorbid conditions, making the pediatric age group the fastest growing subpopulation to suffer the ravages of obesity (2). Data demonstrating a 10-fold increase in excess morbidity among the morbidly obese (body mass index [BMI] >35 kg/M²) lend support to a potential deceleration in life expectancy, rising obesity-related healthcare costs (70–300 billion dollars/year) and obesity-related deaths (300,000/year) (2,3). These health-related effects, as well as potential socio-economic impact (i.e., diminished educational attainment, reduced income trajectory,

and job absenteeism) have resulted in a public health crisis never before seen in this country.

Since its introduction in the 1960s and subsequent refinements, including the introduction of minimally invasive surgical techniques, bariatric surgery has been shown to be both safe and effective in the adult population. The most commonly used weight-loss procedures rely on a combination of gastric restriction and/or intestinal malabsorption to produce 35–70% loss of excess body weight (EBW) (4–8). Data on adolescent bariatric surgery consist of small case series, which offer almost no meaningful insight regarding the potential clinical impact on obesity-related comorbidities (9,10). Furthermore, there is little to no data examining potential differences seen in the adolescent population when compared to an adult cohort (11–13). Despite the current paucity of data regarding the applicability of various bariatric surgical techniques for the treatment of adolescent morbid obesity, an increasing number of pediatric tertiary care centers have committed to establishing adolescent bariatric surgery programs, which will undoubtedly produce the evidence-based data required to help shape the future of weight-reduction surgery in this controversial population. In response to the increasing interest in the applicability of bariatric surgery in the adolescent population, the following chapter addresses complications that develop in both the short-term (within the first month) and long-term (after 1 month) time periods.

Although there are numerous bariatric procedures that can be applied to the morbidly obese adolescent population, this chapter will focus on the complications and subsequent need for reoperative surgery limited to Roux-en-Y gastric bypass (RYGB), the current “gold standard” weight-reduction procedure in the United States and most commonly reported procedure for the treatment of childhood obesity, and the laparoscopic adjustable gastric banding (LAPBAND®; Inamed, A Division of Allergan, Goleta, CA), a purely restrictive procedure that has been gaining wide attention in the United States since its approval for the adult population by the Food and Drug Administration (FDA) in 2001.

In light of the emerging nature of weight-reduction surgery in the adolescent population, and subsequent paucity of large adolescent bariatric series, the authors recognize that much of the information reviewed in this chapter is based on the adult experience and may therefore be subject to change in the next several years as larger scale multicenter longitudinal outcome data becomes available. In accordance with the current consensus that the surgical treatment of adolescent morbid obesity should be undertaken by a team consisting of both an experienced pediatric and adult bariatric surgeon, several institutions, including the Ohio State University School of Medicine, ascribe to a model of close collaboration with our adult colleagues. Because of the complex nature of various surgical weight-reduction procedures as well as an increasing body of literature defining the correlation between surgical complications and clinical outcomes with the technical experience of individual surgeons, the authors recommend close clinical collaboration with adult colleagues at the same institution in an effort to avoid technical misadventures and possibly improve the learning curve (14). In addition to the consideration of this model for primary operations, the authors strongly recommend close collaboration with adult bariatric surgical colleagues before undertaking any reoperative bariatric procedures.

GENERAL CONSIDERATIONS

Although the number of bariatric procedures has been increasing over the past several years and will most likely continue to increase with the better understanding of surgical weight loss in the morbidly obese adolescent population, one should consider several aspects of revisional surgery before undertaking this challenging set of patients. As in the case of other complex gastrointestinal procedures, it is recommended that one identify clear goals and expectations well in advance of potential revisional surgery. Specifically, a clear determination of factors leading to the failure of any primary procedure is imperative before a secondary surgical intervention can be undertaken. Although early postoperative complications are often more clear cut and pertain to wound infections and/or anastomotic problems, the management of long-term complications require significant planning and consideration both on the part of the surgeon, the patient, and, in the case of adolescent bariatric surgery, the patient's family. In addition to the general considerations delineated in Table 1, ethical considerations including the potential scenario of reversing or altering a previous operation in the face of a new or "better" procedure should be anticipated as the use of bariatric surgery gains further acceptance among the adolescent population.

PRIMARY EVALUATION

Evaluation of a patient who has previously undergone bariatric surgery and presents with one or more complications requiring a secondary operation can be a significant diagnostic and therapeutic challenge. In an effort to minimize these difficulties, a step-wise approach should be undertaken. Initial evaluation should consist of a detailed history and physical examination in conjunction with a routine diagnostic work-up (Table 2) in order to determine the associated causes, direct additional diagnostic evaluations, and set clear therapeutic goals. Specific attention should be applied to the patient's perception of the problem with specific emphasis on the degree of weight reduction (i.e., too much or not enough) as well as the presence or absence of several

Table 1
General Considerations

-
1. Review all anatomic considerations of prior surgeries.
 2. Formulate a thorough understanding regarding the suboptimal results of previous operations.
 3. Consider endoscopy and/or contrast radiography in an attempt to better understand possible associated anatomic abnormalities.
 4. Discuss the relevant findings with the patient and family in order to make clear your understanding of why the previous operation failed.
 5. Lay out specific goals of revisional surgery for the patient and family, including expected recovery time, possible complications, and expected future weight loss.
 6. Consider reinforcing educational components such as nutrition and exercise with the patient and family prior to performing a revisional operation.
 7. Consider early consultation and/or referral to a more experienced bariatric surgeon when faced with extremely complex revisional considerations.
 8. Employ behavior counseling early in the course of planning a revisional operation in order to assess potential pitfalls that may lead to further failure.
-

Table 2
Preliminary Patient Evaluation

-
1. Detailed history and physical examination
 2. Nutritional evaluation
 3. Psychological screening
 4. Complete blood cell count
 5. Serum electrolytes, amylase, lipase
 6. Liver and thyroid function panel
 7. Ferritin, prealbumin, and total iron-binding capacity
 8. Vitamin A, D, B₁₂, folate, zinc
 9. Coagulation profile (prothrombin time, activated partial thromboplastin time)
 10. Pregnancy test
 11. Chest radiograph
 12. Bone densitometry
 13. Consider upper gastrointestinal barium study
 14. Consider abdominal CT scan
 15. Drug and alcohol screening
-

commonly encountered postoperative symptoms (i.e., nausea, vomiting, abdominal pain, malaise, and so on). In addition, a review of previous operative notes and, if applicable, communication with the surgeon responsible for the primary procedure and/or previous secondary operations should be sought in order to complete the primary evaluation.

As shown in Tables 3 and 4, a list of differential complications that one might encounter in a postoperative RYGB and/or LAPBAND[®] patient should be considered

Table 3
Complications of RYGB

-
1. Staple-line dehiscence
 2. Diarrhea/Steatorrhea
 3. Poor weight loss
 4. Pouch fistula formation
 5. Bile reflux
 6. Chronic blood loss
 7. Enlarged gastric pouch
 8. Anastomotic stricture
-

Table 4
Complications of LAPBAND[®]

-
1. Stomach slippage
 2. Stoma obstruction
 3. Gastric pouch and esophageal dilatation
 4. Band erosion
 5. Gastric necrosis
 6. Poor weight loss
-

early in the course of a primary evaluation in order to direct diagnostic and therapeutic efforts in a timely and efficient manner.

DEFINITIONS OF SUCCESS

Before embarking on further discussion about revisional bariatric surgery, one must define success and failure in terms of both the immediate postoperative period as well as the long-term postoperative time frame. Failure in the immediate postoperative period is more easily addressed and for the purposes of this discussion, will focus on anastomotic complications (i.e., leakage and/or stenosis) as well as internal herniation (which may be encountered both during the immediate and nonimmediate postoperative periods).

The definition of long-term success is a more complex issue and must take into consideration not only the overall efficacy of the primary operation (typically measured by the degree of EBW loss) but the relative amelioration of various obesity-related comorbid diseases. Even more complex, and beyond the scope of this chapter, is the determination of success as a measure of “quality of life” indicators. As an example of the individual considerations that need to be applied to definitions of success, patients who fail to obtain optimal weight loss within 2 years of undergoing RYGB (i.e., 50–70% EBW) may not require surgical revision if there has been a significant improvement in obesity-related comorbid conditions and overall improvement in quality of life. As alluded to earlier, a team-oriented approach with input from various healthcare disciplines is highly recommended in order to accurately define success and determine the need for reoperation.

REVISION OF RYGB

The RYGB (Fig. 1) is an extremely effective means of establishing durable weight loss and reversing associated comorbid conditions in the morbidly obese population. Frequently referred to as the “gold standard” among numerous surgical weight-reduction surgeries, this procedure is the most commonly performed bariatric procedure in the United States as well as the primary procedure of choice among the various bariatric procedures currently available. In addition, it has also been cited as the preferred secondary procedure for patients requiring revisional surgery following several well-described alternative primary operations. Despite the general safety and efficacy and of this procedure, several well-recognized complication of the RYGB have resulted in a predictable need for revisional surgery (Table 3).

Immediate Postoperative Period (Anastomotic Complications)

Although patients undergoing any form of bariatric surgery are subject to a host of potential complications during the immediate and late postoperative period (i.e., deep venous thrombosis, wound infection, pulmonary embolism, and so on) information in this section will focus on more bariatric-specific issues that may result in the need for a secondary intervention (including reoperation) in the immediate postoperative period.

ANASTOMOTIC LEAK

Leakage from the various enteric anastomoses in the immediate postoperative period may pose a significant diagnostic challenge and should be considered in patients even

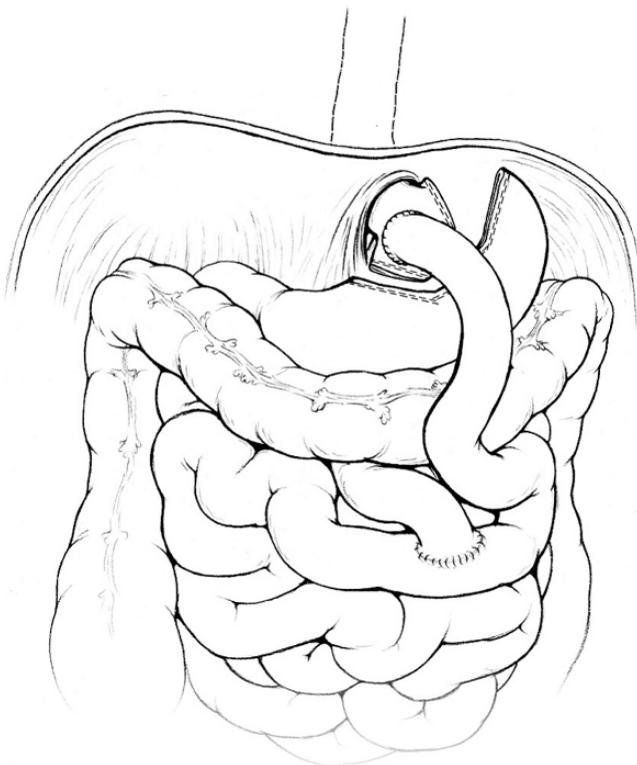


Fig. 1. Roux-en-Y gastric Bypass.

in the face of a technically unremarkable operation (open or laparoscopic), a negative intraoperative leak test, and/or a negative postoperative contrast study. Leakage may occur at either the gastrojejunostomy or the jeunojejunostomy. Although patients with early anastomotic leakage may present with gross peritoneal signs with or without associated hemodynamic embarrassment, more subtle clinical findings may be the only clue that a complication has occurred. Such patients may be noted to have persistent tachycardia with or without fever, leukocytosis, poor oxygen saturation, left shoulder pain, hiccups, and a feeling of restlessness or agitation. Any combination of subtle findings warrants the serious consideration of abdominal reexploration within the early postoperative period in order to rule out an anastomotic complication.

ANASTOMOTIC NARROWING

In addition to leakage, narrowing of the anastomosis secondary to technical error or postoperative inflammation may result in intestinal obstruction at the level of the gastrojejunostomy or jeunojejunostomy. This complication may occur in the early postoperative time period and result in signs and symptoms of acute bowel obstruction. As with patients experiencing postoperative anastomotic leakage, patients with intestinal obstruction often require the ability to distinguish associated symptoms (i.e., nausea, vomiting, and abdominal pain) from common complaints often encountered during the normal postoperative recovery period. Again, interpretation of subtle findings (i.e., persistence of abdominal discomfort for a longer than usual time period) should be

interpreted as having the potential to represent a patient with an intestinal obstruction and, as stated earlier, should prompt early surgical reexploration.

INTERNAL HERNIA

In addition to the anastomotic complications discussed previously, the creation of several potential sites for intestinal herniation can also serve as a cause for problems in both the immediate and late postoperative periods (15–17). Intestinal obstruction may result from herniation of small bowel through three commonly recognized sites: (1) the mesocolic defect through which the Roux limb passes in cases of a retrocolic approach; (2) the “Petersen” defect between the posterior portion of the transverse mesocolon and the Roux limb; and (3) the mesenteric defect created at the jejunojejunostomy site (Fig. 2).

The initial goal of management of postoperative small bowel obstruction should be patient resuscitation (fluid replacement and correction of metabolic derangement) and include intestinal decompression with an endoscopically placed nasogastric tube (NGT). The use of flexible endoscopy rather than blind placement of an NGT allows for inspection of the gastric pouch and enteric limb while avoiding the risk of inadvertent perforation of the gastric pouch. In addition to providing proximal decompression, the use of an NGT will allow for further assessment using contrast radiography. The use of such conservative measures should be carefully weighed against a low threshold for return to the operating room in the face of no improvement within 24 hours or signs of deterioration and/or relative operative indications, including increasing leukocytosis, fever, or persistent abdominal tenderness. Once the decision to proceed to surgery has been made, one should decide whether to use a minimally invasive techniques

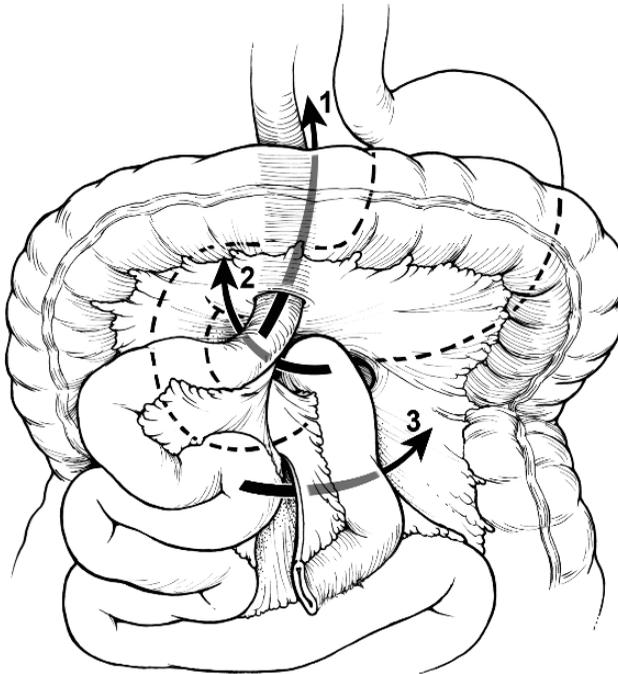


Fig. 2. Potential mesenteric defects involved in postoperative small bowel obstruction. (1) Transverse mesocolon window; (2) Petersen’s space; (3) enteroenterostomy mesenteric defect.

(i.e., laparoscopic) or an “open” approach based on the comfort level and experience of the individual surgeon, because deciphering the abnormal anatomy in an effort to correct the herniation can be quite challenging. Once the defect is identified, careful reduction of the herniated bowel and closure of the defect should be carried out in order to prevent recurrent herniation in the future.

Late Postoperative Period

In contrast to the complications associated with the early postoperative period outlined previously, the remainder of this chapter is devoted to a discussion of a number of well-recognized anatomic and functional derangements that may be encountered months or years after a seemingly successful primary operation.

STAPLE-LINE DEHISCENCE

Most often encountered in the setting of a nondisconnected RYGB, dehiscence of the staple line results in direct communication between the gastric pouch and the remaining gastric remnant. As the restrictive component of the original operation is subsequently compromised, the patient often experiences weight gain, as well as the possibility of bile reflux and ulceration within the Roux limb and/or anastomotic sites. Because of the 5–10% incidence of staple-line dehiscence encountered when using this technique, most bariatric surgeons advocate complete gastric division when fashioning the restrictive pouch (Fig. 1). In the event of a patient that presents with staple-line dehiscence subsequent to a nondisconnected RYGB, complete division of the full staple line with repair of the dehiscence is the revisional procedure of choice. In addition, one should carefully assess the size of the proximal gastric pouch and be prepared to revise it with linear staplers in order to insure that it has total volume no greater than 30 mL.

DIARRHEA/STEATORRHEA

When presented with a patient experiencing significant diarrhea and/or steatorrhea, one must take a very systematic approach and address both dietary concerns and potential nutritional deficiencies. Before definitive surgical therapy can be undertaken, it is advisable to assess the degree of protein, calorie, and fat deficiencies as well as potentially dangerous electrolyte derangements. Once the patient is nutritionally and hemodynamically resuscitated, a systematic assessment of potential causes can be explored. As alluded to previously, a comprehensive review of dietary input should be undertaken by a registered dietician in order to rule out the possibility of diet-related dumping syndrome. This is recommended prior to undertaking potentially time-consuming diagnostic endeavors and possibly subjecting the patient to an unnecessary reoperation in the face of an easily correctable problem. If dietary influences can be ruled out as a potential cause, one should consider the overall length of the Roux limb as a potential cause for this complication. Recent evidence has advocated for the use of progressively longer limb lengths (150–200 cm) in association with increasing BMI (50 kg/M^2) (18,19). The resultant shortening of the common channel, as a consequence of the creation of a long Roux limb, is the most common anatomic factor responsible for persistent nondiet-related diarrhea and/or steatorrhea. Once nutritional and metabolic derangements have been fully addressed, the patient should undergo a lengthening of the common channel in an effort to improve the absorptive capacity of the small bowel

while maintaining acceptable weight loss. In addition to lengthening of the common channel, one should consider insertion of a feeding tube into either the gastric remnant or proximal small bowel in an effort to help optimize the patient's nutritional status during the recovery phase.

POOR WEIGHT LOSS

As with the scenario described previously, patients who present to a bariatric surgeon with poor weight loss or weight gain months or years after under going RYGB require immediate nutritional assessment, because many of these patients can attribute poor weight reduction or weight regain to suboptimal dietary habits. A strong indication that dietary habits are potentially playing a role in this scenario is inferred when encountering a patient that demonstrated adequate weight loss during the early postoperative period (i.e., within the first 6–24 months). In addition to nutritional assessment, one should consider behavioral counseling with the bariatric team psychologist in order to address poor eating habits. Recommendations regarding reoperation for poor weight loss, in the face of an anatomically intact RYGB, are controversial at best owing to the paucity of meaningful clinical data. Anecdotal reports of improved weight loss as a result of limb lengthening have been reported however.

GASTROGASTRIC FISTULA FORMATION

Formation of a fistula from the gastric pouch to the gastric remnant can present as a formidable problem that is typically seen as a result of an anastomotic leak. Unlike several of the complications described earlier, a gastrogastic fistula cannot be controlled through nonoperative means. Definitive surgical therapy in this situation may require ligation of the fistula with limited or total proximal gastrectomy.

BILE REFLUX

In the case of RYGB bile reflux is typically encountered as a result of a Roux limb that is too short. Most bariatric surgeons advocate making the Roux limb at least 85–100 cm in length in order to avoid this complication. If it is determined that a Roux limb is too short, reoperation should consist of revision of the enteroenterostomy in order to create a final limb length of at least 150 cm. As stated previously, other causes of bile reflux may be seen in the case of gastrogastic fistula formation as well as staple-line dehiscence in the case of a nondisconnected RYGB. Reoperative surgical strategies designed to address these particular anatomic complications (described previously) should resolve exposure to bile.

CHRONIC BLOOD LOSS

Chronic blood loss may be encountered in a small subset of patients who have undergone RYGB. Associated causes include antral gastritis and duodenal ulceration. Although the exact pathophysiology involved in post bypass antral gastritis is uncertain, a trial of oral antibiotics for the treatment of presumed *Helicobacter pylori* infection may be warranted. This recommendation supports the argument for routine preoperative screening and treatment of *H. pylori* in morbidly obese patients undergoing bariatric surgery. In rare instances, chronic blood loss secondary to postbypass antritis may require complete gastrectomy of the gastric remnant. As in the case of postbypass antritis, the development of ulceration at the level of the duodenum is another classic cause for chronic anemia, with an associated incidence of approximately 1–2 %. Again,

treatment with antibiotics for presumed *H. pylori* infection is recommended prior to any reoperative surgical option. In cases when antibiotic therapy fails to improve the situation, reoperative surgical options should be predicated on the classic surgical approach to chronic bleeding ulceration of the duodenum recalcitrant to medical therapy (i.e., antrectomy). In cases of an acutely bleeding duodenal ulceration one should consider radiographic embolization or direct ligation of the associated feeding vessel.

ENLARGED GASTRIC POUCH

Although the causes of pouch enlargement remain unclear, several authors have suggested that this phenomenon may be more likely caused by the creation of a pouch that is too large at the culmination of the original operation (i.e., greater than 30 mL capacity) rather than an actual enlargement of the pouch subsequent to the procedure. A larger than normal pouch has been attributed to suboptimal weight loss as well weight gain during the postoperative period. As discussed in the section on poor weight loss, reoperative surgery in the face of an anatomically intact RYGB is controversial. Surgical options consist of decreasing the size of the pouch after taking down the gastrojejunostomy and excluding any gastric fundus before reestablishing intestinal continuity. In addition, one should consider lengthening the size of the Roux limb by at least 50 cm in an effort to increase the degree of intestinal malabsorption capacity. A third but more controversial consideration consists of recent reports of the application of a laparoscopic adjustable band to the gastric pouch in the case of suboptimal weight loss and/or weight gain in patients with RYGB (20).

ANASTOMOTIC STRICTURE

Anastomotic stricture following RYGB has been attributed to localized ischemia at the level of the gastrojejunostomy owing to either tension on the anastomosis or ischemia at the terminal end of the Roux limb. In addition, stricture formation has been seen in association with overuse or abuse of nonsteroidal antiinflammatory agents (NSAIDs). The clinical consequences may be mild or may result in acute obstructive symptoms that may dictate the appropriate level of surgical intervention. The first-line treatment for patients with significant stricture formation is endoscopic balloon dilatation. This procedure should only be performed by a bariatric surgeon or endoscopist familiar with this complication in order to avoid the mistake of overdilating the anastomosis. The use of this technique is often very successful in the immediate postoperative period (i.e., first several months); however, it becomes significantly more challenging later on as a result of significant ulcer formation and associated fibrotic changes. Failure of endoscopic balloon dilatation in the setting of severe fibrosis may dictate the need to perform a formal revision, if the gastrojejunostomy with resection of surrounding fibrotic tissue. Fibrotic reaction involving the proximal pouch may be so severe that an esophagojejunostomy must be performed.

REVISION OF LAPBAND

As mentioned to earlier, placement of the LAPBAND® system is the most widely performed bariatric surgical procedure in Europe and Australia, and has been gaining popularity within the United States since its approval by the FDA in 2001. Several perceived advantages, including the lower overall complication rate, purely restrictive design (Fig. 3) and potential for reversibility when compared to a RYGB, make this option increasingly attractive to both the morbidly obese adult and adolescent populations.

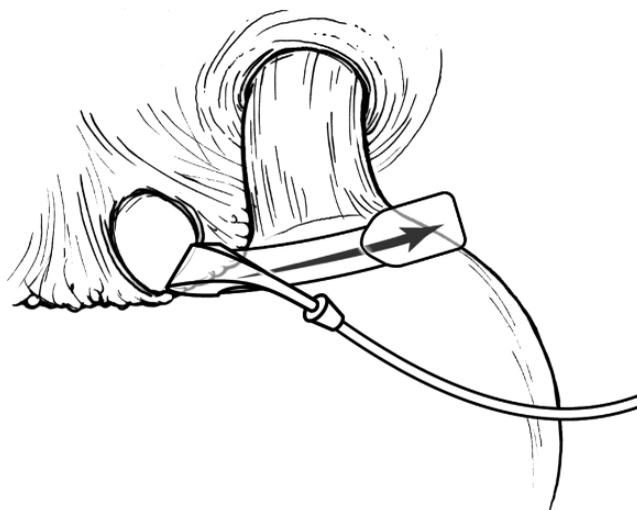


Fig. 3. Laparoscopic-adjustable gastric band.

Immediate and Late Postoperative Period

In addition to acute complications generally associated with any abdominal surgery, specific complications related to the use of the LAPBAND system are seen in both the early and late postoperative periods, and include dilatation and perforation of the stomach and/or esophagus, necrosis of the proximal gastric pouch, stomal obstruction, band erosion, and malfunction and/or disruption of the silastic tubing and subcutaneous port system (Table 4).

Gastric Perforation

With an incidence ranging from 0.2–3.5%, perforation of the stomach after LAPBAND[®] insertion is typically encountered in the immediate postoperative period and has been associated with technical errors (i.e., perforation caused during the retrogastric dissection) seen during the learning-curve period (21–26). Symptoms are typically the result of associated peritonitis and may range from vague abdominal tenderness to gross peritoneal signs. In the event that a gastric perforation is encountered in the postoperative period, one should completely remove the band before performing a formal repair. Reapplication of the LAPBAND[®] system in this setting should be considered on an individual basis, but should be discouraged in the case of large peritoneal contamination and/or associated gastric inflammation.

Stomach Slippage

Widely recognized as the most commonly encountered complication of the LAPBAND[®] insertion, and typically seen in the late postoperative phase, slippage of the stomach (anterior or posterior) through the band is sometimes mistaken as pouch dilatation but rather, is deemed to be the result of an oversized gastric pouch created during the primary procedure. Although initial reports cited an incidence as high as 10%, modification of the surgical dissection technique from the previously endorsed “perigastric” technique to the more recently recommended “pars flaccida”

(Fig. 3) dissection (encompassing minimal tissue dissection and a diminished incidence of posterior slippage) has led to a recently improved incidence of 2–5% (26,27). Consideration that gastric slippage (both anterior or posterior) has occurred should take place when one encounters typical associated symptoms. Specifically, patients may complain of one or several symptoms similar to those encountered with classical gastroesophageal reflux disease (i.e., wheezing, dyspepsia, regurgitation, coughing or choking spells, difficulty swallowing solid food, and so on).

Diagnosis of either posterior (the most common) or anterior slippage of the stomach is best confirmed by obtaining an upper gastrointestinal series. When performed after the band has been completely deflated, this radiographic modality can help differentiate dilatation of the gastric pouch (which may improve with simple band deflation) from true gastric slippage (which is unlikely to resolve following this diagnostic maneuver). Once it has been established that the associated symptoms are caused by true gastric slippage, surgical options include readjustment or complete replacement of the LAPBAND system. Reoperation (laparoscopic or open) in the case of both anterior and posterior gastric slippage begins with careful intraoperative inspection in an effort to confirm the radiological diagnosis, assess the viability of the involved stomach, and determine the specific anatomy (i.e., anterior versus posterior slippage).

Anterior gastric slippage is the result of inadequate fixation of the anterior portion of the band (which is obtained by the placement of several strategically placed fixation sutures). Once confirmed, and assuming that complete removal of the LAPBAND® system is not required, the surgical goal should be to gently pull the herniated portion of stomach back into an optimal position prior to replacement and/or supplementation of previously placed anterior fixation sutures to resecure the band (Fig. 4).

Rather than attempting to pull the stomach back into place, as in the reoperative treatment of an anterior slippage, posterior herniation invariably requires complete removal of the band with its reapplication in a more cephalad position using the pars flaccida technique (incorporation of a portion of the lesser omentum) as shown in Fig. 3.



Fig. 4. Application of gentle traction on the stomach in an attempt to reduce an anterior slip.

Gastric Pouch and Esophageal Dilatation

The pathophysiologic definition of gastric and/or esophageal dilatation in the case of a patient with a LAPBAND is somewhat confusing, because there appears to be overlap with band malposition and slippage (*see* previous section). Whether or not dilatation of the esophagus and proximal gastric pouch are independent entities or the consequence of suboptimal band positioning versus band-related outlet obstruction remains controversial. In addition to such etiologic uncertainty, other potential causes that require consideration include overinflation of the band as well as consumption of inappropriate food or noncompliance with proper mastication techniques (i.e., failure to chew food well before swallowing), resulting in a state of chronic partial obstruction of the esophagus or proximal gastric pouch. Assuming that the patient has been thoroughly evaluated (including an upper gastrointestinal series, careful assessment of the degree of band inflation, and potential patient compliance issues) and has failed to show improvement after a trial of band deflation, one should assume that gastric slippage and/or band malposition has occurred and proceed as described earlier.

Band Erosion

The incidence of band erosion, defined as a breach of the gastric wall underlying the band, is currently reported to be between 0–14 (22,25,28,29). This complication is thought to be associated with various etiologic factors including local tissue ischemia (possibly secondary to microperforation during the primary procedure), chronic inflammation, and chronic ingestion of NSAIDs. Patients do not typically exhibit signs of overt sepsis but rather present with fatigue, weight regain and/or inadequate weight loss, failure to improve weight loss after multiple adjustments or “tightening” of the inner diameter of the device, and port site infection. Bands that have eroded must be removed either operatively (open or laparoscopic), endoscopically, or using a combination of both modalities. The decision whether to insert a new band immediately or at a later time must take into consideration based on individual circumstances, including the surgeon’s overall level of technical experience.

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