Pediatric Abdominal Disorders

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Wilms tumor arises from the renal parenchyma and in most cases is asymptomatic. Neuroblastoma develops from the adrenal gland or along the sympathetic chain and may thus be manifested as either a midline or flank mass. It is an aggressive malignancy and has often metastasized at diagnosis. Ultrasonography and computed tomography (CT) are useful for investigation of abdominal lesions. A pediatric surgeon and oncologist should be consulted early for management of abdominal tumors.

GASTROINTESTINAL BLEEDING

As a general rule, gastrointestinal (GI) bleeding is not usually as severe in children as in adults. In particular, the vast majority of cases of neonatal GI bleeding are benign. The first step in evaluation is to confirm that the suspicious material found in the stool or diaper is actually blood by performing a guaiac filter test for occult blood. Children routinely consume both foods (watermelon) and liquids (antibiotics such as cefdinir, fruit punch, and other juices) that turn the stool red, which may falsely lead both parents and health care workers to believe that GI blood loss is occurring. The benign pink or orange urate crystals in the urine of some neonates and young children are sometimes mistaken for blood when seen in diapers. Urinary tract infections and urethral prolapse may also result in deposition of blood in the diaper that could be confused with GI blood.

Upper GI bleeding produces dark brown, black, or simply heme-positive stool. However, because of the fast transit time in neonates, some upper GI bleeding may be bright red. The most common cause is swallowed maternal blood, acquired either during delivery or as a result of breastfeeding from irritated or cracked nipples.

Esophageal varices are rare in children. In contrast to adults, in whom primary hepatic disease is the leading cause, the most likely cause of varices in children is splanchnic and portal vein obstruction. Varices may develop secondary to umbilical vein catheterization, dehydration, sepsis, or omphalitis. Less common causes are hepatic parenchymal conditions such as biliary cirrhosis secondary to biliary atresia, cystic fibrosis, α1-antitrypsin deficiency, and hepatitis. Early onset of inflammatory bowel disease occurs in the teenage years and is uncommon in younger children.

Perirectal skin breakdown and external rectal fissures both produce blood-streaked stool and are easily identifiable on

KEY POINTS

- The most common cause of intestinal obstruction in patients younger than 6 years is intussusception.
- Henoch-Schönlein purpura is a vasculitis that causes abdominal pain, purpura, and arthritis. Renal involvement is present in up to 50% of cases and is manifested as microscopic hematuria and proteinuria.
- Bilious emesis in infants is suggestive of highly morbid conditions such as malrotation with volvulus, necrotizing enterocolitis, sepsis, or small bowel obstruction.
- In up to 90% of children younger than 2 years with appendicitis, the appendix has perforated by the time of diagnosis. These patients will be found to have generalized peritonitis and shock more often than older children or adults with appendicitis.

EPIDEMIOLOGY

Abdominal pain is a common complaint in children. Up to 25% of children will experience discomfort severe enough to interfere with activity, and annually, one in every seven children in the United States will visit a physician because of abdominal complaints, yet most will have no organic cause identified. Between 2% and 4% of all pediatric outpatient visits are related to abdominal complaints. Discerning the presence of serious underlying disease can be challenging. This chapter describes several of the most significant pathologic abdominal conditions in pediatric patients.

ABDOMINAL MASSES

Obstructive uropathy and renal cysts are the most likely causes of abdominal distention in infancy. Abdominal tumors are slow growing and usually recognized incidentally (e.g., when a parent or physician feels an abdominal mass). The two most common malignancies are Wilms tumor and neuroblastoma. The majority of affected children are younger than 5 years.
physical examination. Sitz baths and stool softeners are useful in treating rectal fissures.

Emergency department (ED) management of GI bleeding is directed at fluid and blood resuscitation.

**MECKEL DIVERTICULUM**

A Meckel diverticulum is the most common omphalomesenteric remnant. The most frequently observed finding is painless rectal bleeding, which occurs as a result of ulceration of the diverticulum or neighboring mucosa by the ectopic tissue. The ectopic tissue is gastric in origin in more than 80% of cases, but it may be pancreatic as well. Symptoms usually occur within the first 2 years of life, and in the majority of affected individuals it is diagnosed by 20 years of age (Box 22.1). A Meckel diverticulum can act as a lead point in intussusception. The diagnostic study of choice is a radiolabeled bleeding study called a Meckel scan. Definitive therapy is surgical excision.

**INTUSSUSCEPTION**

Intussusception occurs when one loop of intestine invaginates into another. Intussusception of the mesentery can also occur and result in edema and vascular congestion. It is the most common cause of intestinal obstruction in children younger than 6 years. The ileocolic region is most often involved. Intussusception is usually manifested in children 6 to 18 months old, with a peak occurrence at 10 to 12 months. The vast majority of cases in children younger than 3 years are idiopathic. One etiologic theory is that inflammation of Peyer patches within the intestine acts as a lead point for the intussusception. In children older than 5 years, a true lead point is found more than 75% of the time. Lead points include polyps, lymphoma, Meckel diverticulum, surgical adhesions, and mucosal inflammation secondary to vasculitis.

Intussusception is often preceded by a viral illness, and the patient may have a low-grade fever at the time of evaluation. Symptoms consist of vomiting and episodic, crampy abdominal pain. Initially, children return to baseline between episodes, but as the condition persists, they may become lethargic. Screaming episodes lasting up to 10 to 15 minutes with hip and knee flexion are routine. The episodes increase in frequency and duration over time, with subsequent shortening of asymptomatic intervals.

The classic triad of symptoms—vomiting, abdominal pain, and “currant jelly” stools—is seen in less than one third of patients. However, more than 75% have two of these findings. Early on, stools test guaiac negative. If bowel ischemia ensues, frank blood mixed with mucus gives the stool a currant jelly appearance.

Some children have only lethargy, which can delay the diagnosis. The most commonly confused entity is constipation because of the similar pattern of colicky abdominal pain. These two conditions can easily be differentiated with a plain abdominal radiograph.

The history is the best guide to the diagnosis of intussusception. On abdominal palpation the right lower quadrant may be empty because the cecum has rotated out of its standard position. The actual intussusception may be palpated as a sausage-shaped mass in the right upper quadrant. Normal physical findings should not dissuade the examiner from proceeding with investigation because most children appear normal between episodes. No laboratory studies are available to confirm the diagnosis, and guaiac test–positive stool is a late finding. Abdominal radiographic findings are most often normal, but a mass may be seen in the right upper quadrant.

Intussusception can be reliably diagnosed with ultrasound. Enemas are both diagnostic and therapeutic. Intussusceptions that cannot be reduced by enema must be reduced surgically. Up to 10% of cases recur, most often within 24 hours. After reduction, the child must be admitted to the hospital for a 24-hour observation period.

Children with a history and physical findings suspicious for intussusception must be evaluated quickly because the passage of time increases both the edema and the difficulty of achieving reduction. A pediatric surgeon should be contacted before the child undergoes attempted enema reduction in case of failure or perforation. Ileoileal intussusceptions may be difficult to visualize and reduce via enema unless there is significant reflux of contrast material. Such intussusceptions are associated with Henoch-Schönlein purpura (HSP), in which the vasculitis acts as a lead point.

**HENOCH-SCHÖNLEIN PURPURA**

HSP is a vasculitis that predominantly affects the capillaries and small vessels. It occurs most commonly in school-age children, with the classic triad consisting of abdominal pain, purpura, and arthritis. Elevations of immunoglobulin A (IgA) are detectable in the blood, along with immune complex deposition in the skin and glomeruli. There is no known etiology, but HSP has often been found to occur after upper respiratory infections. Recurrences are seen in up to 50% of cases.

Abdominal pain occurs in half the patients and is most frequently colicky in nature. Intussusception should be strongly considered in children with guaiac test–positive or frankly bloody stools accompanied by severe pain. The subtype of intussusception associated with HSP is often ileoileal, which is difficult to visualize and reduce with a contrast enema. CT is the imaging modality of choice for ileoileal intussusception associated with HSP, and reduction must often be achieved surgically.

The rash associated with HSP is petechial, purpuric, and usually located on the buttocks and lower extremities. It is seen in dependent areas, so the scrotum and hands may also be affected. In younger children, the rash is displayed on the
dorsal surfaces of the extremities, trunk, and head. The lesions are palpable and appear and progress like common bruises.

Arthritis occurs in two thirds of patients and most often involves the large joints of the lower extremities. The knees and ankles may be edematous and tender to palpation or with movement. Joint pain may be so severe that it interferes with ambulation.

Renal involvement occurs in 25% to 50% of patients and is usually manifested as microscopic hematuria and proteinuria.

The diagnosis of HSP is based on the characteristic rash and accompanying symptoms. The results of a complete blood count and clotting studies are normal.

Nonsteroidal antiinflammatory drugs are the traditional therapy for the arthralgias observed with HSP. Use of corticosteroids for the management of abdominal pain and renal involvement is controversial. Outpatient management is usually sufficient, although substantial joint or abdominal discomfort may require inpatient admission. Severe abdominal pain, especially in conjunction with guaiac test–positive or grossly bloody stools, merits an evaluation for intussusception.

**GASTROESOPHAGEAL REFLUX**

Gastroesophageal reflux (GER) is caused by a loose esophageal sphincter with retrograde passage of food into the esophagus. It is usually manifested in the first few weeks after birth as emesis during or soon after the cessation of feeding. The emesis may be blood streaked but should never be bilious. Some children in whom GER is eventually diagnosed may have previously been incorrectly labeled as having colic, feeding difficulties, or formula intolerance.

Symptoms of GER range from small “wet burps” to discomfort during feedings with arching of the back. One particularly severe form of reflux is the Sandifer syndrome, in which the child has opisthotonic movements and unusual head and neck positioning. The head movements may be an attempt to reduce pain by elongating the esophagus and protecting the airway from aspiration. Complications of GER include failure to thrive, apnea, laryngospasm, and aspiration pneumonia. Reflux esophagitis may be the culprit in children with guaiac test–positive stools or iron deficiency anemia. In most cases, GER resolves spontaneously by 1 year. The diagnosis is most often made from a careful history. Esophageal pH probe, nuclear milk scan, barium swallow study, and direct feeding under fluoroscopic observation are all diagnostic options.

Nonpharmacologic interventions are frequently sufficient to relieve the majority of cases of GER. Smaller-volume feedings with breaks for burping are often helpful. Caregivers should be instructed to keep children semiupright for 30 to 45 minutes after feedings. Thickening of feedings with cereal reduces crying and improves symptoms.

Ranitidine, a histamine blocker, reduces gastric acid and aids mucosal healing. Metoclopramide stimulates esophageal and gastric motility, thereby decreasing the volume of gastric contents that may reflux. GER that is severe and resistant to medical therapy may require surgical intervention with a Nissen fundoplication. This procedure involves wrapping and surgically fixing a portion of the stomach around the esophagus.

**PYLORIC STENOSIS**

Pyloric stenosis results from idiopathic hypertrophy of the antrum of the stomach. The male-to-female ratio is 5:1, and familial occurrence is present in up to 50% of patients. It is usually diagnosed in children at 2 to 5 weeks of age and rarely after 3 months. They feed normally at first but later experience vomiting in the midst of or soon after feeding. The emesis begins as mild and small and then progresses to voluminous and projectile. It is never bilious, although it may be blood streaked. The patient appears to vomit the entire bottle and then refeeds ravenously. After eating, peristaltic waves may be visible across the abdomen. If the condition continues, the child loses weight and becomes dehydrated, with sunken eyes, loose skin, and lethargy. Electrolyte analysis shows a hypochloremic, hypokalemic metabolic alkalosis. As the child continues to vomit, hydrogen and chloride are expelled. The kidneys attempt to maintain normal pH by eliminating potassium and hydrogen ions.

The differential diagnosis includes GER, overfeeding, and gastritis. Careful examination may detect the pylorus or “olive,” especially in a thin, dehydrated child with prolonged illness. Pyloric stenosis is identified with either ultrasonography or an upper GI series. Ultrasonography is the preferred modality because in addition to its high rate of accuracy, no radiation exposure is involved. Ultrasonographic diagnosis is made by visualization of a thickened pylorus. An upper GI radiographic series will demonstrate a “string sign” as the contrast agent squeezes through a narrowed pylorus.

Surgery is the treatment of choice for pyloric stenosis. However, diagnosis of this disorder does not represent a surgical emergency. ED management consists of rehydration and correction of electrolyte status before surgical intervention.

**MALROTATION WITH VOLVULUS**

Early in normal embryonic development, the intestine rotates around the superior mesenteric artery. The duodenum and cecum become widely displaced and fixed into position by the mesentery does not fan outward. This narrow stretch of mesentery, which contains the superior mesenteric artery, crosses over the duodenum. It can easily twist on itself and cause duodenal obstruction and arterial compression. The result is ischemia with the potential for intestinal necrosis within 1 to 2 hours.

The common manifestation is an acute onset of abdominal pain with bilious emesis. Abdominal distention may or may not be present, depending on the anatomic level of the obstruction. Affected children are usually seen in the first year of life, with the majority of cases occurring within the first week to month. Older children may have a history of intermittent episodes of vomiting and abdominal pain that suddenly become more severe. Bloody stools should raise the level of concern for bowel ischemia and impending gangrene. Patients are often quite ill and may be in shock at initial evaluation.

The differential diagnosis of bilious emesis in infants includes a short list of highly morbid conditions (Box 22.2).
In addition to malrotation, sepsis, small bowel obstruction, and necrotizing enterocolitis (NEC) must be considered.

A loop of small bowel overlying the liver may be visible on plain abdominal radiographs. Distal bowel gas is limited or absent. The “double bubble” sign can be visualized on an upright film; it is produced by a dilated stomach and duodenum. An upper GI radiographic series, the diagnostic study of choice, demonstrates abnormal anatomy with a coiled spring appearance of the jejunum in the right upper quadrant.

All children with bilious emesis need immediate evaluation by a surgeon. Malrotation with midgut volvulus is a surgical emergency because bowel infarction can occur rapidly.

**NECROTIZING ENTEROCOLITIS**

NEC is generally a disease of premature infants and is usually diagnosed in the intensive care unit. However, up to 10% of infants with NEC are born at full term, so initial evaluation in the ED is possible. Signs and symptoms range from feeding intolerance and vomiting to pneumatosis intestinalis (air within the intestinal wall), perforation, shock, and disseminated intravascular coagulation. Most affected newborns experience vomiting, which may or may not be bilious. The clinical findings may be limited to guaiac-test–positive stools and feeding intolerance, but in severe cases, infants have massively distended, rigid abdomens in the setting of shock. Gas within the biliary tract is present in 10% to 30% of cases. With improved survival of premature infants, children may seen in the ED with sequelae of NEC, such as strictures, obstruction, fistulas, and short gut syndrome.

**APPENDICITIS**

Appendicitis is typically accompanied by diffuse or periumbilical abdominal pain. Within 8 to 24 hours, vomiting begins, and the pain migrates to the right lower quadrant. Abdominal pain, vomiting, and fever are the classic symptom triad for the disease. Although the pain precedes other symptoms in adult studies, such may not be the case in many children with appendicitis. Up to one third of pediatric cases do not follow this order of symptoms, and vomiting is often reported as the first sign. Fever is routinely low grade; a temperature higher than 39° C reduces the likelihood of appendicitis, except in cases of perforation.

The position of the appendix dramatically affects the location of the pain and symptoms. A normally placed appendix produces discomfort at the McBurney point. A low-lying pelvic appendix may irritate the sigmoid colon and mimic enteritis with diarrhea. A retrocecal appendix may produce flank or posterior pain and may be confused with pyelonephritis or septic arthritis of the hip.

Movement increases the discomfort (e.g., bumps in the car ride to the hospital, walking, jumping) such that patients may walk hunched over, limp, have a shuffling gait, or put weight preferentially on the left leg. Although anorexia is a classic finding, children can be enticed by their favorite foods and may even want to eat.

Examinations should be gentle, and the right lower quadrant should be palpated last to allow evaluation of reportedly nontender regions and to gain the patient’s confidence. Bowel sounds are usually normal or hypoaecitive. An external genital examination must be performed to exclude testicular disorders and incarcerated hernia.

Appendicitis is particularly difficult to identify early in the course of the illness and in the very young (Box 22.3). Approximately 90% of patients younger than 2 years with appendicitis have a perforated appendix at the time of diagnosis. Children have a thinner appendiceal wall and a less well-developed omentum. Therefore, rupture occurs more readily and results in more diffuse bacterial dissemination. Pediatric cases of appendiceal perforation have more severe and diffuse peritonitis than do adult cases. Although the mortality from appendicitis has improved dramatically, the rate of perforation has not changed significantly in the past few decades.

A complete blood count showing leukocytosis and a left shift is supportive of the diagnosis, but many children have normal white blood cell (WBC) counts. Most patients with appendicitis have an elevated WBC count in the range of 11,000 to 15,000 cells/mm³. An appendix in close proximity to the ureter can produce sterile pyuria and mild hematuria. A positive urine Gram stain response and the presence of leukocyte esterase and nitrites can help differentiate a true urinary tract infection from inflammatory hematuria secondary to appendicitis.

In the case of perforation, the pain initially resolves but then becomes more generalized with peritoneal symptoms. It may be most severe in both lower quadrants as the purulent material settles. Young children may simply have nonspecific
symptoms such as fussiness, inconstable crying, irritability, and grunting respirations. Once perforation occurs, the child may have poor perfusion, tachycardia, high fever (>39°C), and even septic shock. Bowel sounds are then absent; the abdomen is rigid with rebound tenderness and involuntary guarding. The WBC count is dramatically elevated with a significant left shift.¹⁰

A fecalith is visible on plain abdominal radiographs in 8% to 10% of patients with appendicitis. A number of other findings can be seen on plain films, including scoliosis concave toward the right side, a sentinel loop overlying the appendix indicating an area of inflammation, air within the appendix, a mass in the right lower quadrant (more often in cases of abscesses), and loss of the psoas shadow on the right.

Ultrasoundography is a useful tool for evaluating children with concerns for appendicitis. The classic finding known as the “target sign” is a fecalith inside a large, inflamed appendix. In obese children, visualization and diagnosis become more challenging. The sensitivity and specificity of ultrasonography exceed 90%, but ruptured appendices are notoriously difficult to identify. No diagnosis can be made if the appendix is not visualized. Ultrasonography is helpful in differentiating appendicitis from other causes of abdominal pain, such as ovarian cysts, but cannot exclude conditions such as mesenteric adenitis. CT has a sensitivity and specificity higher than 95% and may be used in cases with a broad differential diagnosis or when the findings on ultrasound are equivocal or it is unavailable.¹¹ Abdominal CT is helpful in diagnosing inflammatory bowel disease and mesenteric adenitis. Admission for serial abdominal examinations is warranted for a child with a compelling history and physical examination findings but equivocal laboratory findings.

A surgeon should be involved early in the ED evaluation of a child with suspected appendicitis because imaging may be avoided in classic cases. Pain must be addressed, and numerous studies have shown that narcotics do not affect the diagnostic accuracy of the examination. Surgeons may choose to evaluate the patient before analgesia is given, but pain medication should not be withheld indefinitely pending surgical assessment. Signs of shock should be addressed aggressively, and children with suspected rupture should receive broad-spectrum antibiotic coverage.

**HIRSCHSPRUNG DISEASE**

Hirschsprung disease is caused by the absence of ganglion cells in the myenteric plexus of the colon. Without ganglion cells, that segment is under constant contraction. The proximal region dilates to compensate, with resultant constipation. Patients have complaints of chronic constipation, abdominal distention, and vomiting. Physical examination demonstrates palpable stool in the abdomen with a tight anal sphincter, but absence of stool in the rectal vault. Hirschsprung disease has a 4:1 male preponderance. In 80% of affected patients it is diagnosed within the first year of life. The most serious complications are toxic megacolon, perforation, and enterocolitis. A dilated colon, fecal impaction, and air-fluid levels are visible on abdominal radiographs. The diagnosis is made from the finding of aganglionosis on biopsy or anal manometry. A barium enema without bowel preparation shows a narrowed colonic segment and a dramatically dilated proximal segment. In uncomplicated cases, outpatient surgical evaluation is indicated. Resection of the aganglionic segment is curative.

The vast majority of cases of constipation are functional and tied to behavioral and psychologic causes. Pathologic causes are rare and, in addition to Hirschsprung disease, include cystic fibrosis and hypothyroidism.

**BILIARY TRACT DISEASE**

Gallstones are an unusual condition in healthy children. The most common type, pigment stones, usually develop in children with underlying hemolytic conditions such as sickle cell disease and hereditary spherocytosis. Cholesterol stones are more prevalent in adolescent girls, obese children, patients with cystic fibrosis, and those who depend on total parenteral nutrition. Because of the large percentage of pigment stones, up to 50% of cases of cholelithiasis in children are visible on abdominal radiographs, in contrast to 10% to 15% of cases in adults.

Acute cholecystitis (inflammation of the gallbladder in the absence of gallstones) is more common in children than cholelithiasis is. Causes include Kawasaki disease; bacterial infections such as typhoid, shigellosis, and scarlet fever; and viral infections such as hepatitis.

Acute cholangitis usually occurs in children with a history of biliary surgery, as used for the management of choledochal cysts and biliary atresia. The clinical picture is similar to that in adults and consists of right upper quadrant pain, fever, and vomiting. Treatment is inpatient admission for therapy with broad-spectrum antibiotics such as ampicillin, gentamicin, and metronidazole.

**MILK PROTEIN ALLERGY**

Milk protein allergy is manifested as blood-streaked, mucous stools in young infants exposed to cow’s milk–based formulas. Significant flatus and mild discomfort with feeding may be noted. However, most children appear nontoxic and otherwise well. Edema, inflammation, and discrete ulcerations are present in the intestinal mucosa. This condition is best described to occur with consumption of milk protein but may develop with any formula, including soy. Treatment is to change to a formula with a different protein source. The symptoms should resolve within 1 week of complete withdrawal of the offending agent.¹²

**REFERENCES**

References can be found on Expert Consult @ www.expertconsult.com.
REFERENCES