Beyond appendicitis: evaluation and surgical treatment of pediatric acute abdominal pain

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Purpose of review
Evaluation of the child with acute abdominal pain is challenging because of the wide range of potential diagnoses. Presenting symptoms, clinical examination, and laboratory findings can guide selection of diagnostic imaging.

Recent findings
Intussusception and intestinal malrotation are potentially serious causes of intestinal obstruction, which are best evaluated by ultrasound and upper gastrointestinal series, respectively. Ultrasound has diagnostic importance in the evaluation of multiple diseases, including appendicitis, by potentially decreasing the need for inpatient observation, cholecystitis and complications of gall stones such as pancreatitis, and ovarian diseases. Pelvic inflammatory disease should be considered in evaluation of a teenage girl with lower abdominal pain. Less common causes of acute abdominal pain include ingested foreign bodies, infected congenital anomalies, and perforated peptic ulcer disease.

Summary
Presenting symptoms and physical examination findings can narrow the number of potential diagnoses in pediatric acute abdominal pain and thereby guide diagnostic imaging selection. Abdominal/pelvic ultrasound, rather than computed tomography scan, is the preferred modality for initial evaluation of many potential causes of pediatric abdominal pain.

Keywords
abdominal pain, diagnostic imaging, pediatric

INTRODUCTION

Due to the variety of underlying causes, acute abdominal pain in children poses a diagnostic challenge. Conditions manifested by acute abdominal pain vary in incidence with age and sex. Accompanying symptoms and signs are inconsistently present, especially in young children; however, these can guide selection of appropriate diagnostic tests, imaging, and definitive treatment. More recently, avoidance of unnecessary radiation exposure from diagnostic imaging, especially abdominal–pelvic computed tomography (CT) scan [1▪], has gained attention due to concern regarding long-term cancer risk [2]. Nevertheless, accurate and timely diagnosis is sought to avoid both inappropriate operations, as many causes of acute abdominal pain are nonsurgical [3–5], and diagnostic delays that increase morbidity. Here, recent developments in the evaluation and surgical treatment of specific causes of pediatric acute abdominal pain are summarized (Table 1).

ABDOMINAL PAIN WITH SYMPTOMS OF INTESTINAL OBSTRUCTION

In a child with acute abdominal pain, the pain pattern and accompanying symptoms can point to intestinal obstruction as the cause. Abdominal pain is often intermittent or 'colicky'; bilious emesis, in particular, implies the presence of an intestinal obstruction until proven otherwise. In cases of prolonged or high-grade obstruction, intestinal ischemia, infarction, or perforation leads to

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KEY POINTS

- The cause of pediatric acute abdominal pain varies with age and sex.
- Overlap of symptoms and examination findings necessitates use of diagnostic laboratory tests and imaging.
- Use of presenting symptoms and signs of intestinal obstruction and inflammation to narrow the diagnoses under consideration can optimize selection of diagnostic imaging.

inflammatory symptoms such as fever and signs of peritonitis. Intestinal obstruction can result from congenital anomalies or acquired lesions.

Table 1. Surgical causes of pediatric acute abdominal pain

Intestinal obstruction	Intestinal malrotation/volvulus
	Intussusception
	Congenital anomalies with internal hernia/volvulus
	Omphalomesenteric duct remnants
	Intestinal duplication cyst
	Mesenteric cyst
	Incarcerated inguinal hernia
	Adhesive bowel obstruction
	Congenital adhesions
	Postoperative
	Intraluminal obstruction
	Foreign bodies
	Distal intestinal obstruction syndrome
Inflammation	Appendicitis
	Congenital anomalies – infected
	Meckel diverticulitis
	Urachal remnant
	Mesenteric/intestinal duplication cyst
	Cholecystitis/Pancreatitis
	Pelvic inflammatory disease
	Other
	Complicated Crohn's disease
	Perforated viscus
Other	Ovarian
	Ruptured cyst
	Torsion
	Omental/epiploic fat infarction

Intestinal malrotation with volvulus

Malrotation occurs when the intestine is abnormally oriented within the peritoneal cavity during embryonic development. The ascending colon is not fixed to the right side of the abdomen; abnormal 'Ladd's' bands kink the duodenum and narrow the base of the intestinal mesentery, which predisposes the bowel to twisting [6]. Intestinal malrotation with or without volvulus can present at any age but is often diagnosed during infancy [7,8]. Presenting symptoms include abdominal pain, especially in children older than 2 years, and nonbilious or bilious emesis [7–9]. Elevated white blood count, glucose, and C-reactive protein (CRP) are associated with bowel infarction [8]. Abdominal radiographs have limited utility in diagnosing malrotation with volvulus, as the bowel gas pattern is often nonspecific [6]; rather, alternate diagnoses may be suggested by findings of distal intestinal obstruction. Upper gastrointestinal (UGI) series is the primary imaging modality to assess intestinal rotation, specifically, the position of the duodenal–jejunal junction. False-positive UGI studies occur when the duodenal–jejunal junction is displaced by hepatomegaly, splenomegaly, gastric overdistension, or scoliosis. Contrast enema demonstrates the position of the cecum, which is nonfixed with malrotation but can be mobile even with normal rotation [6]. Abnormal relationship between the superior mesenteric artery and vein in malrotation can be detected by ultrasound, which is less specific and sensitive than UGI studies. On CT scan, volvulus leads to the 'whirlpool' sign, created by the twisted bowel mesentery; overlying bowel gas can obscure this finding on ultrasound.

Because of the risk of massive bowel infarction, intestinal malrotation with volvulus requires emergent surgical treatment. In a 10-year review of intestinal malrotation operations in Ireland, 76.4% were performed emergently, and volvulus was identified in 31% [7]. The Ladd procedure consists of reduction of the volvulus, division of Ladd bands, placement of the small bowel on the right and colon on the left side of the abdomen, and incidental appendectomy. The procedure can be performed laparoscopically even in the presence of volvulus [9,10]. Postoperative complications occur with variable frequency depending on the approach, and include adhesive bowel obstruction (up to 13.3% with open approach) and postoperative volvulus (up to 5.6% with laparoscopic approach) [9].

Intussusception

A common cause of intestinal obstruction in young children, intussusception may be idiopathic

or associated with an anatomic lesion, such as Meckel diverticulum, benign tumor (hamartoma), lymphoma, or hematoma (Henoch-Schonlein purpura). In recent studies from Germany [11▪] and Taiwan [12], intussusception was most common in children under 3 years of age with a male preponderance. Viral infection may have a pathogenic role. Increased intussusception following older generation rotavirus vaccine (RotaShield, RRV-TV, Wyeth Laboratories, Inc., Marietta, PA) administration has not been observed with current vaccines (RotaTeq, RV5, Merck & Co., Whitehouse Station, NJ; Rotarix, RV1, GlaxoSmithKline, Research Triangle Park, NC) [11▪,13]. Interestingly, adenovirus serotypes that typically cause respiratory symptoms have been identified within stool specimens from children with intussusception [14▪]. Pathologic lead points are more frequently found in older children [15,16].

Presenting symptoms in children with intussusception vary with age. Abdominal pain (especially intermittent or cramping) and emesis were more common in older children in studies from Japan [14▪] and the United States [16]. Children less than 2 years of age with intussusception often present with bloody stools and lethargy, although this is less consistently seen in older children [14▪]. Only a minority of intussusception cases from Germany had the classic triad of intermittent abdominal pain, abdominal mass, and bloody stool [11▪].

The optimal diagnostic imaging for pediatric intussusception has been explored in several recent studies. Abdominal radiographs have utility under select circumstances. Weihmiller *et al.* [17▪] found a low likelihood of intussusception among evaluated children who had normal abdominal radiographs in combination with other clinical features ($\leq$5 months old, or $>$5 months old without bilious vomiting and with diarrhea). Intraluminal mass, nonvisualization of the cecum, and obscured liver margin/right upper quadrant mass on abdominal radiographs interpreted by pediatric radiologists are highly specific but not sensitive for intussusception [18]; in contrast, both sensitivity and specificity are low with abdominal radiographs interpreted by pediatric emergency medicine physicians [19]. When combined with symptoms such as lethargy, abdominal pain, and vomiting, radiographic findings of soft tissue mass, bowel obstruction, visible intussusception, or paucity of colonic gas have high specificity for intussusception, similar to ultrasound [20]. Nonetheless, ultrasound is the preferred modality for diagnosis of intussusception due to high sensitivity and specificity [21] and avoidance of potentially unnecessary radiation exposure from diagnostic contrast enema [22].

Treatment of ileocolic intussusception is often achieved by contrast enema reduction [11▪,12,23]. Predictors of successful contrast enema reduction include performance in hospitals that treat a high volume of intussusception ($\geq$7 cases per year) [24], and location of the intussusception proximal to the splenic flexure of the colon [25]. Operative reduction is indicated in children with signs of intestinal compromise at presentation or who fail enema reduction. Older children are more frequently treated with operative reduction, likely due to concern for a pathologic lead point [15,16]. Following successful contrast enema reduction, children are typically observed for signs of recurrent intussusception or bowel compromise; recently, the necessity of inpatient observation [26] and fasting [27] has been questioned. Recurrent intussusception occurs in 8–12% of children [12,26,27] with less than half occurring within 24 h, and is associated with treatment by enema compared with operation [12,23].

Congenital anomalies with internal hernia or volvulus

Abdominal congenital anomalies, such as omphalomesenteric duct remnants, intestinal duplication cysts, and mesenteric cysts, can cause intestinal obstruction by internal herniation or volvulus. Omphalomesenteric duct remnants, including Meckel diverticulum, may have peritoneal bands tethered to the umbilicus or mesentery. In a review of children treated with Meckel diverticulectomy, half were less than 4 years old and 30% presented with bowel obstruction [28]; 25% of children with symptomatic omphalomesenteric duct remnants in Turkey had bowel obstruction due to bands, volvulus, or internal hernia at a median age of 42 months [29]. Abdominal radiographs and CT scan generally reveal small bowel dilation with paucity of bowel gas in the distal ileum and colon without intraabdominal inflammation. Ultrasound, CT scan, and MRI are diagnostic for both intestinal duplication and mesenteric cysts [30]. On contrast studies, intestinal duplication cysts may fill if communicating with the intestinal lumen or exert mass effect; ultrasound can demonstrate the muscular wall [30]. Multiple, thin-walled loculations typical of mesenteric cysts are well demonstrated on ultrasound and MRI.

Incarcerated inguinal hernia

When intestine is incarcerated within inguinal hernias, emergent reduction is necessary to avoid ischemic damage to the intestine, and, in boys, the

testis. Prompt repair of inguinal hernia infants has been advocated with the intent of avoiding incarceration; however, the actual risk of incarceration may be lower than previously thought. In a study of premature infants with inguinal hernia, 81% were boys and 4.6% had incarceration [31]; another study identified incarceration in 8% of children under 2 years of age who underwent inguinal hernia repair [32]. Potentially unavoidable, the incarcerations occurred at initial presentation with an inguinal hernia or in premature infants who were still hospitalized postnatally [31,32].

Adhesive bowel obstruction

Both congenital and acquired intraabdominal adhesions can cause small bowel obstruction. Internal herniation within congenital mesenteric defects is suggested by whirlpooling of mesenteric vessels on CT scan or ultrasound, combined with findings of intestinal obstruction by CT scan or contrast studies [33]. In a review of postoperative course following appendectomy, 2.2% of children developed bowel obstruction at a median interval of 6.1 months; appendiceal perforation and open approach were associated with higher risk of bowel obstruction [34]. Operative treatment was used in 81% of postappendectomy bowel obstructions, but was not associated with initial operative approach or perforation status [34]. Water-soluble antegrade contrast study may expedite care of partial adhesive bowel obstruction by demonstrating resolution; Bonnard *et al.* [35] found a shorter length of hospital stay and time to first feeding in children who underwent contrast study compared with historical controls, although these results were potentially biased by a lower rate of operative intervention in the contrast study group.

Intraluminal bowel obstruction

Ingested foreign bodies, including bezoars and magnets, may cause intestinal obstruction. Undigestible materials, such as hair (trichobezoar) or vegetable/fruit (phytobezoar), may form a mass within the stomach that extends or moves distally [36]. Symptoms include abdominal pain, nausea/emesis, early satiety, decreased appetite, and weight loss [37]. Endoscopic, laparoscopic, and open approaches are used for bezoar removal [37–39]. Ingestion of multiple, strong, rare-earth magnets can cause intestinal volvulus and fistula [40]; as a consequence, recommended treatment includes monitoring progression of single magnets through the intestinal tract, and endoscopic or operative removal of multiple magnets [41].

Distal intestinal obstruction syndrome (DIOS) results from impaction of viscous stool within the distal small bowel and colon of individuals with cystic fibrosis. DIOS diagnostic criteria include evidence of intestinal obstruction (bilious emesis and/or air–fluid levels on abdominal radiographs), fecal material in ileum/cecum, and abdominal pain and/or abdominal distension [42]. In a multicenter European study, DIOS occurred infrequently; however, the incidence increased with age, and 20% of patients had recurrent DIOS [42]. Treatment includes oral laxatives, enema with polyethylene glycol lavage, and retrograde water-soluble contrast enema; surgical intervention is rarely needed (4%) [42].

ABDOMINAL PAIN WITH INFLAMMATORY SYMPTOMS

An intraabdominal inflammatory process is suggested when a child has fever and abdominal tenderness in conjunction with abdominal pain. Difficult to distinguish from rarer conditions, appendicitis is the most common surgical cause of abdominal pain in children.

Appendicitis

Since the review by Hennelly and Bachur [43], several studies have further examined the evaluation and treatment of appendicitis. The Alvarado and Pediatric Appendicitis (PAS) Scores are two systems that estimate the likelihood of appendicitis based on symptoms (pain migration, anorexia, nausea/emesis), physical examination findings (fever, right lower quadrant tenderness), and laboratory criteria (leukocytosis and neutrophilia). Wu *et al.* [3] found greater accuracy of the Alvarado score compared with PAS; optimal cutoff for both scores varied with symptom duration. Elevated inflammatory biomarkers, such as interleukin 6 (IL-6) [44,45▪], IL-8 [44], CRP [4,44], and CD-64 [45▪], have been associated with appendicitis compared with noninflammatory and nonsurgical causes of acute abdominal pain. In a retrospective study in Israel, ultrasound appeared to streamline appendicitis evaluation, as increased use correlated with decreased hospital admissions [46]. For perforated appendicitis treatment, a single-center, randomized, prospective study found more rapid recovery with immediate operation compared with antibiotics/delayed appendectomy [47▪]; of note, patients with a well-formed abscess were excluded from this study.

Cholecystitis and pancreatitis

Cholecystitis and pancreatitis generally present with upper abdominal pain in children. Obesity

and Hispanic ethnicity are risk factors for symptomatic gall bladder disease in children [48]. Symptomatic cholelithiasis and complicated obstructive disease are the most common indications for pediatric cholecystectomy [48]. Point of care ultrasound within the emergency department may be helpful for diagnosis of cholecystitis [49]; sonographic criteria include gall bladder wall thickening (>3 mm), pericholecystic fluid, gall stones, and sonographic Murphy's sign.

Pancreatitis often presents with abdominal pain, vomiting, and epigastric tenderness [50,51], although these manifestations are less common in infants and toddlers [52]. Atlanta diagnostic criteria for acute pancreatitis are presence of at least two of the following three parameters: typical abdominal pain, elevated amylase/lipase more than three times the upper limit of normal, and/or confirmatory findings on cross-sectional abdominal imaging [50]. Serum cationic trypsinogen and urine trypsinogen activator peptide are not commonly available but may be helpful in the future [50]. Imaging includes ultrasound to assess for gall stones and alternate causes of abdominal pain; contrast-enhanced CT scan is reserved for delayed assessment of pancreatitis complications such as necrosis [50]. Common causes of pancreatitis are gall stones/sludge, medications, systemic disease, and trauma [50,51,53]. Ma *et al.* [54] found association of gallstone pancreatitis with increased aspartate aminotransferase, obesity, and Hispanic ethnicity. Specific medications implicated in pancreatitis include valproic acid, L-asparaginase, prednisone, and 6-mercaptopurine [50,53]; forms of systemic disease include sepsis, shock, hemolytic–uremic syndrome, and systemic lupus erythematosus [50,53]. Severe outcomes, including mortality, surgery on the pancreas, acute renal failure, respiratory failure, severe gastrointestinal bleed, or shock during the initial admission and/or subsequent pseudocyst development, are more likely with white blood count greater than 18 500/μl, calcium less than 8.3 mg/dl, and blood urea nitrogen rise higher than 5 mg/dl [53].

Methods to image and treat pediatric biliary disease include MRI, endoscopic retrograde cholangiopancreatography (ERCP), and single-incision laparoscopy (SIL). Functional MRI has promise as a mode for imaging biliary and pancreatic disease [55]. Dynamic biliary tree flow patterns can establish the diagnosis of cholecystitis, pancreatitis, and choledocholithiasis. ERCP in children appears safe with low complications; post-ERCP pancreatitis occurs with low frequency (3%) [56]. SIL cholecystectomy in children has favorable outcomes compared with the standard laparoscopic approach; in a single-center review, operative time was longer but appeared to shorten with increased surgeon experience, and postoperative narcotic requirement was lower [57].

Pelvic inflammatory disease

In an older girl, pelvic inflammatory disease (PID) should be included among potential causes of acute lower abdominal pain. In the USA, older children and young adults have a disproportionate incidence of sexually transmitted diseases (STDs); although 15–24-year-olds account for 25% of the sexually experienced population, almost half of new sexually transmitted infections occur in this age group [58]. Because PID causes variable and nonspecific symptoms, the Centers for Disease Control has established PID diagnostic criteria: one or more findings of cervical motion tenderness, uterine tenderness, or adnexal tenderness on pelvic examination; and one or more additional findings of oral temperature higher than 38.3°C, abnormal cervical or vaginal mucopurulent discharge, presence of abundant numbers of white blood cells on saline microscopy of vaginal fluid, elevated erythrocyte sedimentation rate, elevated CRP, or laboratory documentation of cervical infection with *Neisseria gonorrhoeae* or *Chlamydia trachomatis* [59]. Balamuth *et al.* [60] recently found poor baseline knowledge of PID diagnostic criteria and standard treatment among pediatric emergency medicine practitioners; two methods to decrease this knowledge gap were tested.

Other causes of peritonitis

Rarer causes of abdominal pain with peritonitis are infected congenital anomalies [29,61,62], complicated Crohn's disease, and intestinal perforation due to foreign bodies [40] and peptic ulcer disease. In pediatric Crohn's disease, phlegmon and abscess formation can present with focal peritonitis; in a multicenter registry, the cumulative incidence of abscess increased with time from diagnosis (4.7% at 1 year, 8.8% at 5 years, 16.1% at 10 years) [63▪].

ACUTE ABDOMINAL PAIN WITHOUT SYMPTOMS OF OBSTRUCTION OR INFLAMMATION

Surgical causes of acute abdominal pain and nonbilious emesis without inflammatory signs such as fever or peritonitis include ovarian torsion in girls, and epiploic fat or omental torsion.

Ovarian torsion and cyst rupture

Ovarian disorders, such as cyst rupture and torsion, must be considered when evaluating girls with sudden onset abdominal pain [64,65]. Abdominal pain may be intermittent and recurrent, and accompanied by emesis [64,66]; pain migration, fever, rebound tenderness, leukocytosis, and elevated CRP are less likely with ovarian torsion than appendicitis [66]. Adnexal abnormalities are most frequently detected by ultrasound, which is highly sensitive [66–68]; abdominal radiographs may suggest the presence of a mass if large, or reveal calcifications if due to a mature teratoma [66,69]. Symptoms and ultrasound findings may not distinguish ovarian or tubal cyst torsion from ovarian cysts with hemorrhage or rupture [67,68]. Inflammatory biomarkers, such as IL-6 and CD-64, may help differentiate surgical from nonsurgical disease, and appendicitis from ovarian torsion, respectively [45▪].

To maximize the chance of ovarian salvage, operative detorsion of the ovary is emergent and may be performed laparoscopically [65]. Oophorectomy is reserved for suspected underlying malignancy, because even blue discoloration following detorsion is not necessarily indicative of ovarian necrosis [65]; oophorectomy may also be performed in a delayed fashion if a neoplasm is suspected on follow-up imaging. Pathology of resected ovaries has revealed predominantly benign diseases [65], such as mature cystic teratomas, and mucinous and serous cystadenomas [64,65].

Omental and epiploic fat infarction

Focal abdominal pain without inflammatory symptoms can occur with twisting and subsequent infarction of omentum or epiploic fat. By CT scan, torsed epiploic fat appears as an ovoid, fat-density area anterior or anterolateral to the colon, with adjacent peritoneal enhancement; with omental torsion, swirling blood vessels within fat anterior to the bowel are visible. On ultrasound, both appear echogenic at the location of maximal tenderness [70]. In several retrospective series, omental infarction occurred in school-aged, overweight, predominantly male children; diagnosis was generally established by CT scan, and ultrasound had low to moderate sensitivity [71–73]. Debate exists over whether omental infarction is best managed by observation or laparoscopic resection [71–73].

CONCLUSION

Distinguishing surgical from nonsurgical causes of acute abdominal pain in children remains a challenge. Careful consideration of patient characteristics, symptoms, and physical examination and laboratory findings can narrow diagnostic considerations, and thereby guide imaging selection and subsequent treatment. Despite widespread use, CT scan is not always the optimal imaging modality for the evaluation of acute abdominal pain in children.

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Conflicts of interest

There are no conflicts of interest.

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- ▪ of special interest
- ▪▪ of outstanding interest

Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 432).

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