Welcome to the pediatric surgery service. Pediatric surgery is primarily concerned with the diagnosis and treatment of surgical diseases of children, and differs from general surgery in that it is an age defined surgical specialty instead of an anatomically defined specialty. Pediatric surgeons are usually trained in the management of general surgical disease of children but also in the treatment of thoracic, head and neck, urologic, and other surgical diseases, whereas most “adult” surgical specialist tend to be confined to the abdomen, chest, or other organ systems alone. As such, pediatric surgery covers a very broad spectrum of disorders and requires one to be familiar with the various organ systems and body areas. On top of this, children are not “little adults”, and their pathophysiologic response to disease frequently differs dramatically from that in adults.

The study of pediatric surgery is often overlooked in medical school but is of significant importance to all physicians, especially those who plan a career in the primary care specialties. Surgical disorders in children are common, and a working familiarity with these disorders will greatly benefit the primary care physician as well as his or her patients.

This monograph discusses the more common surgical disorders of infants and children. A more thorough discussion can be found in the various pediatric surgical texts; two popular and useful texts are:

Rowe et.al. Essentials of Pediatric Surgery Mosby-Year Book, Inc. (1995): This is a good, general, easy-to-read textbook which summarizes the majority of pediatric surgical disorders (edited by my pediatric surgery mentor, Dr. Marc Rowe).

O’Neil, et.al. Pediatric Surgery, 5th Edition Mosby-Year Book, Inc. (1998): This two-volume text is the current “bible” of pediatric surgery. Most chapters are very detailed, and you can find information on virtually all of the more rare disorders.

Another useful media in the 21st century is the Internet, and pediatric surgeons have been very prolific in creating on-line educational sites. Some good sites to check out are:

http://www.eapsa.org/ – the official website for the American Pediatric Surgical Association.


www.vesalius.com – an online pediatric surgical tutorial, with photos and mpegs of pediatric surgical diseases and procedures.

http://www.vh.org/Providers/TeachingFiles/CAP/CAPHome.html – The Virtual Children’s Hospital Correlapedia, an online collection of pediatric surgical disorders catalogued to age and organ systems; a great review tool, and excellent photographs.


http://www.pedinfo.org – PEDINFO site; a directory of internet sites that are dedicated to pediatrics and pediatric subspecialities. A great place to start when looking for pediatric stuff on the net.

http://home.coqui.net/titolugo/ - Pediatric Surgery Update, an online, continually updated syllabus for pediatric surgical disorders.

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ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA (EA/TEF)

EMBRYOLOGY

At approximately 4 weeks of gestation, the trachea develops as a ventral diverticulum of the foregut. The trachea gradually separates from the foregut through the partitioning of the two structures by the esophagotracheal septum. Failure of the development of this septum produces a persistent communication, or fistula, between the trachea and the esophagus. The etiology behind the development of esophageal atresia is poorly understood, but this appears to occur at the same point in development. Because this is a relatively early embryological event, EA/TEF frequently is associated with other developmental anomalies, including the VACTERL association anomalies. The overall incidence of EA/TEF is approximately 1 in 5000 live births.

CLASSIFICATION

There are five standard types of EA/TEF, described as the Vogt-Gross classification system. The most common type of anomaly is type C, which is esophageal atresia with a distal TE fistula; this is seen in over 85% of cases. Pure esophageal atresia (type A) is the next most common, being found in up to 8% of cases. The remaining types of EA/TEF are quite rare, although their treatment is similar to the more common types.

Diagnosis

EA/TEF is a common surgical cause of respiratory distress in the neonate. Classically, infants will develop excessive salivation, with choking and coughing when trying to drink due to an inability to swallow, with resulting aspiration into the lungs. Infants may also develop severe chemical or bacterial pneumonitis due to the reflux of gastric contents into the lungs through the fistula. The diagnosis is confirmed when one is unable to pass a feeding tube into the stomach, with x-ray confirmation of the tube coiling in the proximal esophageal pouch. Contrast x-ray studies can confirm the esophageal atresia but should be avoided in most cases due to the risk of aspiration of contrast into the lungs. EA/TEF is occasionally diagnosed by prenatal ultrasound; the usual findings are maternal polyhydramnios; the presence of an enlarged proximal esophageal pouch; and small or undetectable fetal stomach.
“H-type” TEF (tracheoesophageal fistula without atresia) is usually not diagnosed in the newborn period, since these children are able to swallow. These lesions are diagnosed later in infancy in the child who presents with recurrent respiratory symptoms during swallowing, or recurrent pneumonia, and is confirmed by performing a contrast esophogram or by discovering the presence of a fistula during bronchoscopy.

**PREOPERATIVE MANAGEMENT**

Infants with EA/TEF are at significant risk of developing pneumonitis from aspiration of saliva and stomach contents, with resulting respiratory distress. Prior to repair, the infant should be maintained with the head elevated in the reverse Trendelenberg position. A sump-type tube (Replogle tube) should be placed into the proximal esophageal pouch on suction in order to prevent aspiration, and the child should be placed on broad-spectrum antibiotics. A search is made for other VACTERL-type anomalies, particularly of the gastrointestinal, renal, and cardiac systems, since many of these disorders will also require surgical treatment in the newborn period as well. Associated anomalies are seen in up to 50-70% of children born with EA/TEF, and frequently involve several organ systems. Pneumonitis is the most serious preoperative complication seen in these infants, and if present should be treated vigorously with intravenous antibiotics, aggressive pulmonary therapy, and prevention of further aspiration.

**SURGICAL TREATMENT**

The two surgical goals for infants with EA/TEF are (1) division of the fistula, with separation of the respiratory and gastrointestinal tracts, and (2) establish continuity of the esophagus so that the child may swallow. This is generally accomplished through a posterolateral thoracotomy (usually on the right, but occasionally from the left side). During the procedure, the distal esophagus and its fistulous connection to the back wall of the trachea is identified. This fistula is then divided, and the tracheal opening is repaired. Next, the proximal esophageal pouch is identified and dissected until the two ends of esophagus are close together, allowing for a tension-free primary anastomosis. A chest tube is placed in the area of the repair, and the procedure is terminated. Occasionally, the "gap" between the two ends of esophagus is very wide, necessitating a variety of maneuvers in order to bring the ends together. Sometimes this is impossible because esophageal length is simply not adequate; in this setting, the child is given a cervical esophagostomy in order to allow the drainage of saliva and a gastrostomy for feeding into the stomach. These children then require another procedure at a later time where some form of esophageal substitute, such as colon or stomach, is placed in order to re-establish esophageal continuity and allow oral feeds.
OUTCOME

Prior to 1941, infants with EA/TEF suffered 100% mortality. Today, infants as small as 1200 gms can undergo primary esophageal repair and expect a survival rate in excess of 90%. The majority of deaths in this patient population are attributable to associated congenital heart lesions, not the esophageal disorder itself. These children do suffer complications, however. Immediate perioperative complications include a risk of anastomotic leak (~15-20%) and anastomotic stricture (~15%). Long-term complications include a very high rate of gastroesophageal reflux, with up to 30 to 40% of children requiring fundoplication to treat their reflux.

CONGENITAL DIAPHRAGMATIC HERNIA

EMBRYOLOGY

The diaphragm forms between the fourth and eighth week of gestation and separates the coelomic cavity into the pleural and peritoneal cavities. The central tendon derives from the septum transversum, and the peripheral muscular portions of the diaphragm develop from the fusion of the posterolateral pleuroperitoneal membranes with the septum transversum. Failure of this fusion produces a persistent posterolateral congenital diaphragmatic hernia (CDH), or foramen of Bochdalek hernia.

ANATOMY

85 to 90% of CDH occurs on the left side; bilateral CDH is very rare and almost uniformly fatal. This disorder is seen in approximately 1 in 5000 live births, but is much more common in stillbirth infants. These defects can vary from a small, slit-like opening in the diaphragm to complete agenesis of the hemidiaphragm.
PATHOPHYSIOLOGY

CDH frequently produces severe respiratory distress as a result of two pathophysiologic processes: pulmonary hypoplasia and pulmonary hypertension (also known as persistent fetal circulation). Pulmonary hypoplasia results from the effects of normally intra-abdominal organs compressing the fetal lungs during critical periods of lung development. This impedes normal lung development, with a decrease in overall numbers of bronchopulmonary segments and alveolar surface area, both of which are critical for gas exchange. Histologically, these lungs appear very immature. Persistent fetal circulation (PFC) occurs due to the failure of the pulmonary circulation to undergo the normal transition from the high resistance, high pressure fetal circulation to the low resistance, low pressure neonatal circulation. This prevents adequate blood flow through the lungs, and the blood instead is "shunted" from the right heart to the systemic circulation via the foramen ovale and the ductus arteriosus. The net result of these changes is a pulmonary circuit with inadequate alveolar surface area for gas exchange and where blood is shunted away from the lungs. Because of this, these infants present early with severe respiratory distress, hypoxia and hypercarbia.

PRESENTATION AND DIAGNOSIS

Most cases of CDH are currently diagnosed in utero using prenatal ultrasonography, and can be detected as early as 20 weeks of gestation. At birth, most infants are symptomatic and exhibit some degree of respiratory distress; this may range from mild tachypnea to severe cardiopulmonary collapse. Some infants are born with relatively little hypoplasia but who develop progressive respiratory symptoms due to the development of worsening pulmonary hypertension; this early period of stability is classically known as the “honeymoon period”. On physical examination, breath sounds are usually not detectable on the ipsilateral side, and the heart sounds may be shifted to the contralateral hemithorax. The abdomen is usually flat, or scaphoid. One must also look for other associated anomalies, particularly cardiac disorders, neural tube defects, or trisomies, since their presence with an associated CDH portends a very poor prognosis. A chest xray is usually diagnostic and will demonstrate loops of intestine in the ipsilateral hemithorax with associated contralateral mediastinal shift; on the left side one may see the nasogastric tube coiled in the intrathoracic stomach.

TREATMENT

The historical treatment for CDH was emergent surgical reduction of the hernia with repair of the diaphragmatic defect. In fact, this has been shown to transiently worsen pulmonary function, and most pediatric surgeons and neonatologists currently feel that a period of stabilization for 24 to 48 hours is beneficial in allowing for resolution of pulmonary hypertension prior to surgical repair. Most infants with a known CDH are intubated and sedated at birth, and
are placed on mechanical ventilation. Many will require vasopressors in an attempt to augment pulmonary blood flow, and numerous vasodilators have been used over the years to attempt to decrease pulmonary vascular resistance; unfortunately, most of these are not selective and will produce systemic hypotension, which may worsen the shunting. The most selective vasodilator used currently is inhaled nitric oxide (NO), which has had mixed success in improving the outcome of these infants. Some infants cannot be stabilized preoperatively, and continue to have worsening pulmonary function despite optimal medical management. In some centers, these children are placed on extracorporeal membrane oxygen (ECMO) support in an attempt to “rest” the lungs and allow for resolution of the pulmonary hypertension.

After stabilization, the CDH is surgically repaired. This is usually performed through a subcostal incision on the ipsilateral side. The herniated viscera are reduced through the diaphragmatic defect and returned to the abdomen. If there is adequate diaphragmatic tissue available, then a primary repair is performed. In many cases there is minimal or no diaphragmatic muscle available, precluding primary repair. In these instances the diaphragm is reconstructed using an artificial substance such as Gore-Tex.

There are currently two major centers that are involved in the prenatal surgical treatment of high-risk CDH fetuses (University of California at San Francisco and the Children’s Hospital of Philadelphia). Both centers use techniques ranging from primary repair of the hernia to tracheal occlusion with augmentation of lung growth and postnatal diaphragmatic repair. While still highly experimental, both groups have had successfully treated cases.

**OUTCOME**

The overall survival rate for all infants with CDH ranges from 50 to 70%, depending to a large degree on the center in which they are managed. Children who survive infancy will usually enjoy normal growth and development, although many continue to have subclinical abnormalities on pulmonary function testing. A significant number of survivors will develop gastroesophageal reflux, with many requiring fundoplication, or a variety of pectus-type chest wall disorders, which may require correction later in life.

**NECROTIZING ENTEROCOLITIS**

Necrotizing enterocolitis (NEC) is the most serious and frequent gastrointestinal disorder seen among premature infants, affecting approximately 2000 to 4000 infants per year. It is a disorder of survivors – extremely premature infants who would not have survived the first hours of birth as recent as 20 years ago.

**PATHOPHYSIOLOGY**

The cause of NEC is not known. The patient population at highest risk includes premature infants and infants with other severe coexisting diseases, such as cyanotic congenital heart disease. The most common proposed etiologies include relative intestinal ischemia and the effects of pathogenic bacteria on the immature neonatal gut. Premature infants are known to
have an incompletely developed gut barrier defense, placing them at an increased risk for intestinal-associated sepsis. Due to other alterations in host defense, such as lack of breast feeding, exposure to pathogenic and resistant bacteria in the ICU setting, and the alteration of normal gut flora by the widespread use of antibiotics, these infants become susceptible to local mucosal injury and inflammation from an abnormal bacterial load, and develop the clinical picture of NEC. It is likely that the true basis for the development of NEC is multifactorial, and that no single etiology exists.

**Pathology**

The most common site of involvement is the terminal ileum and right colon; in up to half of affected infants there will be multiple segments of intestine involved, and 20% of infants will have pan-necrosis of the bowel, where more than 75% of the gut is destroyed. Gross findings include marked distension of the bowel; discoloration of the serosa; subserosal pockets of gas; and frank necrosis and perforation.

**Clinical Symptoms**

NEC may present as a subacute illness or as a rapidly fulminating septic state. Most commonly, one will see a premature infant who has been doing well who suddenly becomes distended and lethargic, and who will display other subtle findings of sepsis including apnea and bradycardia, hypothermia, poor perfusion, and increased gastric residuals. In advanced cases the abdomen may appear erythematous, with palpable thickened bowel loops. Gross rectal bleeding is uncommon but may occur.

**Laboratory and X-ray Findings**

Infants with NEC will frequently display a variety of laboratory abnormalities, including leukopenia, thrombocytopenia, and progressive metabolic acidosis. The characteristic x-ray finding in NEC is pneumatosis intestinalis, although the most common finding is non-specific dilatation of the bowel. Pneumatosis appears as “soap bubbles” scattered throughout the wall of the bowel in either a cystic or linear form, and is a very characteristic finding. Portal vein gas may be seen, which carries a poor prognosis. Ascites may occur and generally is associated with severe necrosis and a poor outcome. Finally, pneumoperitoneum (“free air”) may be found and signifies that an intestinal perforation has occurred.

**Non-surgical Treatment**

Unless there is evidence of intestinal perforation or necrosis, the initial treatment for NEC is nonoperative. The mainstay for medical treatment of uncomplicated NEC is cessation of enteral feedings; nasogastric decompression and bowel rest; broad-spectrum antibiotics; and correction of hypovolemia and other derangements of sepsis as needed. The progress of the disease is then monitored by frequent physical examinations, abdominal x-rays, serial blood counts, and
serial blood gas determinations. Indications for surgical treatment are frequently subjective and include failure to improve on medical therapy alone; progressive hemodynamic instability; progressive metabolic acidosis; worsening thrombocytopenia/leukopenia; abdominal wall erythema or crepitance; and a “fixed”, or non-motile loop of intestine on KUB or physical examination. Absolute indications for surgery include portal venous gas or pneumoperitoneum.

Infants who improve with medical therapy will usually require 7 to 10 days of conservative treatment prior to reinitiating oral feeds and stopping antibiotics. Once feeds are started, they are slowly advanced in order to minimally stress the bowel.

**Surgical Treatment**

Infants who fail medical treatment (~1/3) will in most cases require laparotomy. There are several important principles behind the surgical management of NEC; these include:

1. conservation of bowel length
2. resection of obviously necrotic bowel
3. creation of a stoma proximal to involved bowel
4. preservation of the ileocecal valve if at all possible

A significant number of infants will have pannecrosis of the bowel. This can be very difficult to deal with, as it is not always possible to determine how much bowel will ultimately survive in these cases. In this setting, many surgeons will not resect any bowel, and will allow the child to expire. Another option is to not resect bowel but to create a very high jejunostomy in order to protect the distal intestine in the hopes that some bowel will recover; this strategy has produced some survivors, but the majority of such infants will ultimately succumb to sepsis or liver failure as a result of requiring long-term parenteral nutrition. For infants who undergo surgery and survive, most will require closure of their stoma within a few months of surgery. Prior to closing the stoma, a distal contrast study should be performed through the defunctionalized intestine in order to detect the presence or absence of distal strictures, which can occur in up to 15% of cases.

**Outcome**

There has been a steady improvement in the outcome of children with NEC over the past two decades. Currently, infants who do not require surgery will enjoy a survival of over 95%, while those requiring surgery have an approximately 75% survival, with the majority of fatalities seen in infants with pannecrosis. Most infants with pannecrosis can still be expected to die of their disease, either in the immediate perioperative period or due to the long-term complications of short bowel syndrome.
DUODENAL ATRESIA

Duodenal atresia is the most common form of intestinal atresia seen in newborn infants. It is seen in approximately 1 in 10,000 live births, and may be seen in up to 30% of children born with trisomy 21.

EMBRYOLOGY

The duodenum undergoes a solid phase of development early in gestation, and then recanalizes with further growth of the gut. The etiology of duodenal atresia and stenosis is probably related to a failure of recanalization of the duodenal lumen from its solid cord stage during the 8th to 10th week of development. This is also a point in gestation where other closely related organ systems are developing, such as the biliary tree, the pancreas, and the rotation of the bowel, and it is common for duodenal obstruction to be seen in association with anomalies of these organ systems.

CLASSIFICATION

Duodenal obstruction may be caused by several types of duodenal anomalies, including:

- complete atresia, with continuous or discontinuous muscular wall
- duodenal stenosis, with a narrowed but patent duodenal channel
- “windsock” deformity, with an elongated mucosal membrane and a small mucosal opening.

CLINICAL PRESENTATION

Duodenal atresia is routinely diagnosed by prenatal ultrasonography and appears as the classic “double-bubble” consisting of a dilated stomach and a dilated proximal duodenum; many of these pregnancies are complicated by polyhydramnios. After birth, most infants will develop bilious vomiting. Since the obstruction is a “high” obstruction, the remainder of the gut does not become dilated, so abdominal distension is not usually present. Some patients with duodenal stenosis may not be diagnosed until much latter in life, since they are not completely obstructed; however, the partial obstruction usually produces some type of symptom, and eventually these patients are discovered when they continue to have vomiting or weight loss. Duodenal atresia is associated with a number of other anomalies, which must be ruled out early in the course of treatment. These include Down’s syndrome; congenital cardiovascular disorders; malrotation; annular pancreas; jejunal atresia; and imperforate anus.

RADIOGRAPHIC DIAGNOSIS

The classic xray finding in duodenal atresia is the “double bubble” with an absence of gas in the remaining intestine; this sign is virtually diagnostic and additional contrast studies are not required. This may be difficult to differentiate from the infant with a high-grade obstruction secondary to malrotation and midgut volvulus, although the latter will generally have some evidence of air within the remaining
intestinal tract. While an upper gastrointestinal series will confirm the diagnosis, this is not usually needed in the presence of classic plain film findings.

**TREATMENT**

Duodenal atresia is not a surgical emergency, and adequate time should be taken to prepare the infant for the operating room and to identify associated anomalies. Early treatment should include nasogastric decompression, replacement of fluid and electrolyte losses, and intravenous antibiotics. Once the child is stabilized, then surgery can be performed. The standard surgical treatment for duodenal atresia includes a primary anastomosis between the duodenum above and below the site of atresia, known as a duodenoduodenostomy. If the proximal duodenum is tremendously dilated, then a partial resection of its antimesenteric border can be performed, known as a tapering duodenoplasty. Duodenal stenosis is treated by performing a duodenoplasty directly through the stricture area, usually through a longitudinal duodenostomy which is then closed transversely; however, if an annular pancreas is present, a side-to-side duodenoduodenostomy is created in order to not injure the ectopic pancreas and risk a pancreatic leak. The windsock deformity entails making a lateral duodenotomy, and then excising the neck of the mucosal membrane taking care to avoid injury to any anomalous biliary openings. It is important to look for associated distal small intestinal atresias, which may be present in up to 10% of cases. Postoperatively, return of gastrointestinal function may take 10 to 14 days, at which point the infants are generally started on an enteral diet. Current survival for duodenal atresia is well over 90%, with most fatalities occurring to associated anomalies.

**INTESTINAL ATRESIA**

Unlike duodenal atresia, jejunal and ileal atresias occur relatively late in gestation and are usually not associated with other congenital disorders. These defects are thought to develop as a result of some form of mesenteric vascular accident, such as microemboli or volvulus, with infarction of the affected portion of the gut. Since the fetal GI tract is sterile, the infarcted bowel simply resorbs, producing a point of obstruction. However, cystic fibrosis is associated with the presence of intestinal atresia in 15% of cases and should be ruled out. Jejunoileal atresias are seen in approximately 1 in 4000 live births.

**CLASSIFICATION**

The current classification system is the Martin-Zarella classification, which divides atresias into 5 separate types based on their morphological appearance. The majority of atresias are single, but multiple atresias are seen in up to 15% of patients and must be sought for. Two types of small intestine atresias are noteworthy: type IIIb, or "apple peel" atresia, and type IV, multiple atresias. Both types are associated with an increased incidence of short gut syndrome due to the
loss of a large amount of small bowel. In addition, type IIIb atresia is associated with a significant risk of developing postoperative necrotizing enterocolitis.

**CLINICAL PRESENTATION**

Infants with intestinal atresia usually present with evidence of intestinal obstruction, including abdominal distension and bilious emesis. Decreased passage of meconium is suggestive of intestinal obstruction, but passage of meconium does not rule out an obstruction. Many infants are diagnosed prenatally by the finding of polyhydramnios in the mother, and multiple dilated loops of small bowel on fetal ultrasound. Patients may not be symptomatic until several days of life, especially for infants with a very distal ileal atresia. Abdominal x-rays will show distended loops of small intestine, and one may get an idea of the level of obstruction based on the relative amount of dilated bowel visible. Abdominal calcifications may be present and suggest that a prenatal bowel perforation occurred, with the development of meconium peritonitis and small bowel atresia.

**DIAGNOSIS**

The diagnosis of small intestine atresia is primarily clinical and based on the findings above. For the infant who clinically is obstructed, with few loops of dilated small bowel on KUB, the diagnosis of a proximal intestinal atresia can be made with some certainty, and preparation for surgical treatment can be initiated; further contrast studies are not necessary. For the infant with a large amount of dilated bowel, the differential diagnosis becomes more complex, including Hirschsprung’s disease, meconium ileus, and other disorders that may not need an operation immediately. These infants are best evaluated with a distal contrast enema using water-soluble contrast agents such as gastrograffin. If contrast cannot be refluxed up beyond the point of obstruction into dilated small intestine, the diagnosis of a distal atresia is probable, and preparations for surgery are made.

**TREATMENT**

The treatment for small bowel atresia is surgical. Prior to surgical repair, the infant must be resuscitated with fluids, started on intravenous antibiotics, and decompressed with a nasogastric tube. During laparotomy, the two ends of the atretic bowel are identified. Typically, there is a very large size discrepancy between the very proximal bowel and the contracted distal bowel. However, a primary anastomosis between the two ends can almost always be performed, and it is unusual to need to create a stoma. The surgeon must evaluate the distal bowel for a second atresia; this is best performed by injecting saline into the open end of the distal bowel and assuring that it can be milked into the cecum. It is also important to measure and record the length of remaining intestine if it appears that a significant length of gut is absent. Postoperatively,
these children can have a very delayed return of bowel function, and usually will require parenteral nutrition for one to two weeks.

MALROTATION AND MIDGUT VOLVULUS

Intestinal malrotation occurs as a result of failure of the bowel to return to the abdomen and become properly fixated early in embryonic life. After the physiologic herniation of the fetal intestine through the umbilical ring at 6 weeks of life, by 12 weeks the bowel returns to the abdomen in an orderly fashion, undergoing an approximately 270° counterclockwise rotation based on the superior mesenteric artery. The cecum becomes fixated in the right lower quadrant, and the beginning of the small bowel (the duodenojejunal flexure) becomes fixated in the left upper quadrant; the net effect of this is to create a broad-based mesentery, which decreases its chance of “twisting”, or developing a volvulus. Since the blood vessels to the intestine run within the mesentery, a volvulus will produce an obstruction of the blood vessels with eventual ischemic injury to the involved intestine.

CLINICAL PRESENTATION

The majority of patients who develop midgut volvulus secondary to malrotation will become symptomatic within the first few months of life. The predominant physical finding is bilious emesis, suggesting an obstruction distal to the ampulla of Vater.

_BILIOUS EMESIS IN AN INFANT IS A SURGICAL EMERGENCY UNTIL PROVEN OTHERWISE!_

Some children may develop chronic symptoms of recurrent emesis that may be overlooked. Other children will have nonspecific abdominal pain, non-bilious emesis, or failure to thrive. Older children are more likely to not present with emesis but instead with chronic abdominal pain, and are discovered to have malrotation during their evaluation. Infants who are diagnosed after ischemic injury occurs to the intestine will frequently have abdominal pain, abdominal wall erythema, and rectal bleeding and cardiovascular collapse.

DIAGNOSIS

The diagnosis of malrotation is based on upper gastrointestinal contrast studies. A normal upper GI study should show the duodenum with a normal rightward curve, which then crosses the vertebral column and ascends to the ligament of Treitz at a point to the left of the spine and level with the pylorus. Intestinal malrotation will typically have the small bowel extending down the right side of the abdomen, without the duodenum crossing the midline; the bowel itself may spiral or “corkscrew”, suggesting the presence of a volvulus. An abrupt cutoff at the duodenal level may imply the presence of dense Ladd’s bands or signify the presence of a complete duodenal obstruction secondary to volvulus. A lower GI study may show the cecum to be on the left side of the abdomen, but may also be normal, and thus is not typically used to diagnose malrotation. Ultrasound of the abdomen has been used to demonstrate a reversal of the positions of the superior mesenteric artery and vein, again suggesting a twisting or
volvulus of the mesentery. The current gold standard, however, is a contrast upper GI. Plain film findings are extremely nonspecific and should never be used to “rule out” a malrotation.

**TREATMENT**

The treatment for malrotation with midgut volvulus is immediate surgical correction; bowel that is ischemic can only remain viable for a few hours before irreversible injury occurs. The procedure is called a Ladd’s procedure, in honor of the father of pediatric surgery and the first surgeon to describe its technical details. During a Ladd’s procedure, the bowel is eviscerated and inspected. If it has undergone a volvulus, the bowel is then untwisted in a counterclockwise fashion to relieve the vascular obstruction. Ladd’s bands, which are avascular peritoneal attachments from the cecum to the right upper quadrant, are lysed and the duodenum is mobilized. The mesentery of the bowel is then dissected and broadened, and the appendix is taken out. 10% of infants will have some form of associated intrinsic duodenal obstruction, and this must be evaluated and treated. The bowel is then returned to the abdomen in a nonrotated fashion, with the duodenum and proximal small intestine draped down the right side of the abdomen and the cecum and colon placed into the left upper quadrant. Most infants can start eating in a couple of days, and recurrences are uncommon.

Occasionally a child will have already undergone significant intestinal ischemic injury prior to surgery, and at laparotomy one may discover a large amount of questionably viable bowel. The correct procedure at this point is to untwist the bowel and attempt to determine its viability clinically. If the bowel remains of questionable viability, one may resect it, which frequently produces the short gut syndrome. A better approach is to replace the bowel into the abdomen and then support the child for 24 to 48 hours with fluids and antibiotics. At this point the child is taken back to the operating room for a “second-look” at which point the clearly non-viable bowel is resected. For infants and children who have lost all of their midgut, historical survival is uncommon, although this may be improved in the current era of bowel transplantation.

**ABDOMINAL WALL DEFECTS**

The two most common abdominal wall defects seen in neonates are gastroschisis (Greek: *gastro-* = abdomen, *-schisis* = cleft), and omphalocele. While these two disorders share many similarities in their treatment, they are unrelated from an embryological standpoint and have a significantly different outcome.

**OMPHALOCELE**
Omphalocele represents a failure of closure of the umbilical ring at approximately 12 weeks of gestation. This allows for the persistent herniation of the abdominal contents through the abdominal wall, although they are almost always covered by the intact amnion and other fetal membranes. The omphalocele may vary in size, from being nothing more than a large umbilical hernia to a complete absence of the abdominal wall. There is almost always a large amount of small and large intestine herniated into the sac, and frequently the liver is herniated as well. Rarely the omphalocele sac will rupture prior to birth, at which point the defect may resemble a very large gastroschisis; however, a good rule of thumb is that if the liver is exiting out of the abdominal wall, then the defect is an omphalocele. These infants have a significant risk of having other associated congenital abnormalities, namely cardiac defects and chromosomal anomalies. There is an association between omphalocele and the Beckwith-Wiedemann syndrome, which is important in the newborn period due to their risk for developing neonatal hypoglycemia. The mortality of omphalocele is chiefly related to the presence of associated congenital defects.

**GASTROSCHISIS**

Gastroschisis represents a later gestational event in which the abdominal wall develops normally, only to rupture and allow herniation of the abdominal contents into the amniotic space. The defect is always to the right of the umbilicus, and may be separated from the umbilicus by a normal skin bridge. There is no covering sac, and the bowel is frequently thickened and edematous due to chronic exposure to amniotic fluid. Compared to the defect seen in omphalocele, the abdominal wall defect in gastroschisis is very small. There is no increased risk of cardiac or chromosomal defects, and overall survival is better than that seen in omphalocele. Due to the increased risk of bowel trauma, infants with gastroschisis have an approximately 15% risk of having a small intestinal atresia, and the rare infant will have suffered a complete prenatal volvulus of the bowel with resultant short gut syndrome.

The primary abdominal pathophysiologic problem with both abdominal wall defects is the lack of intraabdominal space to allow for a safe reduction of the viscera and abdominal closure, known as the loss of abdominal domain.

**DIAGNOSIS**

Both conditions are readily diagnosed by prenatal ultrasound. In the case of omphalocele, the prenatal diagnosis of other associated anomalies, including cardiac and neural tube defects can also be made; after birth, the diagnosis can usually be readily made on physical examination. Other associated anomalies should be sought for, particularly in the case of omphalocele, and these can usually be achieved with physical examination, echocardiography, and chromosomal analysis.
TREATMENT

The ultimate goal of the treatment of these two conditions is return of the abdominal viscera and effective closure or reconstruction of the abdominal wall. Infants born with gastroschisis will require immediate protection of the bowel from further injury, including a reduction of any postnatal volvulus that may have occurred. These infants may be dehydrated, and will usually require vigorous fluid replacement. The bowel should be untwisted and covered with a moist, sterile gauze; our practice then is to place the infant’s lower torso into a sterile “bowel bag” to minimize fluid evaporation and heat loss. The infant should be positioned in the right lateral decubitus position, which minimizes any kinking of the superior mesenteric vessels as they exit through the defect. Since 2000, the initial treatment for gastroschisis has evolved to our current practice of immediate bedside silo placement using sedation only, followed by sequential silo reduction and abdominal wall closure within 5-7 days. This has produced outcomes which are clearly superior to the older “emergent” closure techniques, and produces an excellent cosmetic and functional outcome.

Infants with omphalocele can generally be treated the same as a gastroschisis, except that there is usually no need to protect the bowel at birth. For those infants who are otherwise healthy and who have a small defect, primary closure can almost always be achieved. For those infants who are “tight”, the delayed secondary closure after silo placement is usually successful. Occasionally a child will be born with severe associated cardiac or pulmonary disease, and even delayed closure may have a deleterious effect on their underlying disease. In these situations, some surgeons have advocated “painting” the omphalocele membrane with some form of scarifying substance, which will eventually produce epithelialization of the membrane. While this almost always results in a ventral hernia, it allows the child to recover from their associated anomalies and then undergo abdominal wall repair at a later date.

The overall survival rate for infants with gastroschisis is better than 90%, with most mortality due to the complications of short gut syndrome, parenteral nutrition, and TPN-related liver disease. The survival of children with omphalocele varies between 75% and 90%, and is primarily related to the presence of associated congenital anomalies.

CONGENITAL AGANGLIONOSIS (HIRSCHSPRUNG’S DISEASE)

Hirschsprung’s disease is one of the most common causes of lower intestinal obstruction in the newborn. The clinical picture develops due to a failure of complete migration of neural crest derived ganglion cells to the distal bowel during early embryologic development; since these ganglion cells are responsible for mediating intestinal relaxation, the distal bowel remains contracted, producing a functional obstruction at the point where ganglion cells are absent (the “transition zone”).
PATHOLOGY

While the transition zone may occur anywhere in the gastrointestinal tract, in 75 to 80% of infants it occurs in the rectosigmoid region, and presents with a distal colonic obstructive picture. In 10% of infants, the transition zone may be found proximal to this point but still in the colon, while a smaller percentage will have more proximal disease, such as total colonic aganglionosis; rarely, an infant will present with total intestinal Hirschsprung’s disease, which is usually fatal without intestinal transplantation. The classic histological finding in Hirschsprung’s disease is the absence of ganglion cells in the myenteric plexi of the affected bowel. Another classic, but not pathognomonic, feature is the presence of hypertrophied neural fibers and a very thickened muscularis. Special stains have been used to aid in the confirmation of the disease, with the most commonly used being the presence of increased staining using the acetylcholinesterase immunoperoxidase stain. Most pediatric surgeons will accept the diagnosis of Hirschsprung’s disease in the absence of ganglion cells and the presence of increased acetylcholinesterase activity.

EPIDEMIOLOGY

Hirschsprung’s disease occurs in approximately 1 in 5000 live births, with a strong male predilection of 4:1; however, in patients with long segment disease the male to female ratio is closer to 1:1. There may be a significant family history in some patients, and subsequent siblings of affected patients have a increased risk of having the disease.

CLINICAL PRESENTATION

Over 50% of infants with Hirschsprung’s disease are diagnosed in the neonatal period, with the majority of the remainder being diagnosed before 2 to 3 years of age. The clinical presentation tends to follow two major patterns: 1) newborn bowel obstruction, frequently with a septic picture and enterocolitis, or 2) chronic constipation in the older child. Over 95% of newborns with Hirschsprung’s disease will fail to pass meconium in the first 24 hours of life, and will present with abdominal distension and vomiting; if untreated, these infants will usually develop a watery, foul-smelling diarrhea, abdominal tenderness, and sepsis. A significant number will go on to develop intestinal perforation if not treated. Children who are not diagnosed in the newborn period will usually be diagnosed during the evaluation of chronic constipation. These children may have failure to thrive, and generally present with a large, impacted colon (hence the other name for Hirschsprung’s disease – congenital aganglionic megacolon). Rarely, an individual will not be diagnosed until the teenage years or even adulthood.

DIAGNOSIS

The early diagnosis of Hirschsprung’s disease requires a high degree of suspicion in any infant who presents with delayed passage of stools, constipation, or evidence of intestinal obstruction. In the past, the initial diagnosis was based primarily on the demonstration of a transition zone on barium enema, and this is still used to help define the extent of disease.
However, most pediatric surgeons will require histologic confirmation of the disease prior to surgical therapy. This is usually obtained by performing a transanal suction rectal biopsy, which is a very accurate and well-tolerated procedure which can be done at the bedside or in the office; another option is to perform a full-thickness rectal biopsy, which must be done in the operating room under anesthesia. Histologic requirements for the diagnosis of Hirschsprung’s disease include the absence of ganglion cells coupled with the finding of increased acetylcholinesterase staining. If the child is undergoing an emergent laparotomy for enterocolitis or perforation, then the diagnosis can be made by obtaining seromuscular biopsies of the affected bowel and looking for ganglion cells using a frozen section method.

**TREATMENT**

The classic surgical treatment for Hirschsprung’s disease included a leveling colostomy (i.e., identifying the level of the transition zone and performing a colostomy at that point), definitive pullthrough procedure, and then closure of the colostomy. This evolved into a two-procedure method of placing a leveling colostomy at the initial operation, followed by a pull-through without a protective colostomy. This two-step method is still used, and indeed is probably the safest course of action in the septic infant or in the infant whose colon cannot be adequately decompressed after diagnosis. Currently, most pediatric surgeons are performing a single-stage pullthrough operation, which is performed in the neonatal period in otherwise healthy infants who can be adequately decompressed preoperatively.

There are a variety of pullthrough operations used for Hirschsprung’s disease, with the most common being the Soave endorectal pullthrough, the Swenson coloanal pullthrough, and the Duhamel procedure. Our preference is the Soave procedure, although all procedures have essentially the same long-term outcome. The basic principle of any surgical procedure for Hirschsprung’s disease is the removal of aganglionic bowel and the approximation of ganglionic bowel to within 1 cm of the anal canal.
**IMPERFORATE ANUS AND ANORECTAL ANOMALIES**

Imperforate anus is a generic term that refers to any of the number of the developmental anomalies of the anorectum, many of which involve the total lack of an external opening of the lower bowel (hence the term "imperforate). Imperforate anus anomalies occur in approximately 1 in 5,000 live births, and are seen more commonly in males than females. There is a wide spectrum of presentation, from the child with an ectopically placed but patent anal opening, to the total lack of an external opening with a fistulous connection to the genitourinary system.

**EMBRYOLOGY**

The anorectum develops very early in embryogenesis by a complicated series of tissue foldings, migrations, and dissolutions which convert the closed embryonic hindgut into the mature bladder, anorectum, urethra, and portions of the external genitalia. Lack of progression of any of these steps can lead to the failure of the rectum to open onto the external surface of the perineum, a persistent connection of the terminus of the GI tract to the urinary system (rectourethral or rectovesicular fistula), or various malformations of the external genitalia. The early embryological development of the anorectum also explains the high number of other congenital anomalies associated with imperforate anus, such as the VACTERL association.

**CLINICAL PRESENTATION**

Imperforate anus is seen more frequently in males, and is also typically more complicated to manage in males. Clinically the disorder can be classified into “high” or “low” categories based on the absence or presence of an external opening or fistula, which allows for decompression of the GI tract. In the current classification system any external opening in males or females is considered a low lesion, except for the cloacal variant seen in females, which is considered a high anomaly. This distinction is important, as all high anomalies will require a decompressive and fully diverting colostomy soon after birth, while most low anomalies can be repaired primarily in infancy without a colostomy. Most males are born with high anomalies, and most females are born with low anomalies.

When an anorectal anomaly is suspected, a gentle but thorough examination of the perineum must be performed. Evidence for an external opening must be carefully sought for, since the opening may be present but just very small and stenotic. Ectopic openings can usually be found near the anal “dimple”, but can also be located anywhere from the base of the penis to the anal dimple in males, or from the posterior vaginal fourchette to the anal dimple in females. This can usually be facilitated by observing the infant over 12 to 24 hours, as any external opening will usually have passed some meconium by that time. Other physical findings that are
important to note will be the overall development of the perineum and external genitalia or meconium presenting from the urethra in males or above the hymen in females (suggestive of a high anomaly). A careful evaluation for other associated anomalies, including the VACTERL association, must be made in each child regardless of the level of the anorectal disorder.

The typical evaluation for any child with suspected anorectal anomaly will require: physical examination; passage of an orogastric tube to exclude esophageal atresia; echocardiography to evaluate for associated cardiac disorders; renal ultrasound to exclude associated obstructive urinary disorders; plain spinal films to evaluate for spinal disorders; and abdominal x-ray to determine the degree of bowel distension secondary to the obstruction. On occasion, it is difficult to determine if a child has a high or low lesion despite physical examination. A dated but still useful radiograph to obtain in this setting is the Wagensteen invertogram, where a true lateral pelvic x-ray is obtained with the child prone and inverted. This allows air in the distal rectum to act as a contrast agent, where it will rise to the most distal part of the bowel. The relationship of the distal bowel to the ossified structures of the pelvis can then be evaluated and can give the surgeon valuable information as to whether the anomaly is high or low. In practice, however, it is generally clear by 24 hours whether the child will require a colostomy or not based on physical examination.
A cloacal anomaly is a special type of imperforate anus seen in approximately 10% of females with the disorder. Cloacal anomalies are the most severe form of imperforate anus, and occurs when the urinary system, reproductive system, and terminal GI tract all empty into one common channel, the urogenital sinus, which opens onto the perineum. These are all considered high lesions, have an extremely high rate of associated, potentially life threatening urological anomalies, and can be very difficult to repair. Cloacal anomalies are characterized on examination by a single perineal orifice and usually poorly developed external genitalia.

**TREATMENT**

With the exception of cloacal anomalies, anorectal anomalies are never an emergency, and the clinician should fully evaluate the anatomy of the defect as well as search for any other more potentially life threatening anomalies. By observing the child over 12 to 24 hours, the true level of the anomaly usually becomes more evident. Low anomalies in the male can usually be repaired in the neonatal period using a procedure known as a posterior sagittal anoplasty. Low anomalies in the female are usually treated by either early posterior sagittal anoplasty, or using early dilations of the fistula to allow for stooling followed by anoplasty within 2 to 3 months. These infants have an extremely good longterm prognosis, and usually are continent later in life. Infants with high lesions will require a diverting colostomy at birth to allow them to decompress their intestinal tract. These infants will then typically have a distal colostogram performed in the next few months to further define the anatomy between the rectum and the urethra, followed by a posterior sagittal anorectoplasty at 3 to 6 months of age. Once the anoplasty has healed, these infants can then have their colostomy closed.

Females with a cloacal anomaly are considered surgical and urologic emergencies, and will typically require an urgent evaluation soon after birth to rule out the presence of an obstructive uropathy, followed by diverting colostomy and urinary diversion if needed. These children typically undergo definitive perineal reconstruction at 6 to 12 months of life with a
procedure known as a posterior sagittal anorectourethroplasty, and frequently will require reconstruction of the vagina at the same time due to severe associated vaginal anomalies. These are typically long and complicated cases, not infrequently requiring 12 to 18 hours of operative time.

OUTCOME

Survival is largely dependent on the presence and severity of associated cardiac and renal anomalies. Longterm fecal continence is dependent on the level of the anomaly as well as the development of the sacrum and other perineal structures. In general, low lesions with normal sacral and coccygeal development are associated with a greater than 90% long term normal fecal continence, while high lesions with poor sacral development may only have a 10-15% continence rate.

HYPERTROPHIC PYLORIC STENOSIS

Pyloric stenosis represents one of the most common abdominal surgical disorders in the term infant. Prior to the advent of surgical correction for this disorder, (Fredet – 1907, and Ramstedt -1912), the majority of infants with pyloric stenosis died of severe dehydration and malnutrition. Today, it is extremely rare for an infant to have a less than favorable outcome after surgical correction.

EPIDEMIOLOGY AND ETIOLOGY

The etiology of pyloric stenosis is unknown. It is considered to be an acquired disorder, in that infants are normal at birth and generally begin to develop symptoms after 2 to 6 weeks of life, although patients are not infrequently symptomatic before or after these time periods. It is seen in about 3 out of 1000 live births, and is more commonly seen in males. There is a strong genetic influence in the presentation of this disorder, with a definite predisposition to firstborn infants with a family history of pyloric stenosis. The etiology is unclear and a variety of proposed causes have been suggested, from increased gastrin levels to disordered nitric oxide synthesis, but no clear cause and effect has been determined.

CLINICAL PRESENTATION

The typical presentation of an infant with pyloric stenosis is the onset of nonbilious emesis at 2 to 8 weeks of age, with a peak occurrence at about 4 weeks of life. The emesis may be minimal at first, but gradually progresses to large amounts of classically “projectile” vomiting. These infants are typically otherwise healthy at first, although delays in diagnosis will generally lead to dehydration and progressive malnutrition. If the diagnosis has been delayed significantly, one may be able to detect an enlarged stomach with visible gastric peristaltic waves in the left upper quadrant.
The classic physical finding in a child with pyloric stenosis is the palpation of the enlarged pyloric muscle, or “olive”. This can be difficult unless the examiner is patient. Since the stomach is usually greatly distended, it is useful to place an orogastric tube to allow for decompression of the stomach, which will greatly facilitate the examination. It is also useful to give the infant a bottle of glucose water, since this will help to calm him during the examination. The examiner should gently flex the infant’s hips in order to relax the abdominal wall. Next, one should gently and slowly palpate the infant’s midepigastrium to right upper quadrant area, progressively placing more pressure on the abdominal contents with the examining fingers. The olive can usually be felt as a distinct, firm, mobile mass in this area, and usually is approximately 1 to 2 cm in diameter. In the hands of an experienced examiner, palpation of a pyloric olive in a child with a suggestive clinical picture is all that is needed to establish a diagnosis of pyloric stenosis.

**Radiological Diagnosis**

An upper gastrointestinal series can frequently be diagnostic in infants with pyloric stenosis, and will generally demonstrate an enlarged stomach with a narrowed and long pyloric channel, with minimal to no passage of barium into the duodenum. This study is particularly helpful in the infant in whom other causes of vomiting may be present, such as malrotation, gastroesophageal reflux, or duodenal stenosis. More recently, ultrasonography has become the “gold standard” for diagnosis of pyloric stenosis when performed by an experienced ultrasonographer. Although each institution will have its own criteria for a positive study, the dimensions typically seen are a pyloric mural (wall) thickness of greater than 4mm and a channel length of greater than 16mm. The downside to ultrasonography is that it is not useful in evaluating for other causes of emesis, so some clinical discretion must be used in deciding upon which test to use.

**Laboratory Findings**

The classic electrolyte abnormality found in children with pyloric stenosis is hyponatremic, hyochloremic, metabolic alkalosis (also known as a “contraction alkalosis”). This occurs due to the loss of sodium, chloride, and intravascular volume. As the kidney attempts to reclaim sodium in the renal tubules, potassium is lost in the urine. Eventually the kidney must exchange protons for sodium ions as the patient becomes progressive hypokalemic and total body potassium depleted. Although the infant has a metabolic alkalosis, the urine will be acidic due to the loss of protons (“paradoxical aciduria”), and will not become neutral until potassium is replaced.

**Treatment**

The initial treatment of pyloric stenosis is aggressive fluid and electrolyte replacement. Infants should be admitted to the hospital and electrolytes drawn. While they should be made
NPO, nasogastric tube decompression is usually not necessary. The initial fluid resuscitation should include a bolus of normal saline (not Ringer’s Lactate!) at a volume of 10-20 cc/kg, depending on the child’s perceived fluid deficit. IV’s are then given as D5W ½ NS without potassium until the child has urinated, at which point potassium should be added to the fluids. Electrolytes are then repeated every 6 to 12 hours until the serum chloride is greater than 90 meq/l and/or the serum bicarbonate is less than 30 meq/l. At this point the child is considered resuscitated and can be taken safely to the operating room.

The surgical procedure for correction of pyloric stenosis is called the Fredet-Ramstedt pyloromyotomy. The classic abdominal incision is a right upper quadrant incision, although most pediatric surgeons will now perform this through an umbilical incision due to its superior cosmetic results. The pylorus is delivered into the wound, and the muscle is split throughout its length, so that the duodenal mucosa protrudes through the myotomy. Complications of this procedure include inadvertent entry through the duodenal mucosa, which can usually be easily repaired. Postoperatively, infants are allowed to start oral feeds starting at about 4 hours after surgery, and are gradually advanced to an ad lib diet. The majority of infants can be discharged to home within 24 hours. Some postoperative emesis is common, but significant emesis is very unusual. Recurrent pyloric stenosis is extremely rare, and is more likely due to an incomplete myotomy.

**BILIARY ATRESIA**

Biliary atresia is a progressive obliterative disorder of the extrahepatic and intrahepatic bile ducts, which presents as obstructive jaundice in the young infant. The incidence is approximately 1 in 15,000 live births.

**ETIOLOGY**

The cause of biliary atresia is unknown. Since associated gastrointestinal anomalies are seen in 15% of patients, an embryologic cause has been suggested. However, infants can clearly be normal at birth, only to develop obstructive jaundice after a few weeks of life, suggesting that this is an acquired disorder. The probable best answer is that biliary atresia may be the final common pathway for a variety of disorders which in the end produce an inflammatory obliteration of the bile ducts.

**CLINICAL PRESENTATION**

The majority of infants who develop biliary atresia are otherwise term, healthy infants. Jaundice may be present soon after birth or develop in the first few weeks of life. Physiologic
jaundice of infancy is extremely common in newborns and must be differentiated from obstructive jaundice, which is almost always pathologic. Obstructive jaundice will usually present with a significant direct component, where the direct bilirubin is at least 10 to 15% of the total serum bilirubin. There is a long differential diagnosis to consider when evaluating a child for pathologic jaundice, including a large number of metabolic, infectious, genetic, and anatomical lesions which must be excluded before considering the diagnosis of biliary atresia, and the student should become familiar with the main points of this differential.

Infants with biliary atresia will present with progressive jaundice in the majority of cases. Since bilirubin and its byproducts are responsible for the pigment in the stool, these patients will classically develop colorless or acholic stools, which are clay-colored and have no dark pigments. The presence of pigment in the stool essentially rules out biliary atresia. With progressive disease, patients will develop cirrhosis; a hard, firm liver; hepatosplenomegaly; and sometimes ascites. Nutritional wasting may occurred in advanced cases, as well as portal hypertension with esophageal varices and gastrointestinal bleeding.

**DIAGNOSIS**

The most important step in the diagnosis of biliary atresia is ruling out other potential causes of non-surgical jaundice. A partial list of the potential causes of pathological jaundice is included below. The major diseases that must be excluded include infections (TORCH organisms), metabolic disorders (galactosemia, thyroid disorders,), genetic disorders (α-1-AT deficiency, cystic fibrosis), primary liver diseases (neonatal hepatitis, Alagille’ s syndrome), or hemolytic anemias. The evaluation for potential surgical disorders can be performed concomitant with these studies, and will generally require a hepatobiliary ultrasound and a hepatic nuclear medicine scan (HIDA or DISIDA scan). In infants with biliary atresia, the intrahepatic and extrahepatic bile ducts cannot be detected by an ultrasound, and the gallbladder is usually contracted and very small; frequently the gallbladder cannot be seen. The nuclear medicine scan is obtained to determine whether any bile drainage occurs into the intestine. In biliary atresia, the liver usually displays good uptake of the radionuclide tracer, but there is failure of tracer to drain into the duodenum. While some surgeons and gastroenterologists feel that a liver biopsy can also help in making the diagnosis preoperatively, our experience suggests that this is infrequently helpful in confirming or excluding the presence of biliary atresia, and we do not routinely obtain them.
Once the evaluation has excluded a nonsurgical cause of the child’s jaundice, and the ultrasound and HIDA/DISIDA scan are consistent with biliary atresia, the next diagnostic test of choice is an operative cholangiogram. This is performed through a small laparotomy incision by placement of a catheter into the gallbladder and attempting to inject contrast into the biliary tree. If the bile ducts cannot be visualized into the liver, the diagnosis of biliary atresia is confirmed and the definitive surgical procedure is performed at the same time.

**Pathology**

Biliary atresia essentially produces a progressive inflammation, fibrosis, and eventual obliteration of the extrahepatic, and frequently intrahepatic, bile ducts. There are several anatomic types described, depending primarily on the degree of patency of the extrahepatic ducts. “Correctable” biliary atresia is seen where the intrahepatic ducts and the very proximal right and left hepatic ducts are patent, with obliteration of the remainder of the extrahepatic ducts; prior to the use of the Kasai procedure, these were the only infants who survived. Most infants (>85%) will have “uncorrectable” disease, where there are no macroscopically patent ducts, although many will have microscopic ducts patent on histological examination. A variant exists where the proximal hepatic ducts are fibrosed, but the gallbladder and common bile duct are patent. Infants with biliary atresia will also frequently have a number of other congenital defects, including cardiac disease, intestinal malrotation, situs inversus, and asplenia/polysplenia complex.

**Treatment**

Prior to 1959, only children with “correctable” biliary atresia could survive. The surgical procedure for this involved creating an anastomosis between the jejunum and the patent proximal hepatic ducts. Kasai and Suzuki first described the definitive procedure for uncorrectable biliary atresia in 1959. In the Kasai portoenterostomy, a limb of jejunum is anastomosed to the surface of the liver where the microscopically patent bile ducts are located (the hepatic plate), allowing for biliary drainage. In approximately 2/3 of patients, the small ducts will allow for adequate early drainage of bile and resolution of the jaundice; in the remainder of these patients, the remnant bile ducts are inadequate in size and bile drainage does not occur.

Of the infants undergoing a Kasai procedure who enjoy early bile drainage, approximately half will go on to progressive bile duct fibrosis and inadequate drainage, with eventual liver failure, while the other half can be expected to have good liver function for a number of years. Many of these children will go on to develop hepatic cirrhosis and portal hypertension due to liver fibrosis despite adequate bile drainage.

Biliary atresia is the leading disease in children who require liver transplantation, and the majority of infants with biliary atresia will eventually require a transplant due to progressive liver failure or cirrhosis. Those children who enjoy an initial good result from their Kasai procedure are
at a distinct advantage when undergoing transplantation in that they are larger and are more likely to receive an appropriate-sized organ.

**INTUSSUSCEPTION**

Intussusception occurs when the bowel (the *intussusceptum*) invaginates into an adjoining portion of the intestinal tract (the *intussusceptiens*), producing a bowel obstruction and localized ischemia of the involved bowel. While in most cases the etiology is unknown, a few patients will be found to have a pathological lead point in the bowel, such as a Meckel's diverticulum or an intestinal mass.

**CLINICAL FINDINGS**

Idiopathic intussusception typically occurs in an otherwise healthy infant between 6 and 24 months of age. Patients will frequently be awakened from sleep by severe intermittent, colicky abdominal pain that alternates between periods where they are relatively asymptomatic. The pain may be followed by repeated bouts of emesis, and if symptoms are prolonged the child may develop bloody, mucous-containing stools (currant jelly stools). Lethargy between painful episodes is a very commonly seen sign. Physical examination will disclose an infant who draws his legs up during paroxysms of pain; a soft tissue mass may be palpable in the right upper quadrant. When the diagnosis is delayed, infants may present with toxicity and evidence of an acute abdomen.

**DIAGNOSIS**

The diagnosis of intussusception can be very subtle, and a high index of suspicion must be entertained in any infant with a compatible history. The primary mode of diagnosis is by radiological examination. Many radiologists now feel comfortable with diagnosing or excluding an intussusception based on abdominal ultrasonography, where the intussusception may appear as a “target” sign or a “pseudokidney”. The classic diagnostic radiological study is the contrast enema, using either barium or air as a contrast agent. Using either agent, the intussusceptum is visualized as a soft tissue mass within the lumen of the bowel, and may appear as a “coiled-spring”.

**TREATMENT**

The diagnosis and treatment of intussusception are closely intertwined, since in the majority of cases the intussusceptum can be hydrostatically reduced during the barium or air contrast study. Once the diagnosis is entertained, the infant should received intravenous fluids and is prepared for a contrast study. While some have recommended administering antibiotics prior to the study, there is no objective evidence that this is required. The contrast study is
performed, and if the presence of an intussusception is confirmed, then an attempt at hydrostatic reduction is made. The contrast agent is insufflated into the rectum under a controlled, monitored pressure, and the abdomen is imaged fluoroscopically. A successful reduction is present when the contrast agent can be refluxed into the terminal ileum with no evidence of an intraluminal-filling defect. This can be repeated several times until completed. If a successful reduction occurs, the child is usually admitted to the hospital overnight for observation. Hydrostatic reduction is successful in over 90% of patients.

For some children, hydrostatic reduction cannot be accomplished, and surgical reduction is required. These children should receive vigorous fluid and antibiotic resuscitation and be prepared for the operating room. Through a right lower quadrant incision, the intussusceptum is gently "milked" from the intussusciens until reduced. In children where manual reduction cannot be performed or where the bowel is of questionable viability, resection of the involved bowel with a primary anastomosis can usually be safely performed.

Older children, or infants with recurrent, recurrent intussusception, are at high risk for having a pathologic lead point such as a Meckel’s diverticulum or small bowel tumor. It is controversial as to whether older children should undergo hydrostatic reduction or primary surgery when diagnosed. If patients in this group are treated with hydrostatic reduction, they should be formally studied with a contrast GI study at some point to rule out the presence of an anatomic lead point.

APPENDICITIS

Acute appendicitis is the most common surgical disorder of the abdomen, accounting for over 250,000 surgical procedures in the United States per year. The lifetime risk for developing appendicitis is between 6% and 9%, with a peak incidence in the adolescent years.

PATHOPHYSIOLOGY

Appendicitis is felt to occur after the development of acute luminal obstruction of the appendix. As the intraluminal pressures rise, tension on the wall of the appendix increases, producing a decrease in lymphatic drainage, and eventually blood flow of the appendiceal wall. This produces localized inflammation and ischemia of the appendix that progresses to gangrene and full-thickness necrosis with eventual appendiceal perforation. The pain of early appendicitis is autonomically mediated, and classically is dull and vague in location. As localized inflammation progresses, the parietal peritoneum becomes inflamed and localized/somatic pain follows. This probably accounts for the classic shift in location of the pain from an early, poorly localized periumbilical discomfort to well-localized, sharp right lower quadrant pain.
CLINICAL FINDINGS

The invariable constant of appendicitis is abdominal pain, and it is nearly always the first symptom to develop. The pain usually migrates from the periumbilical region to the right lower quadrant, but may also be located in the back or the pelvis. Movement such as sitting or coughing frequently reproduces the pain. Many patients will have associated nausea and vomiting, and anorexia is seen in the large majority of patients. While the presence of diarrhea may suggest acute gastroenteritis, many children with pelvic inflammation from appendicitis will develop loose stools due to rectosigmoid irritation.

On examination, there may be nondescript lower abdominal pain in early cases, although most patients will eventually develop well-localized point tenderness over the right lower quadrant (McBurney’s point). Pain in the right lower quadrant can also frequently be reproduced by deep palpation in other areas of the abdomen, which is a particularly valuable sign. While well described, the obturator and Rovsing’s signs are not particularly helpful.

On rectal examination, one may be able to reproduce the tenderness when palpating on the right side, particularly in the presence of a pelvic appendix. In prepubertal females the adnexa and uterus are easily examined via a rectal exam, and a formal vaginal exam is both unnecessary and very uncomfortable in children. However, a rectal exam is usually not necessary and does not contribute significantly to the diagnosis of appendicitis in the majority of children.

If appendicitis is allowed to progress, then most patients will develop diffuse peritonitis or a walled-off abdominal abscess.

LABORATORY FINDINGS

The most common useful laboratory tests in evaluating appendicitis are the white blood cell count with a differential and a urinalysis. The WBC is usually only slightly elevated in cases of uncomplicated appendicitis, whereas it is usually very elevated in those cases where gangrene or perforations have occurred. A significant left shift or bandemia is almost always seen, even in early appendicitis. A urinalysis is useful in excluding a urinary tract infection. Other laboratory tests, such as liver function tests or serum amylase are unnecessary and usually not helpful unless clinically indicated. A urine pregnancy test should always be obtained in adolescent females.

RADIOLOGICAL TESTS

Most cases of appendicitis can be accurately diagnosed by a careful history and physical examination, and in these cases xray studies add little to the diagnosis. The routine use of plain x-rays, ultrasound, or CT scans of the abdomen should not be encouraged since they add unnecessary expense and delays to the treatment of patients with appendicitis. In those patients where the diagnosis is unclear we will selective use
these studies to aid in the diagnosis. Plain x-rays are rarely helpful unless they demonstrate a fecalith-appendicolith, seen in approximately 15% of patients with appendicitis. Ultrasonography has been shown to be very accurate in the hands of a skilled ultrasonographer, particularly when one finds a noncompressible fluid filled structure in the right lower quadrant which is painful upon compression with the sonography probe. Ultrasound is more helpful in excluding other diagnoses, such as ovarian cysts or torsion. CT scans may show a fluid-filled structure in the right lower quadrant and/or periappendiceal inflammation.

**THE OBSERVATION**

The pediatric patient with acute abdominal pain and possible appendicitis represents one of the classic scenarios of physical diagnosis in children. Because appendicitis is so common, it must always be considered in the differential diagnosis of pediatric abdominal pain. However, the large majority of pediatric patients with abdominal pain do not have a surgical disorder. Learning to differentiate those who need surgery from those who don’t remains one of the last “arts” of surgery, in that the goal of treatment is early surgery for those who require it but avoiding a negative operation in children who are otherwise well. Children who have obvious appendicitis require fluid, antibiotics, and surgery – any type of delay in obtaining confirmatory studies is wasted time. In those children where the diagnosis is questionable, admission to the hospital with observation and serial abdominal exams represents one of the mainstays of diagnosis and treatment. Appendicitis typically progresses in its symptoms, making serial observation and examination critical to its diagnosis in many children. In this setting, the diagnosis usually manifests within 24 hours. Using this policy, most pediatric surgeons will have a “negative” appendectomy rate of less than 5%, while the acceptable rate for adult general surgeons is more on the order of 15-25%.

**TREATMENT**

The treatment of appendicitis is surgical. Most patients with uncomplicated appendicitis will present with mild dehydration but are otherwise in good condition. These patients are rapidly hydrated, given intravenous antibiotics, and taken to the operating room for appendectomy. The antibiotic selection should be directed against the organisms expected to reside in the colon, namely anaerobes and gram-negative organisms; our preference for these patients is cefoxitin or ampicillin-sulbactam. Appendectomy can be accomplished either by a right lower quadrant incision, or by laparoscopic techniques. Patients are generally given a dose of antibiotics postoperatively, and can usually be discharged to home within 24 to 48 hours.
Patients with complicated (gangrenous or perforated appendicitis) have a much higher risk of developing postoperative infectious complications due to the contamination of the surgical field. These patients are generally committed to a 5 to 7 day course of IV antibiotics postoperatively.

Neuroblastoma *(under construction)*

Wilm’s tumor *(under construction)*

Branchial cleft anomalies *(under construction)*

**INGUINAL HERNIA AND HYDROCELE**

Inguinal hernias and hydroceles are probably the most common surgical procedure performed in pediatric patients, and will occur in approximately 5 to 7% of the population at large.

**ANATOMY AND EMBRYOLOGY**

The fetal testis develops from the embryonic kidney (the metanephros) within the abdomen, and descends during development. At approximately 7 months of gestation, the testis begins to traverse the abdominal wall through the inguinal canal into the scrotum. As the testis passes through the internal inguinal ring, a diverticulum of peritoneum known as the *processus vaginalis* descends with it; this processus eventually obliterates, although a remnant naturally persists as the *tunica vaginalis* of the testis. While testicular descent obviously does not occur in females, the development of the female inguinal canal is similar to that of the male. In a significant number of infants, the processus vaginalis remains patent, allowing intra-abdominal fluid or structures to pass into the inguinal canal, producing a hernia.

Pediatric inguinal hernias are considered indirect, in that the defect passes through the inguinal canal lateral to the epigastric vessels, but the floor of the inguinal canal (Hesselbach’s triangle) is intact; “adult” hernias are usually direct, in that the floor of the inguinal canal is attenuated, and the hernia defect occurs medial to the epigastric vessels. These anatomic differences dictate the type of repair that is usually needed in pediatric hernias compared to adult hernias. Although they do occur, direct hernias are rare in children.

Hydroceles occur when the processus vaginalis has closed its communication with the peritoneal cavity, but peritoneal sac remains which becomes filled with fluid. These can occur anywhere along the spermatic cord, although the majority occurs around the testicle within the tunica vaginalis. A communicating hydrocele occurs when the hydrocele “communicates” with the peritoneal cavity via a small, attenuated patent processus; this is technically a hernia, although usually only peritoneal fluid can transit the opening into the hydrocele.
CLINICAL PRESENTATION

Inguinal hernias are more commonly seen in males than females, and have a much higher incidence in premature infants. Other conditions which may predispose to an inguinal hernia include abdominal wall defects, connective tissue disorders, infants receiving peritoneal dialysis, and infants who have a ventriculoperitoneal shunt.

The most common presentation of an inguinal hernia is a bulge seen in the inguinal region, scrotum (males), or labia (females). Typically the mass will enlarge with Valsalva maneuver, and may spontaneously reduce when the child relaxes. Uncomplicated hernias are usually nontender and should be relatively easy to reduce. Occasionally one may feel a firm mass in the hernia of a female, and this will usually represent a herniated ovary or adnexa as part of a "sliding" hernia. Hernias that are painful and erythematous and cannot be reduced are incarcerated hernias, and require immediate surgical attention.

Hydroceles typically are fluid filled structures that are most commonly found in the scrotum, but if large enough can extend up to the inguinal canal. They can be small or very large, and frequently are confused with an incarcerated hernia. A chronic hydrocele is nontender and nonerythematous, and does not fluctuate in size. Hydroceles can occur more proximally in the spermatic cord, and when seen in a female are called a hydrocele of the canal of Nuck. A communicating hydrocele will typically show a fluctuation in its size as fluid enters and exits via the persistent peritoneal opening.

TREATMENT

The majority of hernias will not resolve without surgical repair, while most simple (noncommunicating) hydroceles will resolve with observation. The surgical repair of pediatric (indirect) inguinal hernias involves excision of the hernia sac, with a high ligation of the neck of the sac at the internal inguinal ring. In most patients this is performed as a simple outpatient procedure. Incarcerated hernias should be treated emergently, as the incarcerated bowel is at risk for ischemic infarction, which may require resection. Simple hydroceles are usually observed until the child is 12 to 18 months of age, at which point a hydrocelectomy is indicated. Children with communicating hydroceles should undergo repair at the time of diagnosis, since this is essentially a form of hernia and will not resolve without surgical treatment.

Undescended Testicles (Cryptorchidism) (under construction)