An Update on Common Gastrointestinal Emergencies

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KEYWORDS

• Malrotation • Appendicitis • Pyloric stenosis • Hirschsprung disease
• Intussusception

KEY POINTS

- Infants with pyloric stenosis often present with progressive nonbilious emesis with an intact appetite.
- Although most of the infants with malrotation and volvulus present in the neonatal period maintain a high index of suspicion in any older infant or child presenting with bilious emesis and abdominal pain.
- Intussusception should be considered in an infant presenting with lethargy or altered level of consciousness.
- Laboratory testing is of limited utility in children with appendicitis; careful history and physical examination are more likely to aid in the diagnosis.
- Chronic constipation and malnutrition are common symptoms in delayed presentation of Hirschsprung disease.

IDIOPATHIC HYPERTROPHIC PYLORIC STENOSIS

Background/Epidemiology

Idiopathic hypertrophic pyloric stenosis (IHPS) is a condition where the pyloric muscle abnormally thickens and as a result, there is delayed gastric emptying. This disease affects 2 to 5 in 1000 live births with a male to female predominance of 4:1.[1–3] The exact cause is unknown, despite a multitude of studies that suggest various genetic and environmental associations. Classically, this condition is described as being more common in firstborn males. Recent epidemiologic studies do not suggest a unique position for firstborns, but rather a decline in risk with the increasing birth order.[3] A notable environmental association with IHPS is the sharp decline in incidence after the back to sleep campaign was promoted in Denmark and Sweden.[3] Similarly, the expanded use of erythromycin for pertussis has been associated with

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an increased incidence of IHPS. There were 2 studies that identified clusters of increased incidence of pyloric stenosis after the use of erythromycin. Although the increased risk is small, there are data to suggest that infants younger than 2 weeks of are at greatest risk for developing IHPS when exposed to erythromycin.[3,4]

Clinical Features

Infants with IHPS typically are healthy at birth. Symptoms begin with small amounts of emesis with progression to large-volume emesis as the pyloric muscle hypertrophies. The most common age of presentation is between 2 and 5 weeks of life. IHPS is rare after 12 weeks of age. The emesis is frequently projectile and nonbilious. Protracted emesis predisposes to a Mallory-Weiss tear and subsequent hematemesis. Despite persistent emesis, these infants usually have a normal, intact appetite. Early in the course of disease, infants are often well appearing. As the emesis advances, infants begin to show clinical signs of dehydration. Infants with emesis for an extended period of time may show signs of growth failure often appearing cachectic with loss of subcutaneous fat and loose skin.[5] Additional physical examination findings include a visualization of gastric peristalsis and a palpable pyloric mass or "olive," present in 44% to 48% of cases.[6,7] In cases of severe contraction metabolic alkalosis, infants may present with apnea.[8]

Laboratory Analysis

Electrolytes may be normal early in disease. As a result of protracted emesis with dehydration, a hypochloremic metabolic alkalosis may develop. With earlier detection of disease, the incidence of a hypochloremic alkalosis has slowly declined.[7]

Radiographic Studies

Plain radiographs

Plain radiographs of the abdomen most often are nonspecific and may appear normal. Although neither sensitive nor specific, the presence of gastric dilatation with a paucity of distal bowel gas may be suggestive of IHPS. Infrequently, gastric peristalsis may be visualized as the "caterpillar sign" giving the false appearance of a "double bubble" sign (**Fig. 1**).

Pyloric ultrasound

Ultrasound (US) of the pylorus is the preferred diagnostic study with high sensitivity and specificity, which approaches 98% and 100%, respectively.[9,10] Classic sonographic findings include a thickened pylorus with a length greater than 15 mm and diameter greater than 3 mm. An additional US finding is a prolapsed, hypertrophied pyloric mucosa protruding into the gastric antrum, also known as the "antral nipple" sign (**Fig. 2**).[11] The advantages of sonography over an upper gastrointestinal series are that it requires no radiation exposure, may be rapidly obtained, and does not depend up on the transit of gastric materials across the pyloric canal.

Upper gastrointestinal series

An upper gastrointestinal series (UGI) is frequently obtained when differentiating between other causes of neonatal emesis. In IHPS, an UGI series may demonstrate a failure of relaxation of the prepyloric antrum and a string of contrast through the mucosal interstices that outlines the canal called the "string sign."[12] The advantage of an UGI series is that it provides additional information regarding esophageal anatomy and motility, especially when considering additional causes of neonatal emesis such as malrotation, gastroesophageal reflux disease, and other intestinal stenoses and atresias.

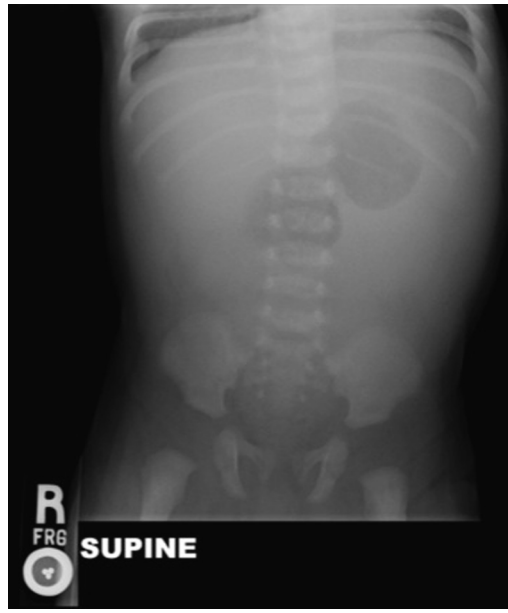

Fig. 1. Pyloric stenosis. Plain abdominal radiograph demonstrates the presence of a caterpillar sign and a paucity of gas distal to the pylorus.

Management

Initial management in patients with IHPS is focused on intravenous hydration with isotonic fluids. Diagnostic workup may include both laboratory testing and radiologic studies as mentioned earlier. Infants are often admitted for intravenous (IV) hydration. Electrolytes abnormalities are corrected before surgical repair to reduce perioperative morbidity. Definitive therapy for IHPS is open or laparoscopic pyloromyotomy. Once surgically repaired, recurrence rarely occurs.

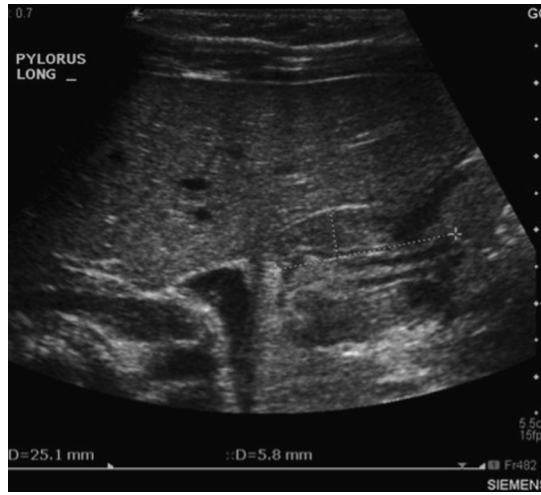

Fig. 2. Pyloric stenosis. Longitudinal abdominal ultrasonography shows thickened muscle and redundant mucosa consistent with the antral nipple sign.

MALROTATION

Background

During normal embryonic development between the 4th and 10th weeks of gestation, a 270° counter-clockwise rotation of the proximal and midgut occurs before settling into the abdomen. Malrotation occurs as a result of incomplete rotation of the bowel with an abnormal fixation of the mesentery of the bowel. It is within this abnormal fixation that a volvulus may occur.

Epidemiology

In the United States, the prevalence of malrotation in infants under the age of 1 year is 3.9 in 10.000 live births.[13] Twenty-five percent of patients with malrotation will present within the 1st month of life and 90% by the 1st year of life. Mortality depends on the degree of bowel ischemia during surgery.[14]

Clinical Features

The classic presentation of malrotation with volvulus is a neonate with sudden onset of bilious emesis. However, there is considerable variability in presentation depending on severity of disease. Bilious emesis indicates any obstruction distal to the ampulla of Vater and is not pathognomonic of malrotation with volvulus. A prospective study identified that 62% of infants with bilious emesis did not have anatomic obstruction; however, further imaging is still necessary to rule out malrotation as a potential cause.[15] Infants frequently present with a normal history or more subtle findings such as feeding problems or gastroesophageal reflux associated with a failure to thrive.[16]

Infants may have a completely normal physical examination. The presence of an acute abdomen, although rare, is a poor prognostic indicator.[16] Approximately 10% to 15% of infants will present with gross hematochezia or guaiac positive stool. The presence of blood is another indicator of poor prognosis due to risk of impending bowel gangrene.[16]

Laboratory Analysis

Laboratory studies are rarely diagnostic; however an elevated white blood cell count, C-reactive protein, lactic acid, and glucose have been associated with bowel ischemia.[17]

Radiographic Studies

Plain radiographs

Plain radiographs of the abdomen often vary from nonspecific findings to a distal bowel obstruction (**Fig. 3**). The presence of a normal bowel gas pattern does not exclude the possibility of malrotation.[14] Plain radiographs and decubitus films of the abdomen are helpful in determining the presence of free intraperitoneal air. The presence of free air requires immediate pediatric surgical consultation and operative intervention.

Upper gastrointestinal series

The preferred imaging modality for determining the presence of malrotation is an UGI series. The diagnostic finding on the UGI series is the abnormal positioning of the duodenal-jejunal junction (**Fig. 4**). Normal positioning of the junction should be located left of the left vertebral pedicle at the level of the inferior margin of the duodenal bulb. In malrotation, the junction is present to the right of the vertebral body.[14] Although the UGI series is the diagnostic imaging of choice, the false-positive rate may be as

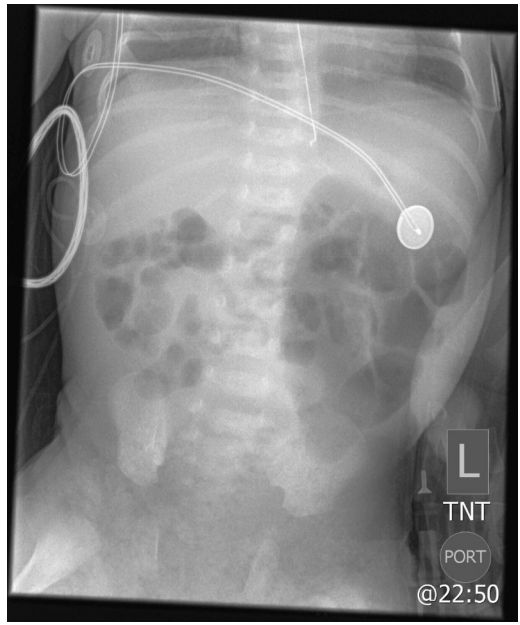

Fig. 3. Malrotation with volvulus. Plain radiographs show the presence of large bowel on the left side of the abdomen.

high as approximately 15% with a false-negative rate of 2% to 3%.[18] Sensitivity of the UGI series for malrotation approaches 93% to 100%.[19] Common reasons for the false positives are usually the result of normal anatomic variation such as a wandering duodenum, duodenum inversum, or mobile duodenum. Additional reasons for the

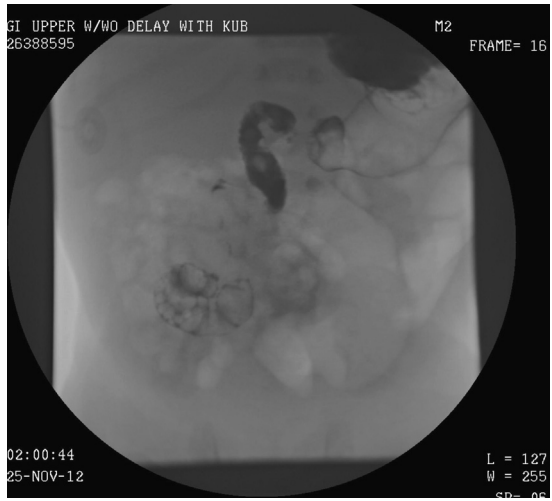

Fig. 4. Malrotation with volvulus. Upper gastrointestinal series demonstrates inferior displacement of the duodenal jejunal junction (DJJ) to the right. DJJ does not pass to the left of the spine and does not rise to the level of the duodenal bulb. Proximal small bowel appears on the right side of the abdomen. Likely corkscrew pattern of duodenum indicating volvulus.

displacement of the junction include a dilated stomach, splenomegaly, renal agenesis, or liver transplantation.[18]

Abdominal ultrasound

US is frequently obtained due to the low risk of radiation exposure and ease of accessibility. Suggestive findings include a reversal of the relationship of the superior mesenteric artery (SMA) to the superior mesenteric vein (SMV). A "whirlpool sign," a swirling, whirlpool-like shape seen when the SMV and mesentery wrap around the SMA in a clockwise direction, may also be visualized on transverse ultrasonography. Several studies have documented variable sensitivity and specificity of this imaging modality.[14]

Management

Once access has been obtained, fluid resuscitation and antibiotics should be initiated, followed by immediate surgical consultation. Unstable patients requiring airway stabilization and aggressive fluid resuscitation will likely require emergent surgical intervention. Further imaging, as described earlier, should be obtained once the patient is adequately resuscitated.

Surgical repair

Originally described by William Ladd in 1930, the Ladd procedure is still performed today with (1) detorsion of the bowel when volvulus is present, (2) lysis of duodenal bands, (3) broadening the mesentery to separate the duodenum and cecum as far away as possible, (4) placement of the small bowel to the right side of the abdomen, and (5) placement of the colon to the left side of the abdomen.[20] The laparoscopic procedure has no difference in complication rates and decrease in length of stay when compared with an open Ladd procedure.[21] Postoperative complications include bowel obstruction from adhesions, volvulus, and incisional hernias.[21,22]

INTUSSUSCEPTION

Background/Epidemiology

Intussusception is the most common cause of pediatric small bowel obstruction and afflicts approximately 56 in 100,000 children annually.[23] The disease involves the telescoping of the bowel into itself, usually including both the large and small bowel. Most intussusceptions are ileocolic and 90% to 95% are presumed to be the result of lymphoid hyperplasia. The remainder is the result of pathologic lead points.[24,25] The typical age of presentation is between 6 months and 2 years, with a peak incidence between 5 and 9 months.[26,27] There have been rare reported cases of infants under 2 months of age with intussusception. Pathologic lead points, such as Meckel diverticulum, benign tumors, or vasculitis from Henoch-Schonlein Purpura, are more common in children older than 2 years of with an incidence of 22% of intussusception cases in this age group.[28]

Clinical Features

The classic presentation of intussusception is a clinical triad of colicky abdominal pain, currant jelly stools, and a palpable abdominal mass. Unfortunately, this triad is present in less than 40% of children.[29–32] Atypical presentations are more common in young infants and older children. In infants younger than 4 months of age, painless intussusception may be present in up to 40%. Other nonspecific neurologic symptoms such as lethargy or altered level of consciousness may also occur.[27,33] Children older than 2 years of age tend to present with more subacute or chronic abdominal pain and few have rectal bleeding.[28]

Laboratory Analysis

Laboratory studies are rarely useful in aiding in the diagnosis of intussusception.

Radiographic Studies

Plain radiographs

In the emergency department, supine and lateral decubitus films are often obtained as screening tools for intussusception. However, the sensitivity and specificity of these studies are very low. Approximately 24% of patients with confirmed intussusception may have normal radiographs.[34] Radiographic findings suggestive of intussusception include the presence of a small bowel obstruction, the appearance of a soft tissue mass in the right upper quadrant, paucity of gas in the right lower quadrant, "target sign," and the "crescent sign." The target sign, seen in approximately 29% of patients with intussusception, is comprised of 2 concentric radiolucencies in the right upper quadrant outlining a soft tissue mass to the right of the spine overlying the kidney.[34] The crescent sign is the presence of a curvilinear mass in the transverse colon beyond the hepatic flexure (**Fig. 5**). The triad of intestinal obstruction, intracolonic mass, and paucity of gas in the right lower quadrant occurs in only 1% of patients.[34] One of the most sensitive indicators of intussusception is the presence of air in the ascending colon visualized on at least 2 of the 3 views of the abdomen reaching a sensitivity of 96% and specificity of 41%.[35]

Abdominal ultrasound

Abdominal US has emerged as the primary diagnostic tool for intussusception with high sensitivity ranging from 98% to 100% and specificity from 88% to 100%.[36–38] Sonographic findings on transverse imaging are a hypoechoic outer rim of homogeneous thickness with a central hyperechoic core designated the "doughnut" or "target" sign (**Fig. 6**).[39] On longitudinal scans, there is an appearance of a

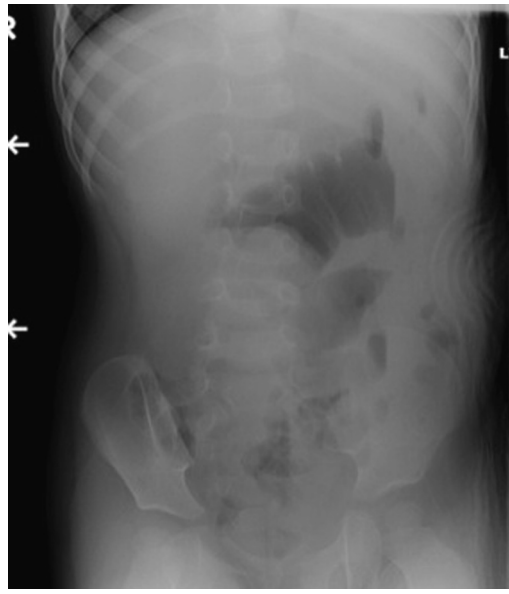

Fig. 5. Intussusception. Plain film demonstration of intracolonic mass. Decubitus plain abdominal radiograph demonstrates soft tissue mass within the colon caused by the head of the intussusception (intussusceptum).

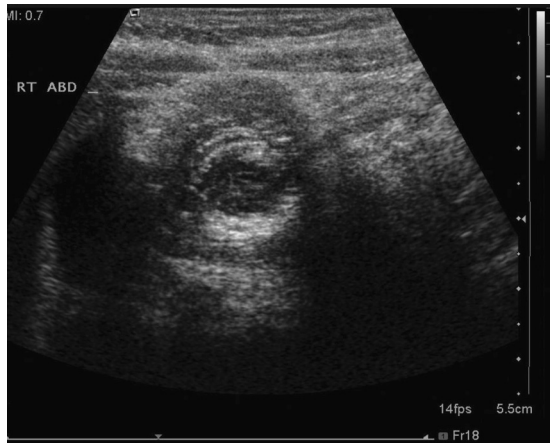

Fig. 6. Intussusception. Abdominal ultrasonography shows an outer hypoechoic region surrounding an echogenic ring also known as the "target sign".

"pseudokidney," a hyperechoic tubular center covered on each side by a hyperechoic rim producing a kidneylike appearance.[26,39]

Management

Initial management should focus on fluid resuscitation. Antibiotics and immediate surgical consultation are required if perforation or peritonitis is suspected. Once a radiographic diagnosis of intussusception is made and perforation is not suspected, nonoperative management is pursued with an air or contrast enema performed by a radiologist (**Fig. 7**). There is considerable controversy in the literature between air and contrast enema with little difference in rates of perforation and recurrence (**Table 1**).

Children at risk for enema reduction failure include infants younger than 3 months, children older than 5 years, duration of symptoms greater than 48 hours, presence of

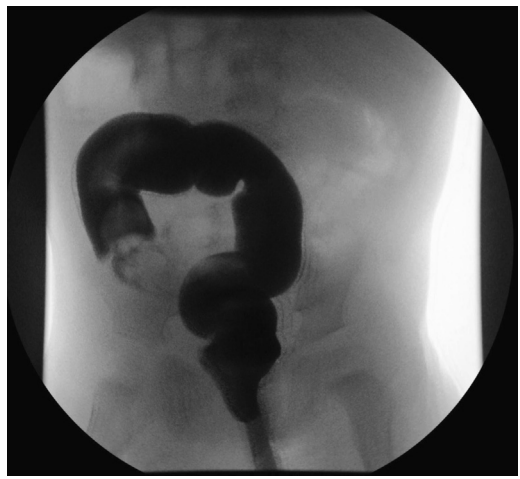

Fig. 7. Intussusception. Contrast enema demonstrating reduction of intussusception.

Table 1
Comparison of air versus contrast enema reduction for intussusception

	Air Enema	Contrast Enema
Success Rate	60%–90%	60%–80%
Rate of Perforation	<3%	<1%
Disadvantage	Tension pneumoperitoneum	Contrast peritonitis
Advantages	Lower radiation dose Better control of intracolonic pressures	Better anatomic definition

hematochezia, significant dehydration, or evidence of small bowel obstruction on plain radiograph.[5] More recent retrospective studies indicate that age may not be a risk factor for failed reduction. These studies demonstrated that a duration of symptoms greater than 24 hours, bloody diarrhea, and lethargy were the most significant risk factors in failed enema reductions.[40,41]

Management after successful enema reduction of intussusception requires observation for recurrent intussusception or bowel perforation. Previous studies recommended admission for 48 hours. This recommendation has not been validated leading to considerable debate on management. The overall recurrence rate for intussusception following enema reduction is 10% with 0% to 5.3% in the first 24 hours.[42] This low early recurrence rate for intussusception demonstrates that few patients would benefit from inpatient observation. More recent studies suggest that, given the low early recurrence rate for enema-reduced intussusception, an emergency department observation for a 6-hour period may be a safe alternative to inpatient management.[43]

APPENDICITIS

Epidemiology

Appendicitis is the most common surgical emergency of childhood afflicting 71,000 children younger than 15 years of age with a male to female ratio of 1.4:1.[44] The incidence is highest in boys aged 10 to 14 years (27.6 per 10,000 population per year) and girls aged 15 to 19 years (20.5 per 10,000 population per year).[45] Children younger than 5 years account for less than 5% of all appendicitis, which poses a diagnostic challenge for clinicians.[46]

Clinical Features

The classic presentation of appendicitis is periumbilical abdominal pain and nausea that migrates to the right lower quadrant followed by emesis and fever.[46,47] Although this sequence of events is present in approximately 50% of adults, it is less common in children.[47] The distinctive physical examination finding of right lower quadrant tenderness is considered to be the single most important diagnostic tool for appendicitis.

Unfortunately, children younger than 5 years are a diagnostic dilemma to clinicians as the result of a lack of communication skills and difficulty in examining these children. Risk factors for missed appendicitis include younger children (5.3 vs 7.9 years), onset of emesis before abdominal pain, constipation, diarrhea, upper respiratory symptoms, lethargy, or irritability.[47] Missed appendicitis not only increases the rate of perforation leading to greater morbidity and mortality but also poses a significant medicolegal risk.[47–50] One study noted that appendicitis was the second most prevalent condition in pediatric malpractice claims from 1985 to 2005 caused by error in diagnosis.[51] Rates of misdiagnosis from initial symptoms vary across age groups.

Most children youger than 3 years old are missed at initial presentation, with some studies noting a missed diagnosis rate approaching 100%.[46,52] In preschool-aged children, the rate of missed appendicitis improves ranging from 19% to 57%. In school-aged children, the rate of missed appendicitis further improves to 12% to 28%.[46,47]

Clinical Prediction Rules

Several clinical predictions rules (CPRs) have been proposed and validated including the Alvarado Score/MANTRELS (Migration, Anorexia, Nausea/vomiting, Tenderness in the right lower quadrant, Rebound pain, Elevation in temperature, Leukocytosis, Shift to the left), Low-Risk Appendicitis Rule, and Pediatric Appendicitis Score (PAS). More recent systematic reviews found the PAS and Alvarado scores to be the most validated CPRs; however, they do not reach the 4-rule performance benchmark of high-performing CPRs.[53] The more recent validation of the Low-Risk Appendicitis Rule yielded a sensitivity of 98%, specificity of 24%, and negative predictive value of 95%when the rule was refined.[54] Components of this refined rule include (1) an absolute neutrophil count of $6.75 \times 10^3/\mu L$ or less and no maximal tenderness in the right lower quadrant or (2) an absolute neutrophil count of $6.75 \times 10^3/\mu L$ or less with maximal tenderness in the right lower quadrant but no abdominal pain with walking/jumping or coughing. Recent commentary on this study noted the low, but not zero, risk for appendicitis; clinicians need to balance the risks of missing appendicitis with the increased risk of negative appendectomies and the potential long-term risks associated with exposure to ionizing radiation.[55]

Laboratory Analysis

Several laboratory studies have been evaluated as potential markers for children with suspected appendicitis. Most data have been equivocal at best.

Complete blood count

A white blood cell (WBC) count and differential is commonly ordered in children, despite limited diagnostic sensitivity and specificity for appendicitis. The WBC count has been studied frequently in the adult population with fewer studies in the pediatric population. Some studies have suggested that neutrophilia may be more sensitive than an elevated WBC count. A more recent study demonstrated that the combination of neutrophilia and increased WBC count results in a higher sensitivity (79%) than either test independently.[56] Neither the WBC count nor the neutrophil count allow for differentiation between perforated from nonperforated appendicitis.[47,57]

C-reactive protein

C-reactive protein (CRP), a nonspecific inflammatory marker, is the most frequently studied biomarker in appendicitis. A meta-analysis performed in the adult population found the WBC count more sensitive than the CRP.[58] Studies in the pediatric population suggest CRP elevations to be more sensitive in children with perforated appendicitis or abscess formation. A study of 209 children with 115 diagnosed with appendicitis established an optimal CRP cutoff value of 3 mg/dL. This CRP cutoff value coincided with a specificity and sensitivity of 65% and 71% respectively. With a CRP value greater than 3 mg/dL and WBC count greater than 12 cells/1000 mm^3, there was a further increase in specificity to 91% and a decrease in sensitivity to 42%.[59] Clinicians should maintain a higher index of suspicion for appendicitis and consider surgical consultation with these elevated values.

Gene expression

Recent advances in the study of gene expression are emerging as diagnostic tools for various disease processes including appendicitis. One study using leukocyte gene expression and cytokine levels identified 80% of the prospective cohorts with appendicitis.[60] Further investigation is warranted.

Radiographic Studies

Plain radiographs

Once thought to be useful in the diagnosis of appendicitis, most recent studies indicate that plain radiographs of the abdomen are often normal or misleading.[46] Previously, a calcified appendicolith identified on plain film was considered diagnostic. More recent studies have demonstrated appendicoliths in only 13% to 22% of cases with appendicitis and 1% to 2% of cases without appendicitis.[46,61,62] As a result of its limited utility, evidenced-based guidelines suggest the use of plain radiographs only when the patient's presentation is concerning for bowel obstruction, free air, mass, or nephrolithiasis.[63]

Abdominal ultrasound

US to evaluate the appendix is the imaging modality of choice in many centers. It is often considered an extension of the physical examination and serves the advantages of being noninvasive, avoiding conscious sedation, and avoiding ionizing radiation exposure.[64,65] The major disadvantages of abdominal US in the evaluation of appendicitis are that it is highly operator dependent and is less accurate than computed tomography (CT).[66] A meta-analysis comparing US to CT in children demonstrated higher pooled sensitivity and specificity for CT at 94% and 95%, respectively, over US, 88% and 94%, respectively.[66] For children with obesity, one study has established that the specificity, sensitivity, and negative predictive value were significantly lower than in nonobese children.[67]

Computed tomography

CT has been the diagnostic imaging of choice, secondary to its widespread availability at most major hospitals and emergency rooms. Compared with US, CT has superior accuracy (**Fig. 8**). An 18-year retrospective institutional review of an adult emergency department found a significant reduction in the negative appendectomy rate following an increase in the proportion of patients who had a preoperative CT.[68] Although CT of the abdomen and pelvis is frequently performed with various methods of contrast, a recent meta-analysis observed that the introduction of high-resolution CT may deter

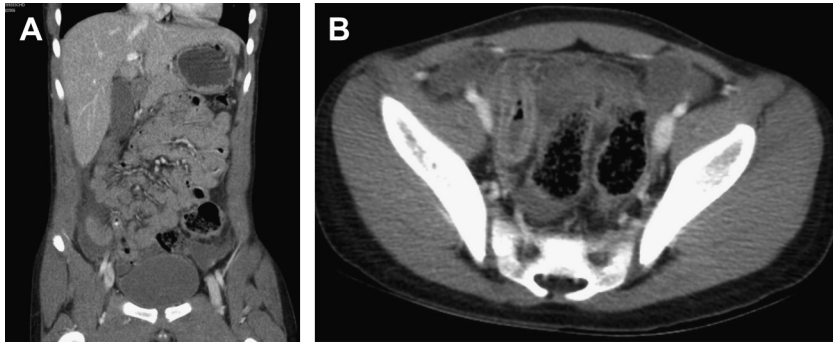

Fig. 8. Appendicitis. Computed tomography. (*A*) Coronal images show appendicolith. (*B*) Images demonstrate dilated and inflammed appendix.

the eventual use of contrast altogether in the adult population.[69] This comprehensive review of 7 studies found the pooled sensitivity and specificity of noncontrast CT to be 93% and 96%, respectively with a false negative rate of 7.3%. Although well described in the adult literature,[70] there have been few studies in the pediatric population that allow the exclusion of appendicitis from a normal CT scan with a nonvisualized appendix. In one retrospective case control study of pediatric patients, the negative predictive value of a normal CT scan with a nonvisualized appendix was 98.7%.[71]

The widespread availability and fairly good sensitivity of CT is offset by the limitations of equivocal scans, exposure to ionizing radiation, and need for sedation in young children. Unfortunately, the risk of exposure to ionizing radiation is a growing concern. A comprehensive review of the effects of ionizing radiation derived from prediction models by Wakeford[72] validated that low levels of radiation exposure are associated with higher risk of childhood leukemia. Before the review by Wakeford, a Markov-based decision model found that a single abdominal CT in a 5-year-old child has a lifetime risk of radiation-induced cancer of 26.1 per 100,000 in women and 20.4 per 100,000 in men. This study also outlined the utility of US followed by CT if the initial US result was negative. This protocol demonstrated a reduction of CT scans with a concomitant reduction in radiation-induced malignancy by more than 50%.[73]

Magnetic resonance imaging

Magnetic resonance imaging (MRI) is emerging as a promising radiographic study in the diagnosis of appendicitis. MRI of the abdomen was originally studied in pregnant women. Given the reduced risk of ionizing radiation exposure, the concept was applied to the pediatric population. A recent study investigated 208 children after the implementation of a four-sequence expedited noncontrast MRI protocol and found the sensitivity and specificity to be 97% and 97.6%, respectively (**Fig. 9**).[74] The high sensitivity and specificity is partially offset by several limitations: a requirement for sedation in young children, potentially high cost, and lengthy procedural time. With the aforementioned study using a noncontrast expedited MR protocol, the median time from procedure request to final report was 164 minutes. Cost analysis

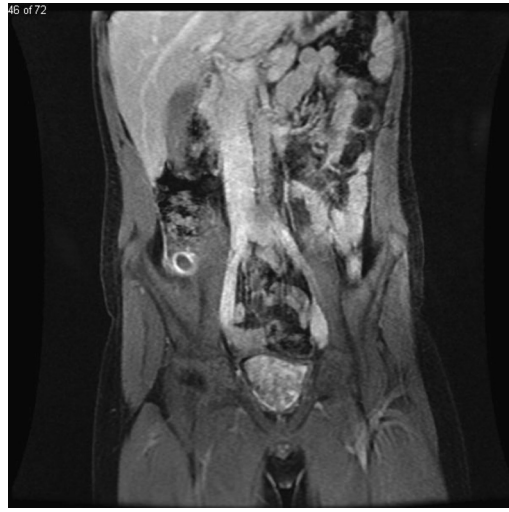

Fig. 9. Appendicitis. MRI of the abdomen and pelvis shows markedly dilated and inflamed appendix.

has been briefly addressed in the European literature,[75] and the most recent pediatric study indicated the cost of a noncontrast MRI to be $104 more than the cost of a CT with IV contrast; however, this is highly institution dependent and warrants further investigation.

Management

Currently, the standard treatment of choice is appendectomy. For nonperforated appendicitis, laparoscopic appendectomy is the preferred surgical approach. Recent advances in surgical techniques have facilitated the single umbilical incision laparoscopy for appendectomy (SILA). A systematic review of 9 studies in the adult population demonstrated no significant difference in operative time, length of stay, pain scores, and conversion or complication rates between SILA and conventional laparoscopic appendectomy.[76] Several pediatric studies have demonstrated similar findings[77]; a prospective study of 415 children using SILA validated its feasibility in the pediatric population.[78]

In cases of perforated appendicitis with abscess formation, the preferred approach is percutaneous drainage of the abscess and IV antibiotics followed by an interval appendectomy.[65] One study comparing early appendectomy to interval appendectomy in cases of perforated appendicitis with abscess formation revealed no differences in length of hospitalization, rate of abscess recurrence, or overall charges.[79]

HIRSCHSPRUNG DISEASE

Background/Epidemiology

Hirschsprung disease (HD), also known as aganglionic megacolon, is the absence of parasympathetic ganglion cells of Auerbach plexus in a variable portion of the distal gut. The most classic form, referred to as "short segment" disease, is limited to the rectosigmoid colon and accounts for 80% of all cases. The incidence is 1 in 5000 with a strong male to female predominance of 4:1.[80] "Long segment" disease extends proximal to the sigmoid colon and can involve the entire large bowel.

Hirschsprung disease may be associated with other congenital cardiac, neurologic, gastrointestinal, or urologic abnormalities. Trisomy 21 is the most common chromosomal abnormality associated in 10% of these infants.[80]

Clinical Features

Eighty percent of infants with Hirschsprung disease present within the neonatal period. The most common presenting symptom in 90% of neonates is the failure to pass meconium in the first 24 hours.[81] Additional symptoms include bilious emesis, infrequent explosive diarrhea, jaundice, and poor feeding. The presenting symptoms in older children vary from the neonatal time period as these patients often have chronic constipation, progressive abdominal distention, and malnutrition.[82] Most of the older children with HD have short segment disease.

Ten percent of children with HD present with fever, abdominal distention, abdominal pain, and sepsis. This is more commonly seen in neonates and infants.[83]

Laboratory Studies

Laboratory studies are of limited utility. WBC count and CRP may be elevated, but these are nonspecific markers.

Radiographic Studies

Plain radiographs are frequently obtained as screening tools.

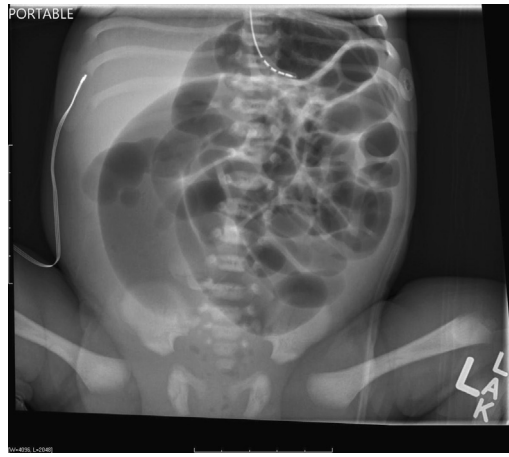

Fig. 10. Hirschsprung Disease. Plain radiograph of the abdomen shows a dilated small bowel and proximal colon with an empty rectum.

Plain radiographs in Hirschsprung disease demonstrate a dilated small bowel and proximal colon with an empty rectum (**Fig. 10**). A contrast enema obtained on an unprepped bowel will reveal a transition zone that reflects the joining of the aganglionic bowel with the dilated ganglionic bowel (**Fig. 11**).[80] Delayed barium evacuation may be noted in plain radiographs taken after the contrast enema is complete (**Fig. 12**).

Management

Fluid resuscitation and antibiotics should be initiated in patients who demonstrate signs of Hirschsprung disease–associated enterocolitis. In stable patients, the

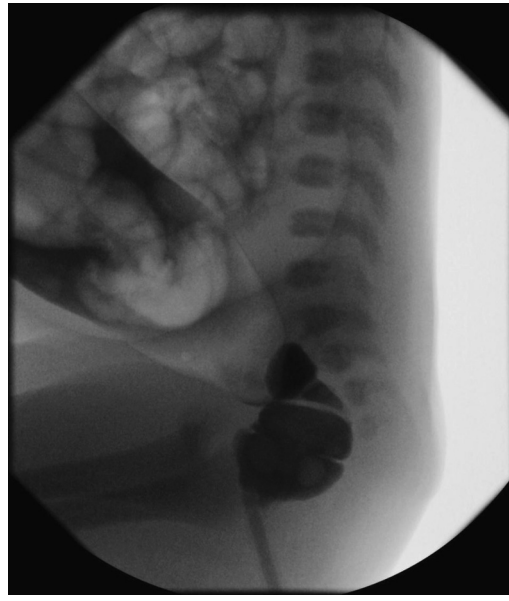

Fig. 11. Hirschsprung Disease. Contrast enema with demonstrating the presence of a transition zone.

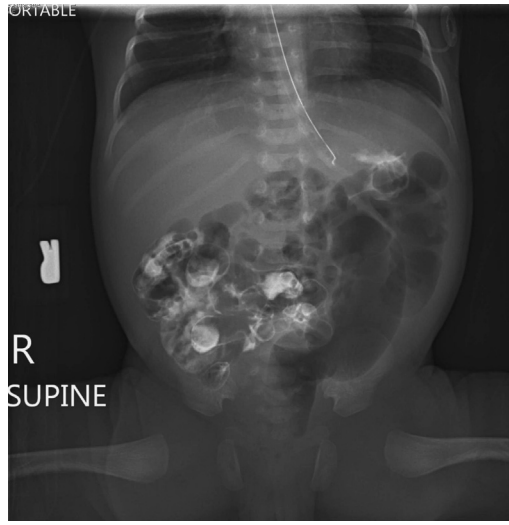

Fig. 12. Hirschsprung Disease. Plain abdominal radiography with delayed barium evacuation after completion of a contrast enema.

diagnosis may be made via a contrast enema or rectal suction biopsy. Discussion with a pediatric surgeon is necessary, but not emergent, for patients presenting without associated enterocolitis.

SUMMARY

Abdominal pain is one of the most common presenting complaints in the emergency department. Early recognition of these conditions requires high indices of suspicion. Despite advances in research and treatment, the diagnosis of pediatric abdominal emergencies remains challenging and can be associated with considerable cost and morbidity. Management often revolves around appropriate fluid resuscitation, electrolyte repletion, obtaining advanced imaging, and prompt surgical consultation.

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