

## Chapter 18

# Gastrointestinal Tract

### Esophageal Conditions

- Tracheoesophageal fistula (TEF) and esophageal atresia (EA)
  - Symptoms
    - ▶ Copious salivation (drooling) and aspiration at first feeding
    - ▶ Choking, coughing, respiratory distress, and atelectasis
    - ▶ Aspiration of acidic gastric secretions results in chemical pneumonia (the most common cause of death)
    - ▶ Abdominal distention as the stomach fills with air passing through the TEF
  - Diagnosis
    - ▶ In a newborn, a nasogastric (NG) tube (radiopaque) cannot be passed into the stomach and polyhydramnios is evident (more common with a pure atresia)
    - ▶ A bowel gas pattern can be seen on a chest radiograph and plain abdominal radiographs
      - ▷ Pure atresia will present with a gasless abdomen
      - ▷ A fistula between the trachea and esophagus appears as a gas-filled stomach and bowel
    - ▶ A contrast swallow is not indicated unless the diagnosis cannot be established by other means
    - ▶ A chest radiograph shows the NG tube curled in the blind upper esophageal pouch
  - Initial treatment
    - ▶ Give nothing by mouth
    - ▶ Pass an NG tube (Repogle sump type, low suction)
    - ▶ Provide intravenous (IV) fluids
    - ▶ Elevate the patient's head to minimize aspiration
    - ▶ Administer antibiotics
    - ▶ Evaluate for associated vertebral, anorectal, cardiac, tracheoesophageal fistula, renal, and radial limb (VACTERL) anomalies
    - ▶ Take plain radiographs to rule out vertebral and limb

- ▶ anomalies
  - ▶ Perform an echocardiogram to define anatomy
  - ▶ Perform a renal ultrasound to detect anomalies
  - ▶ Operative repair is best performed at a tertiary medical facility
- Corrosive injury of the esophagus
  - Strong liquid alkalis (hydroscopic) are the most common cause of corrosive injury to the esophagus, coagulating protein and causing liquefaction necrosis (solids are more difficult to swallow and are usually expectorated). Agents become firmly attached to a moist mucosal surface and may involve the entire thickness of the esophageal wall, but perforation is rare
  - Common injury-causing agents
    - ▶ Potassium hydroxide, sodium hydroxide, and sulfuric acid are associated with severe injury
    - ▶ Sodium hypochlorite (bleach) and detergents seldom cause clinically significant injuries
  - Injury to the esophagus is much more common than injury to the stomach in alkali burns, but acidic corrosives will injure the stomach by coagulation necrosis
  - Symptoms
    - ▶ Vomiting
    - ▶ Dysphagia
    - ▶ Drooling, inability to swallow saliva
    - ▶ Abdominal or upper abdominal pain
    - ▶ Burns of the mouth or pharynx
  - Diagnosis
    - ▶ Perform esophagogastroduodenoscopy using a flexible endoscope, with the patient under general anesthesia, within 24 hours of injury
      - ▷ This is the most accurate means of identifying the extent and severity of the burn
      - ▷ Injured areas appear as a whitish coagulum surrounded by an area of hyperemia
      - ▷ The endoscope should be passed only up to (but not through) the level of injury
      - ▷ Avoid instrumentation after 24–36 hours following injury

- ▷ Remember that deep, circumferential burns have a high incidence of stricture
- ▶ Cineesophagram is the most accurate means of assessing the extent of esophageal injury and motility and can help predict later stricture formation
- ▶ Perform chest and abdominal radiographs
- Treatment
  - ▶ If patient does not have a burn, discharge with follow-up in 3–4 weeks
  - ▶ If patient has a minimum to moderate burn, give antibiotics (ampicillin and gentamicin) and perform esophagram within 48 hours
  - ▶ If motility is undisturbed:
    - ▷ Give clear liquids for 72 hours
    - ▷ Advance to regular diet
    - ▷ Stop antibiotics on the 14th day
    - ▷ Discharge with regular follow-ups on the 21st day
  - ▶ In severe cases:
    - ▷ Pass a string at the time of esophagogastroduodenoscopy
    - ▷ Place a gastrostomy tube
    - ▷ Steroids have no therapeutic value; give antibiotics (ampicillin, 150 mg/kg/day)
    - ▷ If the cineesophagram is normal:
      - Give nothing by mouth until the patient can swallow saliva, then clear liquids; advance to a regular diet
      - Stop antibiotics and repeat the contrast swallow; discharge if normal on the 21st day
    - ▷ If the cineesophagram is abnormal:
      - Give nothing by mouth until a gastrostomy tube can be used
      - On the 21st day, stop antibiotics and repeat contrast swallow; if swallow is normal, discharge with monthly follow-up for 1 year; if a stricture is present, start dilatations
  - ▶ Long-term complications
    - ▷ Stricture (treat with monthly dilatations for 1 y)
    - ▷ Achalasia

- Squamous cell carcinoma (latency may be > 20 y)
- Esophageal perforation
  - Etiology is most commonly iatrogenic
  - Symptoms
    - Respiratory distress (newborns and infants)
    - Drooling
    - Subcutaneous emphysema
    - Substernal chest pain
    - Septic shock
    - Pleural effusion
  - Diagnosis
    - Chest radiograph will show pneumothorax, pneumoperitoneum, pneumomediastinum, and pleural effusion
    - Perform contrast study with water-soluble contrast
  - Treatment
    - For traumatic perforation of the cervical esophagus in a newborn, place an NG tube and give broad-spectrum antibiotics
    - For submucosal perforation of the intrathoracic esophagus with symptoms of mediastinitis (increased temperature, increased white blood count):
      - Perform left posterolateral thoracotomy at the sixth intercostal space (for a low esophageal perforations)
      - Administer broad-spectrum antibiotics
      - Perform primary repair of the esophagus if the perforation is < 24 hours old (interpose pericardial, pleural, or strap muscle patch)
      - Place a chest tube for mediastinal drainage
    - For submucosal perforation of the intrathoracic esophagus without symptoms of mediastinitis, antibiotics alone are often sufficient
    - If the perforation is > 24 hours old with marked contamination, treat as above and perform end cervical esophagostomy and gastrostomy; provide mediastinal drainage
- Gastroesophageal reflux
  - Antireflux barriers
    - Several anatomic factors serve as antireflux barriers, including the sling formed by right crus of the diaphragm, the phrenoesophageal membrane, the Angle of Hiss

(formed by the sharp angle between the lower esophagus and the gastric fundus), and the rosette-like configuration of gastric mucosa

- ▶ Physiologic factors that serve as antireflux barriers include the high-pressure zone, which includes the intraabdominal esophagus and a segment of the supradiaphragmatic thoracic esophagus
- ▶ The intraabdominal length of the esophagus (3–4 cm is optimal), intraabdominal pressure, and hormonal and pharmacological factors can also serve as antireflux barriers
- Etiology: reflux is normal in infants until 10–15 months of age, when children assume a more upright posture, ingest a more solid diet, and develop increased lower esophageal sphincter tone
- Symptoms
  - ▶ Recurrent vomiting with failure to thrive, coughing, stridor, and laryngospasm
  - ▶ Recurrent aspiration pneumonia
  - ▶ Asthma-like symptoms
  - ▶ Esophagitis (occult blood loss causing iron-deficient anemia; may result in eventual stricture)
  - ▶ Apnea, sudden infant death syndrome, retrosternal burning (older children)
  - ▶ Sandifer syndrome (voluntary contortions of the head, neck, and trunk associated with reflux esophagitis, producing iron-deficiency anemia)
- Diagnosis
  - ▶ Use cineesophagram, contrast swallow, upper gastrointestinal (UGI) series, or gastric emptying scan
  - ▶ Perform esophagoscopy with biopsies (to diagnose esophagitis and Barrett esophagus)
  - ▶ 24-hour pH-probe testing is the preferred diagnostic study; esophageal pH should be  $< 4.0$ ,  $< 4\%$  of the probe test time
  - ▶ Use a radioisotope “milk scan” to assess gastric emptying time; the normal quantity emptied over 1 hour should be greater than 30%; over 2 hours, quantity emptied should be  $> 80\%$

- ▶ Perform esophageal manometry
- Nonsurgical treatment (80% success rate)
  - ▶ Thicken feedings
  - ▶ Position patient upright
  - ▶ Give bethanechol or metoclopramide (0.1 mg/kg/dose, bid–qid before meals) to increase lower esophageal sphincter tone, oral erythromycin (2–3 mg/kg/dose before meals), an antacid or H<sub>2</sub>-blockers (eg, ranitidine) to decrease acidity, and proton pump inhibitors (eg, omeprazole)
  - ▶ Give frequent, small-volume feeds
- Treatment (surgical)
  - ▶ Nissen fundoplication (360° wrap)
    - ▷ Indications
      - Esophageal stricture (especially if it occurs following a TEF repair and does not respond to dilations)
      - Severe esophagitis (especially with secondary anemia)
      - Barrett’s metaplasia (a metaplasia of esophageal mucosa resulting from chronic irritation secondary to gastroesophageal [GE] reflux)
      - Recurrent aspiration pneumonia
      - Failure of medical management
      - Repeated vomiting with failure to thrive
      - Severe apneic spells
    - ▷ Intellectually disabled children with GE reflux who need a gastrostomy tube for feeding have a high incidence of delayed gastric emptying and may require pyloromyotomy or pyloroplasty at the time of fundoplication; erythromycin (10 mg/kg tid) may increase gastric emptying
- Postoperative complications
  - ▶ Wrap breakdown resulting in recurrent reflux
  - ▶ Excessively tight wrap resulting in dysphagia (usually responds to dilation)
  - ▶ Slippage of the wrap onto the stomach
  - ▶ Herniation of the wrap into the mediastinum
- Strictures

- Etiology
  - ▶ Reflux esophagitis
  - ▶ Corrosive ingestion
  - ▶ Anastomotic scarring (may be aggravated by GE reflux)
- Treatment
  - ▶ Serial dilatations
  - ▶ Antireflux procedure
  - ▶ Local resection (if stricture is short and circular)
  - ▶ Esophageal substitution (as a last resort)
- Types of dilators
  - ▶ Tucker (passed over a string)
  - ▶ Hurst, Maloney (mercury-weighted)
  - ▶ Jackson
  - ▶ Pneumatic balloon

### **Stomach and Duodenum**

- Infantile hypertrophic pyloric stenosis (HPS)
  - Etiology: multifactorial with proven X-linked factor
  - Male-to-female ratio is 4:1 (most common in firstborn male); occurs more frequently in Caucasians than African Americans
  - Symptoms
    - ▶ Usually occurs at 2–6 weeks of age
    - ▶ Nonbilious, projectile vomiting
    - ▶ Ravenous appetite shortly after an episode of emesis
  - Physical examination
    - ▶ Visible peristaltic waves (left to right) during a test feed of glucose water
    - ▶ A palpable olive (hypertrophied pyloric musculature) above and to the right of the umbilicus is felt in 80% of cases
  - Diagnosis
    - ▶ Physical examination findings as above
    - ▶ If physical examination is doubtful, perform an ultrasound
    - ▶ HPS ultrasound criteria for pyloric stenosis are as follows:
      - ▷ Diameter of pylorus > 14 mm
      - ▷ Muscular thickness > 4 mm
      - ▷ Length > 16 mm

- ▶ Contrast UGI series should be performed only if the physical examination and ultrasound are not diagnostic; “string sign” shows a thickened, narrow pyloric canal
- Laboratory tests
  - ▶ Typically, the patient’s electrolytes show a hypochloremic, hypokalemic metabolic alkalosis
    - ▷ Hydrogen and chlorine are lost by vomiting, producing a metabolic alkalosis
    - ▷ Because chlorine is depleted, bicarbonate is reabsorbed with sodium to increase the metabolic alkalosis
  - ▶ Paradoxical aciduria: sodium is conserved because of extracellular fluid loss, and potassium is lost in the urine to compensate for sodium reabsorption
    - ▷ The resulting hypokalemia enhances excretion of hydrogen in the urine
    - ▷ Decreased potassium induces a worsening alkalosis, resulting in a further decrease in potassium and increased hydrogen excretion
    - ▷ This results in paradoxical aciduria, which is a late sign indicating depletion of total-body potassium
  - ▶ Increased bilirubin (indirect) is due to decreased glucuronyl transferase activity
- Treatment
  - ▶ Correct fluid, electrolyte, and acid-base abnormalities using  $D_5\frac{1}{2}$  normal saline plus potassium chloride (2–4 mEq/kg after patient urinates) at 150–175 cc/kg/day
  - ▶ Resuscitation is adequate when patient achieves a urine output about 2 cc/kg/h, electrolytes are normal, and serum bicarbonate is < 27 mEq/L (patients with uncorrected metabolic alkalosis are at risk for dysrhythmias and apnea from general anesthesia)
  - ▶ An anesthesiologist should aspirate the stomach before giving anesthesia to decrease the risk of aspiration on intubation
  - ▶ Ramstedt pyloromyotomy
    - ▷ Surgical approaches include transverse right upper-quadrant incision, periumbilical incision, and laparoscopic pyloromyotomy (a longitudinal incision is made through the hypertrophied muscle to relieve

- the constriction while avoiding mucosal perforation)
  - Postoperative feeding
    - ▶ Initiate small-volume oral feedings of glucose and water at 4–8 hours following operation. For example:
      - ▷ 15 cc oral electrolyte solution every 2 hours × 3, then
      - ▷ 30 cc oral electrolyte solution every 2 hours × 3, then
      - ▷ 30 cc full-strength formula or breast milk every 2 hours × 3, then
      - ▷ 60 cc full-strength formula or breast milk every 3 hours × 3, then
      - ▷ 90 cc full-strength formula or breast milk every 4 hours
    - ▶ If vomiting occurs, wait 2 hours and retry the last volume tolerated
  - Treating complications
    - ▶ Mucosal perforation
      - ▷ Close in two layers using a polyglactin 919 (absorbable, synthetic, braided) suture over an omental patch
      - ▷ Consider a second myotomy at a different site 45° from the first site
      - ▷ Give nothing by mouth and maintain an NG tube on suction for 24 hours before starting feeds
    - ▶ Vomiting is usually secondary to gastritis or reflux and resolves in 1–2 days in almost all cases
    - ▶ In the case of wound infection, prescribe antibiotics
    - ▶ Bleeding may indicate the need for reexploration
    - ▶ Consider incomplete myotomy only if vomiting continues for > 7–10 days postoperatively, is forceful, or follows every feeding. Redo the myotomy at a site 45° from the original site
    - ▶ Arrhythmias and apnea are associated with general anesthesia in patients with uncorrected metabolic alkalosis (ie, serum bicarbonate < 27 mEq/L); monitor pulse oximetry and apnea postoperatively
- Gastric antral web
  - Produces incomplete obstruction with an insidious delay in symptoms
  - Pathology: submucosal web in the distal antrum is covered by gastric mucosa (no muscular layer present)

- Symptoms include postprandial, nonbilious vomiting; failure to thrive; and epigastric pain
- Diagnosis: UGI series
- Treatment
  - ▶ Intraoperatively place a balloon-tipped catheter through the web, inflate the balloon, and gently retract the catheter to identify the site on the gastric wall where the web originates
  - ▶ Make a longitudinal gastrotomy incision over the site, excise a portion of the web, and close the gastrotomy transversely
- Gastric volvulus
  - Pathology: abnormal rotation of one part of the stomach around another
    - ▶ Organoaxial: rotation occurs around a plane joining the hiatus and pylorus
    - ▶ Mesentericoaxial: rotation occurs around a line joining the greater and lesser curvatures (most common)
  - Usually seen in infants
  - Symptoms: Borchardt triad (acute gastric distention, difficulty passing an NG tube, and nonproductive vomiting)
  - Diagnosis must be made early to prevent hemorrhage and necrosis of the stomach. Mesentericoaxial UGI series shows obstructed, upside down stomach, with antrum often in the chest
  - Treatment
    - ▶ Fluid resuscitation
    - ▶ Emergency operation to prevent necrosis; perform Stamm gastrotomy with fixation of the stomach to the anterior abdominal wall
- Gastric perforation
  - Spontaneous neonatal
    - ▶ Neonate is usually healthy in first week of life
    - ▶ Symptoms include poor feeding, abdominal distention, lethargy, and peritonitis
    - ▶ Abdominal radiograph shows massive pneumoperitoneum
    - ▶ Usually occurs on the greater curvature
    - ▶ Treatment

- Give nothing by mouth
- Place NG tube
- Give IV fluids
- Give antibiotics
- Perform 2-layer closure
- Perform gastrostomy for prolonged obstruction or perforation
- Posttraumatic (eg, secondary to an NG tube or postoperative endoscopy)
  - ▶ Diagnose using water-soluble contrast
  - ▶ Usually occurs on the lesser curvature
  - ▶ Treatment
    - Provide IV fluid resuscitation (20 cc/kg bolus Ringer lactate)
    - Place an NG tube under fluoroscopy
    - Give antibiotics (ampicillin plus gentamicin)
    - Perform operative closure (2 layers)
    - Perform gastroduodenostomy (Billroth I operation) if distal stomach is necrotic
- Bezoars
  - Types
    - ▶ Trichobezoar (hair; usually seen in intellectually disabled children)
    - ▶ Phytobezoar (vegetable fibers)
    - ▶ Lactobezoar (milk; caused by improper milk preparation, such as with powdered formulas or concentrated milk requiring reconstitution or dilution)
  - Symptoms: nonbilious vomiting, dehydration, and failure to thrive
  - Palpable epigastric mass is evident on physical examination
  - Diagnosis: UGI, endoscopy
  - Treatment: gastrotomy and surgical removal if patient is symptomatic and endoscopic removal is not possible
- Duodenal obstruction
  - Etiology
    - ▶ Duodenal atresia or web
    - ▶ Annular pancreas
    - ▶ Preduodenal portal vein
  - Anomalies associated with duodenal atresia

- ▶ Down syndrome (30%)
- ▶ Cardiac, renal, central nervous system, annular pancreas, malrotation, and anterior portal vein
- Symptoms
  - ▶ Bilious vomiting after starting feeds
  - ▶ Infants have usually passed meconium by 24 hours of age
- Diagnosis
  - ▶ Plain abdominal radiographs will show “double bubble” sign, representing air in the stomach and duodenum
- Treatment
  - ▶ Emergency operative correction
  - ▶ Malrotation discovered while fixing an intestinal atresia must be repaired (see Intestinal atresias, page 204)
- Preoperative preparation
  - ▶ Place an NG tube; suction to decompress the stomach and duodenum
  - ▶ Administer IV fluid resuscitation (10–20 mL/kg of crystalloid or 5% albumin); urine output should be about 2 mL/kg/h
  - ▶ Administer antibiotics (eg, ampicillin and gentamicin)
  - ▶ Rule out cardiac anomalies
- Operation
  - ▶ Perform transverse, right supraumbilical incision, extending to the midline
  - ▶ If a “windsock web” is present,
    - ▷ Pass a Foley catheter (about size 8 Fr) through the length of the duodenum into the proximal jejunum and slowly withdraw
    - ▷ Perform duodenotomy along the lateral aspect to avoid injury to the ampulla of Vater, which is located medially
    - ▷ Excise the web
  - ▶ In the case of duodenal atresia and annular pancreas
    - ▷ “Kocherize” the duodenum
    - ▷ Perform a duodenoduodenostomy (one layer, diamond-type anastomosis) if possible, or duodenojejunosomy (often required for annular pancreas); never perform a duodenogastostomy, which carries a risk of marginal ulceration

- ▷ If the proximal duodenum is significantly dilated, consider suture plication (interrupted Lembert sutures) or stapled tapering duodenoplasty (using an endostapler) on the antimesenteric side
- ▶ Return of normal bowel function is frequently delayed; consider using a central venous catheter for total parenteral nutrition (TPN)

### **Small Intestine**

- Malrotation with volvulus (rotational anomaly)
  - Anatomy: ligament of Treitz extends from the second lumbar vertebrae to the sacroiliac joint in the right lower quadrant
  - Etiology: mesenteric fixation failure permits the midgut to rotate the last 90° counterclockwise around the narrow mesenteric pedicle, which contains its entire blood supply (superior mesenteric artery and superior mesenteric vein)
  - The risk of volvulus in patients with malrotation does not decrease with age and should be surgically corrected, even in asymptomatic patients
  - Symptoms of midgut volvulus
    - ▶ Bilious vomiting (always rule out malrotation with volvulus and duodenal atresia in a newborn with bilious vomiting)
    - ▶ Abdominal distention is possible
    - ▶ Painful, tender abdomen
    - ▶ Hematemesis, hematochezia
    - ▶ Child appears acutely ill
    - ▶ Sepsis
    - ▶ Shock (hypoperfusion state)
  - Diagnosis of midgut volvulus
    - ▶ Plain abdominal radiographs show dilated stomach (double bubble), gasless abdomen
    - ▶ Ultrasound may be used to determine the relative positions of the superior mesenteric artery and superior mesenteric vein
    - ▶ UGI is only indicated if acute abdominal findings are absent on physical examination, but will show duodenum and jejunum lying to the right of the spine,

entire opacified bowel on the right side, and duodenal obstruction with the proximal duodenum appearing as a corkscrew at the obstruction point

- Treatment of midgut volvulus
  - ▶ Perform a transverse upper-abdominal incision
  - ▶ Eviscerate and inspect the entire small bowel for areas of atresia or perforation
  - ▶ Reduce a volvulus by counterclockwise rotation of the midgut
  - ▶ Divide Ladd bands, which extend from the cecum across the first and second portions of the duodenum to the retroperitoneum at the right gutter
  - ▶ Widen the mesentery (the most important step to prevent recurrence)
  - ▶ Separate the duodenum from the cecum and place the cecum in the left lower quadrant; position the duodenum, the proximal jejunum in the right lower quadrant, and the ascending colon in the left upper quadrant
  - ▶ Perform an appendectomy (because of the potential delay in the future diagnosis of acute appendicitis that results from the new, nonanatomical location of the cecum and appendix)
- Treatment of ischemic or necrotic small bowel
  - ▶ If a short segment is necrotic but the remainder is normal, perform conservative resection and primary anastomosis
  - ▶ If a short segment is necrotic but the remainder is of questionable viability, perform conservative resection with stoma formation
  - ▶ Reexamine in 12–24 hours if large portions of the gut or stomas appear ischemic
- Intestinal atresias
  - General
    - ▶ Most common cause of congenital intestinal obstruction in the newborn
    - ▶ Etiology: fetal gut infarction (mesenteric vascular accidents)
    - ▶ Esophageal atresia
    - ▶ Duodenal atresia

- ▶ Small-intestinal atresia: distal ileum is the most common site; diagnose using UGI series with small-bowel follow through
- ▶ Colonic atresia: represents only 5% of intestinal atresias, most commonly occurring at the transverse colon; diagnose using contrast enema
- Diagnosis
  - ▶ Prenatal: ultrasound will show polyhydramnios (> 2,000 mL total amniotic fluid volume)
  - ▶ Postnatal
    - ▷ Early, bilious (85%) vomiting is associated with duodenal and proximal jejunal obstruction
    - ▷ Delayed (hours to days) vomiting is associated with distal intestinal obstruction
  - ▶ Failure to pass meconium in the first 24–48 hours of life
  - ▶ Presence or absence of abdominal distention depends on the level of the obstruction
  - ▶ Jaundice: increased indirect bilirubin;  $\beta$ -glucuronidase in the intestinal mucosa deconjugates direct bilirubin and enhances enterohepatic recirculation of bilirubin in the presence of bowel obstruction
  - ▶ Radiographs
    - ▷ Double-bubble sign is associated with duodenal obstruction (atresia, annular pancreas)
    - ▷ Triple-bubble sign shows air in the stomach, duodenum, and bowel proximal to the area of atresia
      - Perform limited UGI series to rule out malrotation or volvulus
      - There will be a paucity of gas in the distal bowel
      - Contrast enema will show microcolon; dilated, air-filled proximal bowel
- Treatment
  - ▶ Preoperative
    - ▷ Place an NG tube
    - ▷ Administer IV fluids
    - ▷ Type and cross-match blood
    - ▷ Administer antibiotics
    - ▷ Rule out associated anomalies
  - ▶ Perform operative treatment for duodenal atresia (see

Duodenal obstruction, see page 201), small bowel atresia, or colonic atresia

- ▷ In the case of small bowel atresia, take the following steps:
  - Inspect the entire small bowel for areas of atresia or stenosis
  - Flush saline into the distal bowel to rule out additional areas of atresia or stenosis
  - Resect dilated proximal bulbous tip to avoid postoperative functional obstruction
  - Perform primary end-to-end oblique anastomosis (a stapled, antimesenteric tapering duodenoplasty [duodenal atresia] or enteroplasty [small bowel] of the dilated proximal bowel is preferred over resection of a significant length of bowel)
  - Preserve distal ileum and ileocecal valve, if possible, to prevent vitamin B<sub>12</sub> and fat malabsorption
  - If primary anastomosis is not possible (eg, in premature infants or in the case of peritonitis), perform an ileostomy with exteriorization of both limbs
- ▷ In the case of colonic atresia, use primary anastomosis if possible; use temporary proximal end colostomy if there is a large size discrepancy
- ▶ Postoperative
  - ▷ Place an NG tube
  - ▷ Parenteral nutrition may be required because of the high incidence of prolonged postoperative ileus (lasting weeks to months)
  - ▷ Give elemental formulas (Nutramigen or Pregestimil [Mead Johnson Nutrition, Glenview, Ill])
  - ▷ Mortality is usually associated with complex congenital heart disease (especially Down syndrome with endocardial cushion defect)
  - ▷ Duodenojejunostomy may be associated with blind loop syndrome; treat by converting to a duodenoduodenostomy
- Complications
  - ▶ Sepsis resulting from pneumonia or an anastomotic leak
  - ▶ Functional obstruction

- Meckel's diverticulum
  - Anatomy
    - ▶ Located on the antimesenteric border of the ileum
    - ▶ Associated with omphalocele, malrotation, atresias, and many other congenital anomalies
    - ▶ Called a "disease of twos"
      - ▷ 2 inches long
      - ▷ Occurs 2 inches from the ileocecal valve
      - ▷ 2% of cases are symptomatic
      - ▷ Condition affects 2% of the population
      - ▷ There are 2 potential types of ectopic mucosa (gastric more common than pancreatic)
      - ▷ Occurs as a 2:1 ratio in males to females
  - Symptoms
    - ▶ The most common presentation in pediatric patients < 5 years old is sudden, painless, lower gastrointestinal (GI) bleeding
    - ▶ Occurs secondary to peptic ulceration in the adjacent ileal mucosa
    - ▶ Rarely results in a life-threatening hemorrhage
    - ▶ Causes grossly bloody or occasionally black, tarry stools resulting in anemia
    - ▶ Most common cause of massive rectal bleeding in pediatric patients (esophageal varices secondary to a history of omphalitis is the most common cause of massive hematemesis in this age group, and can be managed conservatively in almost all cases)
    - ▶ In Meckel's diverticulitis, pain is indistinguishable from that caused by acute appendicitis
  - May cause intestinal obstruction by acting as a lead point for intussusception
  - Littre hernia is a Meckel's diverticulum trapped in an incarcerated umbilical or inguinal hernia
  - In a hostile environment, diagnose using ultrasound, computed tomography scan, and clinical observation
  - Complications
    - ▶ Hemorrhage (most common in children)
    - ▶ Obstruction (most common in adults)
      - ▷ A common lead point for ileocolic intussusception

- ▶ Attachment of diverticulum to abdominal wall by fibrous bands
- ▶ Perforation
- ▶ Volvulus
- Treat symptomatic cases with surgical excision (treatment of asymptomatic cases is controversial, but most pediatric surgeons feel that incidental diverticulectomy is not indicated)
  - ▶ In the presence of a narrow base, perform a wedge resection with transverse closure
  - ▶ In the case of a wide base or significant degree of inflammation in the adjacent intestine, perform a sleeve resection with end-to-end ileoileostomy
  - ▶ In the case of intussusception, manual reduction is rarely possible; perform segmental resection
- Necrotizing enterocolitis
  - Most common surgical emergency in the neonate (infants < 30 days old)
  - Usually occurs after the tenth day of life when coliform bacteria colonize the GI tract, but can occur at any time in the neonatal period
  - Pathophysiology
    - ▶ Ischemic intestinal mucosa is susceptible to reperfusion injury and bacterial translocation of gut flora, especially involving *Escherichia coli*, *Klebsiella*, *Clostridium difficile*, *Enterobacter*, and *Proteus*, which produce hydrogen gas within the gut wall (pneumatosis intestinalis)
    - ▶ The most commonly affected sites are the terminal ileum and right colon
  - Histology
    - ▶ Most common pathology is bland ischemic necrosis of the superficial mucosa
    - ▶ Microthrombi occur secondary to platelet aggregation throughout the mesentery
    - ▶ Characterized by “skip areas” of involvement
    - ▶ Pneumatosis intestinalis
  - Reperfusion triggers agents that produce cellular injury (eg, anion radicals, superoxide, hydrogen peroxide)
  - Radiographic findings
    - ▶ Plain abdominal films demonstrate intramural gas

- ▶ In the presence of pneumatosis intestinalis (pathognomonic), hydrogen gas (from *E coli*) will be evident
- ▶ Air in the portal venous system (pylephlebitis) is a poor prognostic sign (however, these findings alone are not indications for operative intervention and are reversible with medical management)
- ▶ Pneumoperitoneum may be present (secondary to GI perforation)
- ▶ Static intestinal loops (persistent loops of adynamic or edematous bowel) can be seen on serial abdominal radiographs
- ▶ The earliest, most common finding is distention of multiple bowel loops
- ▶ Bowel gas is diminished
- ▶ Ascites is a grave sign
- ▶ Abdominal wall ecchymosis is usually indicative of underlying dead bowel
- Physical examination
  - ▶ Patient's first symptom will be feeding intolerance (high feeding residuals)
  - ▶ Abdominal distention is the most common sign
  - ▶ Lethargy
  - ▶ Bilious vomiting
  - ▶ Rectal bleeding (plus guaiac-positive stool)
  - ▶ Coagulopathy
  - ▶ Bradycardia
  - ▶ Apnea
  - ▶ Edema and erythema of the abdominal wall
  - ▶ Thermal instability
  - ▶ Carbohydrate intolerance, causing the presence of reducing substances in the stool
- Laboratory studies
  - ▶ Thrombocytopenia (secondary to microvascular plugging and binding to gram-negative endotoxin) is the most relevant
  - ▶ Leukopenia (especially in severe cases)
  - ▶ Metabolic acidosis (pH < 7.2, with an anion gap), especially with decreased serum sodium
  - ▶ Increased breath hydrogen excretion

- Medical treatment
  - ▶ Nothing by mouth for at least 10 days
  - ▶ NG tube decompression of the UGI tract
  - ▶ IV (not enteral) antibiotics (ampicillin, gentamicin, clindamycin) to cover *E coli*, *Klebsiella*, and *C difficile*
  - ▶ Parenteral nutrition
  - ▶ Serial abdominal radiographs
- Surgical treatment
  - ▶ Pneumoperitoneum is an absolute indication for surgical management
  - ▶ Relative indications for peritoneal drainage or laparotomy include:
    - ▷ Positive peritoneal lavage (brown color, positive Gram stain), which suggests a gangrenous or perforated bowel
    - ▷ Palpable abdominal mass
    - ▷ Progressive peritonitis (edema or erythema of the abdominal wall)
    - ▷ Deterioration on medical management (increasing acidosis, decreasing platelets)
    - ▷ Intestinal obstruction
    - ▷ Fixed, dilated loop of intestine (unchanged after 24 h) on serial abdominal radiographs
    - ▷ Portal vein gas
    - ▷ Ascites
  - ▶ Perform a right, transverse, supraumbilical incision
  - ▶ Conservative resection of frankly gangrenous bowel to prevent short-bowel syndrome
  - ▶ Preserve the ileocecal valve if possible
  - ▶ Take a second look at marginally viable bowel at 24–48 hours
  - ▶ Administer antibiotics and TPN postoperatively
  - ▶ In very small, ill, premature infants, set up peritoneal drainage using a ¼-inch Penrose drain placed while patient is under local anesthesia (this may be an effective temporary or permanent measure in an unstable, high-risk, premature infant with peritonitis, respiratory failure, or shock)
  - ▶ If resection of multiple necrotic segments is necessary,

- perform a proximal stoma and multiple anastomoses of the distal, defunctionalized bowel
- Postoperative complications
  - ▶ Most common complication is stricture due to cicatricial healing of the injured mucosa (usually involving the left colon)
    - ▷ This often presents weeks to months later with a bowel obstruction
    - ▷ Diagnose by contrast enema
    - ▷ Treat with segmental resection and primary anastomosis
  - ▶ Short-bowel syndrome resulting from extensive bowel resection
    - ▷ This may be prevented by conservative resection and “second look” operations
    - ▷ Treatment includes TPN and dietary manipulation
  - ▶ Malabsorption (usually reversible) resulting from extensive mucosal injury
  - ▶ Interloop fistulae: treat with octreotide (3–5  $\mu\text{g}/\text{kg}/\text{h}$  IV) or 1 mg/kg bid (up to 4 mg/kg/8 h)
  - ▶ Survivors of severe necrotizing enterocolitis remain at high risk for growth and neurologic morbidity
- Intussusception
  - Seen in healthy children 6–24 months old, usually due to hypertrophy of Peyer’s patches in the terminal ileum from an antecedent viral infection (eg, rotavirus, reovirus, or echovirus); seasonal incidence (midwinter and early summer) corresponds with upper respiratory infections and GI viral disease
  - Lead points (only 5% of children)
    - ▶ Polyps (juvenile, hamartoma)
    - ▶ Malignant tumors (eg, lymphoma, lymphosarcoma)
    - ▶ Meckel’s diverticulum (most common), appendix
    - ▶ Peutz-Jeghers syndrome
    - ▶ Intestinal duplications
    - ▶ Hemangioma
    - ▶ Cystic fibrosis (due to inspissated feces in the terminal ileum)
    - ▶ Henoch-Schönlein purpura (hamartomas in the intesti-

- nal wall act as lead points for ileal-ileal intussusception)
- Site
  - ▶ Most common site in the pediatric age group is ileocecal and ileocolic
  - ▶ When resulting from postoperative abdominal and thoracic operations, ileal-ileal site is the most common
- Pathophysiology
  - ▶ Proximal portion of bowel (intussusceptum) is drawn into the distal loop (intussusciens) by peristaltic activity
  - ▶ Mesentery of the proximal bowel is strangulated and compressed, leading to venous obstruction that results in edema of the bowel wall; this causes arterial obstruction, leading first to gangrene and then perforation and peritonitis
- Symptoms
  - ▶ Characteristically seen in an otherwise healthy, robust child
  - ▶ Child experiences paroxysms (lasting around 10–15 minutes) of abdominal cramps and intermittent vomiting (may or may not be bilious); screams and pulls legs up to abdomen
  - ▶ Dark red (bloody) mucoid (“red currant jelly”) stools
- Physical findings
  - ▶ Elongated (sausage-shaped) mass in the right upper quadrant, absence of bowel in the right lower quadrant (Dance sign)
  - ▶ Hyperperistaltic rushes during episodes of pain
  - ▶ Symptoms include tachycardia, fever, and hypotension
  - ▶ If peritoneal symptoms are present, perform a laparotomy (do not attempt radiographic reduction)
  - ▶ Radiographic diagnosis: take flat and upright abdominal films; cecal gas shadow will be absent in right iliac region and small bowel obstruction will be evident (air–fluid levels may be present on upright abdominal radiograph)
  - ▶ Ultrasound will show target sign
  - ▶ Perform air enema or contrast enema (will show coiled spring appearance of the lead point)
- Treatment

- ▶ Initial stabilization
  - ▷ Administer IV fluids (10 mL/kg of 5% albumin solution or Ringer lactate)
  - ▷ Give nothing by mouth
  - ▷ Administer antibiotics (ampicillin, gentamicin, and metronidazole)
  - ▷ Place NG tube to suction
- ▶ Laboratory tests: complete blood count, electrolytes, blood urea nitrogen, glucose, and creatinine; type and cross-match blood
- ▶ Attempt hydrostatic reduction using diatrizoate sodium or air enema (do not attempt in the presence of peritonitis or pneumoperitoneum)
  - ▷ Suspend enema bag < 1 meter (~ 3 ft) above the level of the anus
  - ▷ Water-soluble contrast is preferred
  - ▷ Other methods of reduction include insufflated air (preferred; maximum pressure of 80–120 mmHg) or saline enema under ultrasonography
- ▶ After successful reduction, contrast must be seen to pass into the terminal ileum; at the same time, feces and contrast should be expelled
- ▶ If reduction is unsuccessful, perform operative exploration
- ▶ Nonoperative treatment is effective > 80% of the time
- Incidence of recurrence is low (< 10%)
  - ▶ Diagnose using air or contrast enema
  - ▶ After the third episode, operative intervention is indicated because of the high likelihood of a pathologic lead point
  - ▶ If contrast or air reduction is unsuccessful, or if physical examination reveals a tender or rigid abdomen
    - ▷ Perform laparotomy using a transverse, right, lower incision
    - ▷ Attempt manual reduction by gently milking the intussusceptum out of the intussusciens (never pull it out)
    - ▷ Perform appendectomy (usually) and resection of frankly gangrenous bowel or lead point (eg, Meckel's)
    - ▷ Perform primary ileocolic anastomosis

- ▶ Postoperative intussusception is evident by a small bowel obstruction in the early postoperative period (around postoperative day 5)
  - ▷ Most commonly occurs as an ileo-ileal intussusception
  - ▷ Treat with manual, operative reduction
- Meconium disease
  - Meconium ileus
    - ▶ Generalized mucoviscidosis of exocrine secretions, resulting in inspissated meconium that may cause intraluminal intestinal obstruction in a newborn
    - ▶ Affects all exocrine glands and is usually associated with cystic fibrosis
      - ▷ Lung: air passages are obstructed by thick mucoid secretions
      - ▷ Pancreas: pancreatic ducts are obstructed
      - ▷ Intestine: obstruction by thick, viscous mucus as water migrates out of the intestinal lumen
      - ▷ Other organs affected include sweat and salivary glands, nasal mucus membranes, and reproductive organs
    - ▶ Differential diagnosis includes Hirschsprung disease (especially total aganglionosis), hypothyroidism, small left colon syndrome, colonic or ileal atresia, and meconium plug syndrome
    - ▶ Radiographic findings (on abdominal radiograph) include the following:
      - ▷ Multiple distended loops of intestine mimicking small bowel obstruction, but air–fluid levels are rarely present because of the viscosity of the intraluminal meconium and because the bowel is completely filled with fluid or meconium
      - ▷ Course, granular, soap-bubble (“ground glass”) appearance (mixture of air within thick meconium; Neuhauser’s sign)
      - ▷ Prenatal perforation with meconium peritonitis (due to liberated lipases and bile salts) results in calcium deposition (saponification)
      - ▷ Prominent air–fluid levels suggest intestinal atresia or volvulus

- ▷ Ascites and pneumoperitoneum are indicative of postnatal colonic perforation
- ▷ There is no clinical or radiographic evidence of perforation or peritonitis with uncomplicated meconium ileus
- ▶ Nonoperative treatment
  - ▷ Ensure patient is well hydrated
  - ▷ Insert an NG tube
  - ▷ Administer antibiotics
  - ▷ Perform fluoroscopically visualized enema with a hyperosmolar agent (meglumine diatrizoate or *N*-acetylcysteine)
- ▶ Operative treatment
  - ▷ Indicated for “complicated” patients (ie, those with perforation or peritoneal signs, volvulus, gangrene, or atresia) or after a failed attempt at nonoperative treatment
  - ▷ Technique
    - Perform enterotomy at ileum or appendectomy (with irrigation and drainage) through the appendiceal stump (to avoid an anastomosis or ileostomy)
    - Run entire length of bowel to rule out areas of necrosis or atresia; if any are discovered, they should be resected
    - Pass a red rubber catheter proximally and distally
    - Irrigate with saline or *N*-acetylcysteine (mucolytic)
    - Perform primary anastomosis, ileostomy and mucus fistula, or Bishop-Koop end-to-side (proximal dilated bowel to side) or Santuli anastomosis (avoided by irrigating through the appendiceal stump)
    - Administer antibiotics for 3 days
    - Continue NG suction
    - Initiate oral feedings and pancreatic enzymes 5–10 days following operation
- Meconium plug syndrome

- ▶ Newborn infants with this condition sustain intestinal obstruction because the colon cannot rid itself of fetal meconium residue
- ▶ Physical findings include an obstructing meconium mass and colonic hypomotility
- ▶ Differential diagnosis includes Hirschsprung disease, decreased potassium, decreased calcium, and increased glycogen
- ▶ Associated with decreased glucose in infants of diabetic mothers, but not usually associated with cystic fibrosis
- ▶ Symptoms include abdominal distention and colonic obstruction (resulting from plug of inspissated meconium, which is usually whitish gray distally, green proximally)
- ▶ Abdominal radiograph will reveal multiple loops of dilated small bowel
- ▶ Diagnose and treat using diatrizoate sodium enema; operation is rarely necessary (rule out Hirschsprung disease by rectal biopsy)
- Meconium ileus equivalent
  - ▶ Mechanical obstruction resulting from thick, putty-like, inspissated stool in the intestine
  - ▶ Seen at any age beyond the newborn period in patients with cystic fibrosis

### **Large Intestine**

- The appendix arises from the posteromedial border of the cecum and has its own mesoappendix; blood supply flows from the superior mesenteric artery to the right colic artery, then from the ileocolic artery to the appendicular branch
- Acute appendicitis is the most common acute surgical condition of the abdomen; it parallels the amount of lymphoid tissue in the appendix (peak incidence is during the mid-teenage years)
  - Fecaliths, which obstruct the lumen, are the most common cause of luminal obstruction and the most common factor in the etiology of acute appendicitis; other causes include lymphoid hyperplasia, seeds, and worms (eg, pinworms, *Ascaris*)
  - Sequence of events in acute appendicitis

- ▶ Begins with an obstruction of the lumen; normal mucosal secretion continues
- ▶ Distention increases
- ▶ Nerve endings of visceral afferent sympathetic pain fibers are stimulated through the celiac ganglion to the tenth thoracic segment, causing vague, dull, diffuse pain in the midabdomen or lower epigastrium; this pain is referred to the umbilical area (tenth dermatome)
- ▶ The parietal peritoneum becomes irritated by the inflamed tip of the appendix, causing pain to be localized to the right lower quadrant
- ▶ The stimulation of peristalsis leads to cramping
- ▶ Appendiceal distention increases secondary to bacterial proliferation, which produces reflex nausea and vomiting
- ▶ Venous pressure is exceeded while arterial inflow continues, resulting in engorgement and vascular congestion
- ▶ Bacteria from the lumen translocate into the bloodstream
- ▶ Reflex nausea and vomiting result, as does severe visceral pain from the increased distention
- ▶ The appendiceal wall becomes infarcted, resulting in perforation and peritonitis
- Symptoms
  - ▶ Localized pain in the right, lower quadrant is the most important diagnostic finding on physical examination
  - ▶ Pain is initially centered in the lower epigastrium or periumbilical area (serosa of the appendix becomes inflamed and pain localizes to the right lower quadrant after 4–6 hours as a result of parietal peritoneum inflammation)
- Anatomical locations
  - ▶ A long appendix with an inflamed tip results in pelvic pain
  - ▶ Occurrence at the retrocecal (two thirds of all cases of appendicitis occur here) location results in flank or back pain (most commonly), nausea, vomiting, and increased white blood cell count; microscopic hematuria may occur if the inflamed appendix lies near the ureter
  - ▶ Occurrence at the pelvic location results in suprapubic pain from an inflamed tip lying against the bladder,

- which causes urinary frequency and dysuria
- ▷ If there is tenderness on a rectal examination, the inflamed tip is lying adjacent to the rectum
- ▷ An abscess presents with severe urinary symptoms and diarrhea
- ▶ Occurrence at the retroileal location results in testicular pain from irritation of the spermatic artery and ureter; anorexia almost always accompanies appendicitis (if a child is truly hungry, appendicitis is unlikely)
- ▷ Vomiting (1–2 episodes) is frequent
- ▷ Symptoms usually move from anorexia to abdominal pain to nausea with vomiting; if vomiting precedes the onset of abdominal pain, the diagnosis should be questioned (in favor of gastroenteritis)
- ▷ Low-grade fever (> 100°F, 38°C) is rare
- ▷ The individual will experience point tenderness over McBurney's point (at the junction of the lateral one third and medial two thirds of a line from the anterior superior iliac spine to the umbilicus), usually accompanied by guarding and muscle spasm; this is the most important physical finding
- ▷ Individual may also show psoas sign (pain on extension of the right thigh, which, when patient is in side-lying position, stretches the irritated ileopsoas muscle)
- ▷ Obturator sign may also be present (passive internal rotation of the flexed right thigh stretches the obturator internus muscle, producing pain)
- ▷ Rovsing's sign (pain in the right lower quadrant upon palpation of the left lower quadrant) may be experienced by some individuals
- ▷ Cutaneous hyperesthesia may be present in distribution of T10, T11, and T12 segments
- ▷ Preoperative diagnosis should be correct in 85%–90% of cases
- ▷ Symptoms secondary to bacterial toxins and absorption of dead tissue toxins include fever, tachycardia, and leukocytosis
- Laboratory findings
  - ▶ White blood cell count will be about 10,000–18,000, with

- left shift (> 18,000 is consistent with a perforation)
- ▶ Infants and elderly patients may be unable to increase white blood cell count, resulting in a delay in diagnosis and increased incidence of rupture
  - ▶ Perform urinalysis to rule out a urinary tract infection and pregnancy
  - ▶ Ultrasound will show the appendix diameter to be > 8 mm and appendix will be edematous with adjacent fluid (rule out gynecological problems such as ovarian cysts and tubal pregnancy)
    - ▷ A gas-filled appendix usually indicates appendicitis with proximal obstruction
    - ▷ Radiopaque fecalith is almost always associated with gangrenous appendicitis
    - ▷ A distended loop of small bowel will be evident in the right lower quadrant
  - ▶ Barium enema findings are pathognomonic of appendicitis; findings include nonfilling of appendix, mass effect on medial and inferior borders of the cecum, and mucosal irregularities of the terminal ileum
  - ▶ Obtain a chest radiograph to rule out right lower lobe pneumonia
  - ▶ Computed tomography scan will often reveal inflammation, fat stranding, and fluid
  - Appendiceal rupture is a common complication
  - Appendicitis during pregnancy
    - This is the most common extrauterine surgical emergency (though incidence of appendicitis is not increased by pregnancy)
    - White blood cell count normally increases in pregnancy, but a left shift is abnormal
    - The appendix moves superiorly and laterally as pregnancy progresses
    - Laparoscopy may be helpful if diagnosis is uncertain
    - The infant mortality rate from maternal appendicitis is 8.5%; the rate of mortality from maternal perforation and peritonitis is 35%
    - Treat by inserting an NG tube and providing IV fluids and preoperative antibiotics appropriate to cover *Bacteroides*

*fragilis* (gram-negative rod; eg, cefotetan or clindamycin)

- ▶ Nonoperative management may be appropriate to treat a periappendiceal abscess
- ▶ Drain the abscess percutaneously or operatively
- ▶ If symptoms regress, manage conservatively
- ▶ If needed, perform an interval appendectomy at 5–6 weeks
- If appendicitis is not found:
  - ▶ Rule out adnexal disease (if tuboovarian abscess is found, incise and drain only, do not resect tube or ovary)
  - ▶ Examine the mesentery for adenopathy (mesenteric lymphadenitis)
  - ▶ Examine the small bowel for a distance of about 3 feet to rule out Crohn disease, ulcerative colitis, terminal ileitis, and Meckel's diverticulitis
- Complications (incidence approximately 5%)
  - ▶ Superficial wound or port-site infection. Treat by re-opening the skin and subcutaneous tissue, and begin saline wet-to-dry dressing (change tid); administer antibiotics
  - ▶ Pelvic abscess is the most common complication of a ruptured appendicitis; treat with antibiotics and drainage, depending on size and location
  - ▶ Ileus (place an NG tube and administer IV fluids)
  - ▶ Small bowel obstruction (place NG tube initially; may require lysis of adhesions)
  - ▶ Appendiceal stump blowout (drain or perform tube cecostomy)
  - ▶ Decreased fertility (due to scarring and fallopian tube obstruction)
- Prognosis
  - ▶ The principal factor determining mortality is rupture
    - ▷ Unruptured: 0.1% mortality
    - ▷ Ruptured: 3% mortality
  - ▶ Cause of death is usually uncontrolled gram-negative sepsis
- Appendiceal rupture
  - Deaths from appendicitis are almost always secondary to complications of perforation, resulting in gram-negative sepsis (*E coli* and *Bacteroides* are the most common organ-

isms); incidence of appendiceal rupture is significantly higher in pediatric and geriatric age groups (due to a delay in diagnosis)

- Rectal or pelvic examinations are essential for all age groups
- Young children may not be able to form a phlegmon to wall off a perforation or abscess because of a paucity of omentum
- Symptoms
  - ▶ Temporary pain relief after perforation is rare; localized pain progresses to encompass the entire right lower quadrant
  - ▶ A tender, boggy mass in the peritoneal area indicates the presence of a phlegmon or abscess
  - ▶ Symptoms usually last > 36 hours and include temperature elevation to 102°F–104°F (39°C–40°C), increased white blood cell count (20,000–35,000 with extreme left shift), and hemoconcentration
- Pylethrombophlebitis (septic thrombophlebitis of the portal vein) is a complication of gangrenous appendicitis heralded by chills, spiking fever, right lower quadrant pain, and jaundice; septic clots may embolize to the liver, producing multiple pyogenic abscesses
- The most common sites of seeding from an appendiceal perforation are the pouch of Douglas (pelvic cul-de-sac) and the right subhepatic space (via the right gutter)
- The accuracy of the preoperative diagnosis should be approximately 85%–90% and depends on 3 major factors:
  - ▶ Anatomical location of the inflamed appendix
  - ▶ Stage of the process (simple or ruptured)
  - ▶ Age and sex of patient (harder to diagnose in young females)
- The most common erroneous diagnosis is acute mesenteric lymphadenitis (especially in children), which is usually associated with an antecedent upper respiratory infection. In acute mesenteric lymphadenitis, pain is less severe and not as sharply localized, there may be generalized lymphadenopathy, and complete blood count may show lymphocytosis
- To treat, observe the patient for a short time, perform serial abdominal examinations, then explore if the diagnosis is still in doubt

- Acute gastroenteritis
  - The viral form is evident by profuse watery diarrhea, nausea, and vomiting. Hyperperistaltic abdominal cramps precede watery stools, and vomiting precedes the onset of abdominal pain. Complete blood count is usually normal or shows a right shift (increased lymphocytes)
  - Bacterial (caused by *Salmonella*) often results from the ingestion of contaminated food. Symptoms include bloody diarrhea, skin rash, bradycardia, leukopenia, chills, and fever
    - ▶ Typhoid fever is usually due to *Salmonella typhosa*, which is cultured from stool or blood. Symptoms include maculopapular rash, inappropriate bradycardia, leukopenia, and ileal perforation (in 1% of cases)
    - ▶ Treat with chloramphenicol
  - Bacterial (caused by *Yersinia enterocolitica*, *Campylobacter*) results from ingesting food contaminated by feces or urine
    - ▶ Symptoms include cervical and mesenteric lymphadenitis, ileus, and colitis
    - ▶ Treat with ampicillin or gentamicin
- Gynecological disorders are important in the differential diagnosis of appendicitis
  - Gynecological disorders compose the highest rate of missed diagnosis in young adult females
  - Differential diagnosis includes pelvic inflammatory disease (the most common erroneous diagnosis), ruptured Graafian follicle (mittelschmerz), and ruptured ectopic pregnancy
- Diseases in the male include torsion of the testis and acute epididymitis (see Chapter 21, Genitourinary Tract)
- Urinary tract infection
  - Urinalysis shows pyuria
  - Symptoms include urinary frequency and dysuria
  - Suprapubic or costovertebral angle tenderness is evident on physical examination
  - Patient's temperature will be  $> 101^{\circ}\text{F}$  (about  $38^{\circ}\text{C}$ )
- Ureteral stone
  - Pain is referred to pelvis
  - Laboratory findings show hematuria but neither fever nor increased white blood cell count

- Primary peritonitis
  - Diagnosis with peritoneal aspiration. If only a single species of cocci is seen, patient has primary peritonitis and needs medical treatment; if mixed flora are seen, patient has secondary peritonitis
- Henoch-Schönlein purpura
  - Usually occurs 2–3 weeks after a *Streptococcal* infection
  - Symptoms include joint pain, purpuric rash, and nephritis with albuminuria
  - Laboratory findings include increased platelets
- Hemolytic uremic syndrome
  - Main symptom is bloody diarrhea
  - Laboratory findings include anemia (due to hemolysis), increased white blood cells, decreased platelets, hematuria, and increased blood urea nitrogen
- Diverticulitis or perforating carcinoma of the cecum
- Constipation
  - Diagnose with abdominal radiograph
  - Treat with enemas
- Pneumonia
  - Physical findings show no point tenderness
  - Diagnose using chest radiograph
- Typhlitis (bacterial invasion of the intestinal wall)
  - Usually seen in oncology patients
  - Symptoms typically include right lower quadrant abdominal pain and neutropenia
- Hirschsprung disease (congenital megacolon)
  - Neural crest cells form neuroblasts, which migrate to the distal rectum in the twelfth week of gestation, becoming enteric ganglion cells (Auerbach myenteric plexus)
  - Pathology
    - ▶ Absence of ganglion cells in submucosa (Meissner plexus) and intramuscular plexus (Auerbach plexus)
    - ▶ Enlarged, hypertrophic, nonmyelinated nerve fibers in submucosa, muscularis mucosa, and Auerbach intramuscular plexus
    - ▶ Increased acetylcholinesterase in muscularis nerve fibers
  - Most common site of aganglionosis is the rectosigmoid due

- to arrested migration of neuroblasts (80%)
- Symptoms
  - ▶ Decreased stool frequency, failure to pass meconium within 48 hours after birth
  - ▶ GI obstruction (distention, bilious vomiting)
  - ▶ Enterocolitis (the major cause of morbidity and mortality)
  - ▶ Fecal soiling is rare
- Differential diagnosis
  - ▶ In the neonate: sepsis, necrotizing enterocolitis-associated stricture, meconium plug or ileus, or intestinal atresias
  - ▶ Habit constipation associated with a full rectal ampulla and fecal soiling (whereas in Hirschsprung disease, the rectal ampulla is usually empty)
  - ▶ Functional constipation (infrequent, large, firm stools accompanied by pain and bleeding from anal fissures), a history of normal stool frequency during infancy is associated with functional constipation, not with Hirschsprung disease
  - ▶ Hypothyroidism
- Enterocolitis
  - Symptoms include abdominal distention, diarrhea, vomiting, sepsis, and perforation
  - Usually occurs in children < 3 years old and is the most common cause of death in affected children
  - May occur before or after colostomy or pull-through
  - Laboratory test results show high levels of *C difficile* toxin
  - Diagnosis
    - ▶ Perform rectal biopsy (suction or full-thickness)
    - ▶ Perform a contrast enema
      - ▷ Conical transition zone from the distal, nondilated, aganglionic colon or rectum to the proximal (ganglionic) dilated colon as demonstrated on contrast enema
      - ▷ A transition zone may be absent in neonates
      - ▷ Excludes other causes of colonic obstruction, including small left colon syndrome, meconium plug syndrome, and atresia
  - Nonsurgical treatment

- ▶ Perform NG tube decompression in the upper GI tract
- ▶ Administer IV fluids
- ▶ Administer antibiotics (ampicillin, gentamicin, or metronidazole)
- ▶ Perform rectal irrigations with 20 cc/kg warm Ringer lactate tid using a soft, rubber catheter
- ▶ If the above measures are unhelpful and patient cannot be urgently transferred to the care of a pediatric surgeon, consider performing a colostomy
- Surgical treatment
  - ▶ Perform a leveling colostomy of normally innervated (biopsy-proven) bowel
  - ▶ If no clear-cut transition zone can be determined, the patient may have total colonic aganglionosis, and if a biopsy diagnosis is unobtainable, an ileostomy should be performed
  - ▶ Refer patient to a facility with pediatric surgery capability, if possible

### **Rectum and Anus**

- Rectal prolapse (proctidentia)
  - True prolapse: circular folds of full-thickness bowel
  - False prolapse: radial folds of mucosa only
  - Treatment
    - ▶ Rule out cystic fibrosis and parasites
    - ▶ Discourage patient from prolonged sitting or straining during a bowel movement
    - ▶ Prescribe stool softeners
    - ▶ Administer submucosal injection of 5% phenol in almond oil, hypertonic glucose (50%), or hypertonic saline (20%)
      - ▷ Administer 5 cc per treatment
      - ▷ Use a #18 spinal needle for four-quadrant sclerosis; inject 2 cc posteriorly and 1 cc in the other three quadrants (almost all cases will resolve spontaneously without an operative procedure)
- Anal fissure
  - Most common cause of rectal bleeding in the newborn
  - Symptoms include blood streaks on outside of stool and constipation

- Physical examination will show a superficial tear of the anal mucosa, usually in the posterior midline. A sentinel skin tag indicates a chronic fissure. Fifty percent of anal fissures are associated with fistula-in-ano (see below)
- Caused by stretching and tearing during evacuation of large, hard stools
- Treatment consists of administering stool softeners, dilatation (may require manual stretch under sedation), and sitz baths
- Perianal and perirectal abscess
  - In infants, these can develop from an infected diaper rash. In children, rule out Crohn disease, leukemia, and immunodeficiency syndromes
  - Symptoms include fever and pain
  - Treat with incision, drainage, and sitz baths
- Fistula-in-ano
  - Results from a perianal abscess extending from a crypt to the perianal skin (suspect when an abscess recurs)
  - Tract is intersphincteric and lateral to the anus
  - Treat with fistulotomy (over a probe), tract curettage, or, if fistula is complex or high, place a seton tie
- Imperforate anus
  - Evaluate for associated VACTERRL defects
  - Initial approach consists of descending colostomy and mucus fistula
- Preoperative mechanical bowel preparation
  - Give polyethylene glycol electrolyte solution (25–35 cc/kg/h for 4–6 h via an NG tube; safe in infants and children)
  - Add oral erythromycin base and neomycin