Diagnosis and Management of Pediatric Appendicitis, Intussusception, and Meckel Diverticulum

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KEYWORDS
• Right lower quadrant • Abdominal pain • Appendicitis • Appendectomy • Meckel diverticulum • Intussusception • Pediatric

KEY POINTS
• A classic presentation of appendicitis begins with the gradual onset of dull periumbilical pain followed by migration of this pain to the right lower quadrant.
• Initially, the appendiceal lumen becomes obstructed, leading to distention and increased intraluminal pressure.
• An increased white blood cell (WBC) count (>10,000–12,000 cells per cubic millimeter) significantly increases the odds of appendicitis. In the absence of leukocytosis and fever, appendicitis is unlikely.

APPENDICITIS
Appendicitis is the most common pediatric abdominal surgical emergency worldwide. It is estimated that 86 cases of appendicitis per 100,000 people occur annually, with an estimated 70,000 pediatric appendectomies performed in the United States each year.¹

In the last several decades, both the diagnosis and management of appendicitis have undergone significant evolution. These changes stem from a variety of causes, including recent advances in laparoscopy, concerns regarding radiation exposure, and advances in pediatric imaging. This article highlights those changes in practice as well as some of the remaining controversies in care regarding pediatric appendicitis.

Diagnosis

Demographics
The peak incidence occurs in the second decade of life with the median age at diagnosis between 10 and 11 years. Male/female ratio is 1.4:1. Appendicitis also seems to have
a seasonal variation with increased presentation of appendicitis in the summer months. However, perforated appendicitis occurs more frequently in the fall and winter.2

**Symptoms**
A classic presentation of appendicitis begins with the gradual onset of dull periumbilical pain followed by migration of this pain to the right lower quadrant. Nausea and vomiting, when they occur, typically follow the onset of pain. Anorexia and fever are also common complaints, with diarrhea occurring less frequently. Classic teaching suggests that perforation occurs at 24 to 36 hours from the onset of the first symptom, which is usually pain. When perforation occurs, the pain may increase greatly and can become more generalized. Because of the increased inflammatory response, perforation is often associated with higher fevers, increased systemic symptoms, and increased laboratory values (WBC count and C-reactive protein [CRP]).

The cause of this progression lies in the physiology of the abdomen and peritoneum. Initially, the appendiceal lumen becomes obstructed, leading to distention and increased intraluminal pressure. This distention causes stimulation of the eighth to tenth visceral afferent thoracic nerves, leading to a mild periumbilical pain. Increasing intraluminal pressure evolves into tissue ischemia, mucosal compromise, and eventual transmural inflammation. This inflammation spreads to the parietal peritoneum, leading to localized somatic pain in the right lower quadrant and constitutional symptoms of fever, nausea, emesis, and anorexia. Necrosis is followed by eventual perforation.

Although this is presented as the classic picture of appendicitis, it occurs in fewer than 50% of children. However, certain findings have been shown to increase or decrease the likelihood of appendicitis. Most significant is the evolution of midabdominal pain migrating to the right lower quadrant (likelihood ratio [LR] 1.9–3.1) and the presence of fever (LR 3.4). The absence of fever lowers the likelihood of appendicitis by two-thirds. This confusion in clinical picture leads to a delay in diagnosis, as well as increasing the rate of perforated appendicitis among pediatric patients. Because of a lack of reliable history, rates of perforation as high as 80% to 100% have been reported in children less than 3 years of age. Children aged 10 to 17 years have a lower rate of perforation at 20%.3

**Signs**
The signs of appendicitis in the pediatric population are equally difficult to interpret. Up to 44% of children present with multiple atypical clinical findings. Classic physical examination findings include tenderness to palpation and guarding in the right lower quadrant, hypoactive bowel sounds, percussive tenderness, and rebound tenderness. Certain maneuvers can be used to assess for appendicitis. The Rovsing sign involves palpating the left lower quadrant and is considered positive when the patient feels referred pain in the right lower quadrant. A positive obturator sign occurs when pain is elicited with internal rotation of the right lower extremity while it is flexed at the knee and hip. The Psoas sign is elicited while the patient lies on his or her left side and is considered positive if pain occurs with extension at the hip. Of these findings, only rebound tenderness has been shown to correlate with increased likelihood of appendicitis (LR 2.3–3.9). Lack of tenderness in the right lower quadrant reduces the likelihood of appendicitis by half.4

**Laboratory studies**
Typical studies ordered for suspected appendicitis include a complete blood count (CBC) and a comprehensive metabolic panel (CMP). Although the CMP has little diagnostic usefulness for appendicitis, it allows assessment of electrolyte status as well as evaluation for potential alternative causes of abdominal pain. An increased WBC
count (>10,000–12,000 cells per cubic millimeter) significantly increases the odds of appendicitis. Kwan and Nager showed an adjusted odds ratio of 6.5 for a WBC count greater than 12,000 per cubic millimeter. In toddlers (<4 years), a normal WBC count has a negative predictive value of 95.6%, whereas the negative predictive value in children (ages 4–11.9 years) is 89.5%. The negative predictive value of a low or normal WBC count among adolescents is 91.9%. A left shift (increased immature forms of neutrophils) also has a strong association with appendicitis, because only 3.7% of pediatric patients without left shift have appendicitis.

**Scoring systems**

Several scoring systems exist for the diagnosis of appendicitis; however, 2 systems have been evaluated in pediatric patients. The first is the Alvarado score, which was initially developed for use in the adult population. The Alvarado score is composed of 8 components with a total score of 10 (Table 1). Alvarado scores of 1 to 4 are negative for appendicitis, whereas scores from 9 to 10 are diagnostic of appendicitis. In cases of intermediate scores of 5 to 8, further diagnostic studies are required. Using these specifications, a 93% sensitivity, 100% specificity, 100% positive predictive value (PPV), and 96% negative predictive value (NPV) are obtained.

The Pediatric Appendicitis Score (PAS) is composed of 8 components with a total score of 10 (Table 2). A score of 1 to 3 is considered negative for appendicitis, whereas scores from 8 to 10 are considered positive. The intermediate scores of 4 to 7 require further diagnostic testing. With these thresholds, the sensitivity for appendicitis is 97% with a specificity of 97.6%, a PPV of 97.2%, and an NPV of 97.6%. Although both of these systems have been shown to be useful in the diagnosis of appendicitis, they do not replace an experienced clinician.

**Differential Diagnosis**

The differential diagnosis is varied, but is divided into 5 general categories: inflammatory, infectious, vascular, congenital, and genitourinary conditions. Inflammatory mimickers of appendicitis include mesenteric adenitis (primary or secondary), inflammatory bowel disease, intussusception, omental infarction, or epiploic appendagitis. When CT (computed tomography) scans are performed for right lower quadrant pain, mesenteric adenitis is the most common alternative diagnosis, followed closely by inflammatory bowel diseases. Infectious causes include viral infections, bacterial infections, and parasitic infections. Among vascular causes, Henoch-Schonlein purpura can initially

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<td>Migration of pain</td>
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<tr>
<td>Anorexia</td>
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<tr>
<td>Nausea/vomiting</td>
<td>1</td>
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<tr>
<td>Right lower quadrant tenderness</td>
<td>2</td>
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<tr>
<td>Rebound pain</td>
<td>1</td>
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<td>Increase in temperature (&gt;37.3°C)</td>
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<td>Leukocytosis (&gt;10,000/µL)</td>
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<td>Polymorphonuclear neutrophilia (&gt;75%)</td>
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present as severe abdominal pain before the characteristic purpuric rash appears. Congenital causes include Meckel diverticulum, Meckel diverticulitis, and duplication cysts. Genitourinary causes include pyelonephritis, nephrolithiasis, ovarian torsion, ovarian tumors, hemorrhagic ovarian cysts, pelvic inflammatory disease, and infected urachal remnants. Constipation cannot be forgotten when evaluating pediatric patients because it is often a culprit in abdominal pain.

**Imaging**

**Radiograph** Radiograph imaging has little usefulness in confirming straightforward appendicitis and should not be performed unless considering alternative diagnoses, such as constipation. Occasionally an appendicolith can be seen on abdominal film, although, without symptoms, it is not an indication for appendectomy.

**Ultrasound** Ultrasound (US) has become an increasingly sophisticated diagnostic modality in the last 20 years with the added bonus of no radiation exposure. US has also allowed improved selection of patients requiring surgery and/or admission from the emergency department, as well as decreasing recurrent emergency room referrals. Multiple studies have focused on the ability of US to be used as an adjunct in the diagnosis of appendicitis in the adult population as well as in the pediatric population. Sonographic criteria for appendicitis include a blind-ending tubular structure with a diameter greater than 6 mm; a wall thicker than 2 mm; or an irregular wall that is rigid, noncompressible, and lacks peristalsis. Other signs that may suggest appendicitis include absence of air in the appendiceal lumen, periappendiceal fat changes, visible appendicolith, complex mass, mesenteric lymph nodes, and free fluid. A wide range of sensitivities and specificities can be found in the literature. Goldin and colleagues showed that increasing the parameters of the diameter to 7 mm and the thickness to 1.7 mm improved the sensitivity to 98.7% and the specificity to 95.4%. Further studies have also shown that surgeon-performed US with clinical evaluation may yield similar accuracy as radiologist-performed US.

One concern regarding US surrounds its use in obese patients. Although some studies state that there is no difference based on weight within the pediatric population, several studies have shown decreased visualization of the appendix with

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<td>Anorexia</td>
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<td>1</td>
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<td>Right lower quadrant tenderness</td>
<td>2</td>
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<tr>
<td>Cough/hopping/percussion</td>
<td>2</td>
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<tr>
<td>tenderness in right lower quadrant</td>
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<tr>
<td>Increase in temperature</td>
<td>1</td>
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<td>Leukocytes &gt;10,000/μL</td>
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<tr>
<td>Polymorphonuclear neutrophilia &gt;75%</td>
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<td>Total</td>
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increased obesity or abdominal wall thickness.\textsuperscript{12,13} Other studies have revealed an increased need for CT to definitively diagnose appendicitis in obese pediatric patients.\textsuperscript{14} Because of the conflicting data on this subject, no firm answer can be provided currently and further prospective studies are needed for evaluation.

**CT** Two types of CT scans can be performed when evaluating for appendicitis. The first protocol consists of oral and intravenous contrast, whereas the second uses rectal and intravenous contrast. Both methods use criteria including nonfilling appendix, appendicolith, fat stranding in the right lower quadrant, appendix diameter greater than 6 mm, appendiceal wall thickening, or arrowhead sign. The sensitivity of CT scan for appendicitis is 97\%, with a specificity of 99\%, PPV of 98\%, and NPV of 98\%. The accuracy of CT scan for diagnosing acute appendicitis is 96\%.\textsuperscript{15,16} Perforated appendicitis is suggested by appendicolith with intraluminal appendiceal air, extraluminal air, bowel wall thickening, ileal wall enhancement, extraluminal appendicolith, abscess, phlegmon, periappendiceal inflammatory stranding, and free fluid. The accuracy of CT scan for perforation is 72\%, with a sensitivity of 62\% and specificity of 82\%. Both the standard prep of Gastrografin and Volumen (1-hour prep) have been shown to be equally efficacious in visualization of the appendix.\textsuperscript{17} Nonvisualization of the appendix has been shown to have a high NPV (98.7\%).\textsuperscript{18}

**US versus CT** Current controversy seems to center around the use of CT versus US. US has the advantage of being low-cost without exposure to dyes and radiation, as well as providing dynamic information. However, it has significant disadvantages including being highly operator dependent and being limited by patient body habitus as well as appendiceal location. In the pediatric population, the sensitivity of US ranges from 78\% to 94\% and the specificity ranges from 89\% to 98\%. Accuracy has been reported between 89\% and 98\%.\textsuperscript{1} Another concern regarding US is the potential lack of availability during the night shifts, when CT scan use has been shown to increase.\textsuperscript{19} CT scans are not affected by the patient’s size, the position of appendix, or availability of experienced technicians.\textsuperscript{20} CT scans also have a higher sensitivity (95\%–99\%) and specificity (83\%–100\%) throughout the literature. Disadvantages include a prolonged preparation time as well as exposure to both intravenous dyes and radiation. One CT scan of the abdomen in a 5-year-old child increases the lifetime risk of radiation-induced cancer to 26.1 per 100,000 in women and 20.1 per 100,000 in men.\textsuperscript{21}

Most of the literature now focuses on a combination of analytical models such as the PAS or Alvarado score with sequential imaging. Most centers first perform an US in the intermediate groups, followed by CT scan or serial examinations if the US is nondiagnostic. This protocol has resulted in a high sensitivity (96\%–99\%) and specificity (83\%–92\%) for the diagnosis of appendicitis.\textsuperscript{22} These pathways have also shown an improvement in the overall hospital costs, as well as the incremental cost-effectiveness ratios.

When evaluating a patient for appendicitis, the importance of clinical judgment cannot be overstated. Although at many centers there is an increased reliance on CT scans, studies have clearly shown that the diagnosis of appendicitis can be made with history, examination, and selective use of US. Williams and colleagues\textsuperscript{23} showed that a pediatric surgeon can differentiate appendicitis from other abdominal disorders with 92\% accuracy. Our center’s experience and the literature confirm that patients with suspected appendicitis can be successfully evaluated without the use of CT scan in most patients.
Treatment

Antibiotics

For more than 30 years, pediatric surgeons used a triple-antibiotic regimen when dealing with appendicitis, consisting of ampicillin, gentamicin, and clindamycin. With the changes in the adult antibiotic regimens, pediatric surgery has evolved from this triple-antibiotic regimen to a simpler single-drug regimen. Bacteriologic epidemiology of appendicitis shows that the most commonly isolated organisms are *Escherichia coli*, *Streptococcus group milleri*, anaerobes, and *Pseudomonas aeruginosa*. Both piperacillin/tazobactam and cefoxitin have been shown to be at least as efficacious as the triple-drug regimen, and may also decrease length of stay and pharmaceutical costs.\(^\text{24}\) Other studies suggest that metronidazole must be added to a third-generation cephalosporin to cover anaerobic isolates.\(^\text{25,26}\)

The length of treatment is determined by presence or lack of perforation. In general, broad-spectrum coverage is recommended before operation. Our center prefers a single dose of cefoxitin (40 mg/kg). In simple appendicitis, the treatment of pediatric patients mirrors that of adults and consists of a single perioperative dose of antibiotics. With perforated appendicitis, at least a 5-day course of broad-spectrum antibiotics, such as piperacillin/tazobactam, is recommended. However, similar results were achieved using a 7-day course that initiated with intravenous antibiotics and finished with oral antibiotics. Total length of antibiotic therapy should be determined by the clinical condition of the patient, including resolution of fever, pain, bowel function, and WBC count.\(^\text{27}\)

Nonoperative management of acute appendicitis

Limited data exist from Europe regarding the management of acute appendicitis with antibiotics alone. One study from Turkey selected outpatients with less than 24 hours of pain and treated them with parenteral antibiotics (ampicillin/sulbactam) with resolution of symptoms within all 16 patients.\(^\text{28}\) The experience with nonoperative management in this fashion is limited and it is not recommended.

Surgical options for acute appendicitis

Incidental appendectomy

Incidental appendectomy is not advocated except in specific situations, including any surgery that has a right lower quadrant incision such as Meckel diverticulectomy or intussusception reduction. Occasionally a patient is found to have appendicolith while being evaluated for other disorders and this is not, in itself, an indication for appendectomy. Incidental appendectomy has not shown any benefit and usually should not be pursued, particularly if the procedure will be converted from a clean to a clean-contaminated case.

Delayed appendectomy

Although traditional teaching was that appendectomy must be performed emergently, recent studies have challenged this belief. When comparing emergent appendectomy (within 5 hours of admission) to urgent appendectomy (within 17 hours), it has been shown that there is no difference in gangrenous/perforated appendices, operative length, readmission, postoperative complications, hospital stay, or hospital charges.\(^\text{29}\) Most centers think that appendectomy can safely be delayed until morning in patients presenting at night, although it is recommended to start broad-spectrum antibiotics during the delay.

Open appendectomy

Traditional appendectomy was first described by McBurney\(^\text{30}\) and is still used for both acute and perforated appendicitis, although our center has not performed one in more than 10 years. Most surgeons use a transverse or oblique right lower quadrant incision. Dissection is carried down to the muscle, which is split.
The mesoappendix is divided, followed by excision of the appendix at its base. Management of the appendiceal stump can involve simple ligation, ligation and inversion using a purse-string, or pure inversion without ligature. The method used is a matter of physician preference.

**Three-port laparoscopic appendectomy** Traditional laparoscopic appendectomies involve one 10-mm to 12-mm port and two 5-mm ports. Traditionally, the larger port is placed at the umbilicus, although variations exist. The two 5-mm port sites can be placed in the left lower quadrant and in the suprapubic midline, with care taken to avoid injury of the bladder. Once ports are in place, the camera is usually placed in the left lower quadrant port. Laparoscopy allows visualization of the entire abdomen, which provides a significant advantage compared with open appendectomies.

Regarding division of the mesoappendix, several methods are available. Studies have shown than division of the mesoappendix in pediatric patients with electrocautery is safe and cost-effective. Some physicians prefer a US-activated scalpel (Harmonic) for division of the mesoappendix. The advantages include decreased exchanges of instruments as well as preventing theoretic current transmission. With either of these methods, an endoloop can be used to effectively ligate the appendiceal stump, which provides further cost saving measures compared with an endostapling device. Alternatively, endostapling devices can be used to divide both the mesoappendix and the appendix.

**Transumbilical laparoscopic appendectomy** Transumbilical laparoscopic appendectomy provides a middle ground of open and laparoscopic appendectomies. The procedure starts with the placement of a 12-mm umbilical trocar site, through which a working laparoscopic camera with single 5-mm instrument port is inserted. Once the appendix has been grasped, it is withdrawn through the umbilical incision (Fig. 1). A traditional appendectomy with division of the mesoappendix and excision of the appendix can then be performed. Multiple studies have shown that transumbilical laparoscopic appendectomies are safe with similar complication rates to traditional laparoscopic appendectomies in both simple and complicated appendicitis. They also provide the benefit of scarless surgery.

**Single-incision laparoscopic appendectomy** Single-incision laparoscopic appendectomies are performed using a 12-mm incision at the umbilicus. Multiple devices

Fig. 1. Transumbilical laparoscopic appendectomy after delivery of the appendix through the umbilical incision. The appendix and mesoappendix may be resected at this point using traditional techniques.
have been developed to provide multiple ports through this single-incision including the SILS port (Covidien, Norwalk, CT, USA) and the TriPort Access System (Advanced Surgical Concepts, Wicklow, Ireland). Dissection and transection of the appendix and mesoappendix can proceed similarly to a traditional laparoscopic appendectomy. Safety and efficacy are noted to be the same as with other methods, but operative times are slightly longer, most likely secondary to the learning process.34

**Laparoscopic versus open appendectomy**

When dealing with either simple or perforated appendicitis, the laparoscopic approach has been found to be safe and efficacious. Overall, laparoscopy leads to decreased length of stay and decreased time to oral intake. Length of stay decreases from an average of 3 days to 2 days for simple appendicitis and 7 to 5 days for complicated appendicitis.35 In many centers, simple appendectomies can lead to discharge within 24 hours. Complication rates are lower overall for laparoscopic appendectomies, although the postoperative intra-abdominal abscess rate is higher with the laparoscopic approach.36 For this reason, laparoscopic appendectomy is thought to be a safe option when dealing with appendicitis in any stage of disease.

**Nonoperative versus operative management of perforated appendicitis**

Complicated or perforated appendicitis remains an area of significant debate. As discussed earlier, laparoscopic appendectomy can safely be performed when dealing with perforated appendicitis. When dealing with complicated perforated appendicitis with abscess or phlegmon, the treatment can either be antibiotics with or without immediate drainage or surgical intervention. Nonoperative management is most commonly used in patients with symptoms for more than 3 days, absence of diffuse peritonitis, absence of obstruction, and mass on imaging or examination. These patients are placed on broad-spectrum antibiotics (Zosyn, triple-antibiotic therapy, gentamicin, and clindamycin) can undergo drainage of the fluid collections greater than 2 cm in size.37 Drainage can be performed through US or CT guidance and may be performed via a transabdominal, transgluteal, or transrectal approach. The transvaginal approach is not recommended in pediatric patients unless the patient is a teenager who is postpartum or admits to sexual activity.

A nonoperative approach to appendicitis is traditionally followed by an interval appendectomy. The need for interval appendectomy has been called into question recently. Retrospective studies have shown that up to 80% of children may not require appendectomy and that 3% of patients suffer a complication secondary to interval appendectomy.38 More recent prospective studies have shown a recurrence rate of 8% to 43%, with an increased rate of reoccurrence among patients with appendicolith.39,40 This finding suggests that the morbidity of surgery may be avoided by selective surgery in patients with certain criteria, such as the presence of appendicolith. Another issue under debate is whether these patients with complicated appendicitis should have conservative management or undergo initial operative management. Recent studies have shown improvement in return to normal activity as well as decreased complications when early appendectomy is performed.41 At our center, we practice selective early intervention depending on patient condition and size or presence of fluid collection.

**Postoperative Care**

Postoperative care depends largely on a patient’s intraoperative findings. With simple appendicitis, early oral intake and discharge within 24 hours is increasingly seen. With laparoscopic approaches, patients are frequently discharged from the
recovery room. Perforated appendicitis requires postoperative antibiotics and more care should be taken with advancement of diet. Routine nasogastric tube placement with perforated appendicitis has been abandoned and should only be performed in cases of ileus or obstruction. Many patients with simple perforated appendicitis tolerate advancement of diet at a similar rate as with nonperforated appendicitis. However, patients with complicated appendicitis often suffer significant ileus. Antibiotic therapy should be continued until the patient’s clinical condition improves, as seen by resolution of ileus, abdominal pain, normalization of WBC count, and lack of fever.

Complications

Postoperative complications include wound infection, deep-space infections, bowel obstruction, and stump appendicitis. Different measures have been advocated to prevent wound infections, including use of wound protectors in perforated appendicitis and Endobag. No benefit has been seen with interrupted closure of wounds or with keeping wounds open. To decrease intra-abdominal abscess formation, many surgeons use peritoneal lavage with saline or sterile water. Drain placement has largely been abandoned.

SUMMARY

1. Appendicitis most frequently occurs between the ages of 10 to 11 years.
2. The classic signs and symptoms of appendicitis occur in less than half of pediatric patients. The most sensitive symptoms include migrating pain to the right lower quadrant and fever. Rebound tenderness on examination also increases the likelihood of appendicitis.
3. Increased WBC count and left shift are the most accurate laboratory values when assessing for appendicitis.
4. Clinical judgment and judicious studies are the best methods when assessing for appendicitis. Scoring systems such as the Alvarado score and PAS have also been shown to be useful in diagnosis.
5. US is the imaging study of choice and CT scans should be avoided in pediatric patients because of radiation exposure.
6. Although both open and laparoscopic approaches have been shown to be safe and effective with simple or complex appendicitis, laparoscopic appendectomies are recommended.
7. The treatment of complex appendicitis with mass or abscess is still controversial, with support for both operative and conservative management. With nonoperative management, controversy also exists regarding interval appendectomy.

INTUSSUSCEPTION

The invagination of a proximal intestinal segment into a distal section of bowel is referred to as intussusception. The portion of bowel that invaginates into the distal bowel is referred to as the intussusceptum, whereas the distal segment is referred to as intussuscipiens (Fig. 2). Intussusception is one of the most frequent causes of bowel obstruction in the pediatric population. Significant complications including bowel necrosis, perforation, and death can occur if there is a delay in diagnosis. For this reason, there is significant onus on clinicians to accurately and promptly diagnose intussusception.
Diagnosis

Demographics
Although intussusception can occur at any age, it most frequently occurs before the age of 2 years, with peak incidence between 5 and 10 months.\textsuperscript{43} The incidence is reported as 56 children per 100,000 per year. The ratio of males to females is 2:1. There is also a seasonal variation, with increasing incidence in the fall and winter months.

Classification and pathogenesis
Intussusception is classified by anatomic location or by pathogenesis. Most commonly, intussusceptions are ileocolic, but can also be ileoileal, colocolic, or located at other gastrointestinal tract locations. Intussusceptions can also be divided into idiopathic intussusception and those secondary to a pathologic lead point.

Idiopathic
As opposed to adult intussusception, intussusception amongst the pediatric population is usually idiopathic because more than 90% of pediatric intussusception is without pathologic lead point. A greater percentage of idiopathic cases occur among patients less than the age of 2 years. Many mechanisms have been proposed for the pathogenesis of idiopathic intussusception. Because of frequent association with current or recent upper respiratory infections or enteritis, viral or bacterial infections have been proposed as a cause. This theory seemed to be supported by both the seasonal variation of intussusception as well as the increased incidence associated with the tetravalent rhesus-human reassortment rotavirus vaccine (RotaShield; Wyeth-Lederle Vaccines, Philadelphia, PA). This vaccine was initially released in 1998, but was subsequently withdrawn from the market when there was shown to be a 20-fold increase in the risk of intussusception during the first 14 days following vaccine administration.\textsuperscript{44} Two further vaccines have been developed since that time: RotaTeq (a live oral human-bovine reassortment rotavirus vaccine; Merck and Co, Whitehouse Station, NJ, USA) and Rotarix (live, oral, human attenuated rotavirus vaccine; GlaxoSmithKline, Brentford, Middlesex, United Kingdom). Multiple studies have shown the safety and efficacy of these vaccinations without increased incidence of intussusception. Current recommendations from the American Academy of Pediatrics advise vaccination with either RotaTeq at ages 2, 4, and 6 months or with Rotarix at ages 2 and 4 months.\textsuperscript{45}

Secondary to lead point
Although intussusception secondary to a lead point is more often associated with the adult population, approximately 10% of pediatric patients also have a pathologic lead point.\textsuperscript{43} There are multiple possible lead points that are divided into anatomic (Meckel diverticulum, appendix, duplication cyst, heterotopic tissues), tumors (lipomas, lymphoma, ganglioneuroma, Kaposi sarcoma), genetic

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Fig. 2. The intussusceptum (dark arrow) invaginates into the intussuscipiens (outlined arrow). In ileocecal intussusception, the ileum is the intussusceptum and the cecum is the intussuscipiens.
hamartomas secondary to Peutz-Jeghers syndrome, cystic fibrosis), vascular (Henoch-Schonlein purpura, hemorrhagic edema, blue rubber bleb nevus syndrome), infectious (pseudomembranous colitis, bacterial), traumatic (secondary to hematoma or dysmotility), secondary to a foreign body (enterostomy tube), and postsurgical lead points. After the age of 3 years, the incidence of pathologic lead point increases with the most common pathologies including Meckel diverticulum, lymphoma, and polyps.

Pathologic lead points are often suggested by an irreducible or recurrent intussusception. When a patient fails enema reduction of the intussusception, care must be taken during surgery to assess the bowel for disorders and treat appropriately, if found. Some studies suggest that recurrent intussusception should increase the clinical suspicion for a lead point, prompting further investigation by radiologic means or by laparoscopy. The appropriate number of repeated nonoperative attempts is still subject to debate.

Patients with lead points secondary to Burkitt lymphoma require special consideration. These children present at a higher median age (10 years) than idiopathic intussusception and are almost always ileocolic intussusceptions. These intussusceptions are usually irreducible by enema techniques. Up to 70% present with stage II disease (primary gastrointestinal tract tumor with or without involvement of associated mesenteric nodes) and can undergo curative resection. These patients benefit from a decreased duration and intensity of chemotherapy, along with an increased survival. Surgeons must therefore be alert for this possibility when operating on irreducible intussusception and thoroughly explore all peritoneal surfaces. Consideration should also be given to collection of ascitic fluid and examination of liver, spleen, and retroperitoneal nodes.

Peutz-Jeghers syndrome (PJS) is an autosomal dominant inherited disorder characterized by gastrointestinal hamartomas, mucocutaneous pigmentation, and an increased cancer risk. It requires special attention secondary to the hamartomas that occur in more than 90% of patients with PJS. These hamartomas occur predominantly in the jejunum, and carry up to a 69% risk of intussusception. Hamartomas greater than 15 mm in diameter have been shown to significantly increase the risk of intussusception and some centers recommend polypectomy at greater than or 10 mm diameter. Surveillance for hamartomas can be achieved through a combination of CT enteroclysis, small bowel follow-through, magnetic resonance enteroclysis, video-capsule endoscopy, and endoscopy with balloon-assisted enteroscopy (BAE). Endoscopy with BAE also allows polypectomy.

**Symptoms and signs**
The classic symptoms of intussusception include abdominal pain, vomiting, and currant jelly stool. Patients experience episodic abdominal pain and present with calmness interspersed with fussiness. Other less common symptoms include fever, diarrhea, and constipation. The patient’s examination may range from completely benign to tenderness in the right lower quadrant, or even frank peritonitis in a patient with perforation. Some patients may have a tubular, sausage-shaped mass on palpation of the abdomen and/or guaiac-positive stool. However, history and physical examination are unreliable in the diagnosis of intussusception and fewer than 25% to 50% of patients have the classic triad of symptoms.

**Laboratory studies**
Although it is useful to obtain a CBC and basic metabolic panel when evaluating a patient with abdominal pain, there are no consistent findings among patients with
intussusception. However, increased white blood count or bandemia should alert a clinician to the possibility of perforation or gangrenous bowel.

**Imaging**

**Abdominal radiograph** Findings consistent with intussusception on plain film include presence of small bowel obstruction, paucity of right lower quadrant gas, presence of an intracolonic mass, and presence of a rim sign. Some studies suggest that plain films are useful only to detect pneumoperitoneum or for the evaluation of other pathologic conditions. Other studies suggest that, in combination with an accurate assessment of the clinical scenario, 2-view or 3-view films of the abdomen can yield similar results to US for sensitivity and specificity. These studies generally require the availability of an experienced pediatric surgeon or pediatric radiologist to achieve these results. Most centers prefer US for confirmation of diagnosis and only use plain films as an adjunct to diagnosis.

**US** US was first identified as a promising diagnostic modality for intussusception in the 1980s, and has become the primary modality for diagnosis at many centers. Although US is highly operator dependent, skilled technicians can achieve a sensitivity of 97.9% with a specificity of 97.8%. Findings that indicate intussusception include a target or bull’s-eye lesion with concentric echogenic layers. Findings that suggest a decreased likelihood of enema reduction include trapped peritoneal fluid, absence of blood flow, enlarged lymph nodes, and intramural gas. US also allows measurement of length and width of these lesions, aiding in differentiation of small bowel intussusceptions from ileocolic intussusception. Small bowel intussusceptions have a mean diameter of 1.5 cm with a length of 2.5 cm. Ileocolic intussusceptions have a mean diameter of 3.7 cm and a mean length of 8.2 cm. This differentiation is thought to be important because small bowel intussusceptions are often transient and some centers choose to monitor these patients.

**Fluoroscopy** Although many centers prefer diagnosis with US before fluoroscopic studies, some centers progress straight from initial evaluation to diagnostic contrast enema. This progression is beneficial in initial cost to the patient, as well as in decreasing time to reduction. However, studies show that the lifetime cost-effectiveness increases because of radiation exposure and increased risk of radiation-induced malignancy from 59.7 cases per 100,000 to 79.3 cases per 100,000. For this reason, our center prefers an initial screening evaluation with US followed by therapeutic air enema when required.

**CT/magnetic resonance imaging** Although CT and magnetic resonance imaging (MRI) both have high sensitivity and specificity for intussusception, the use of these modalities is limited. CT scans increase the risk of radiation-induced malignancy. Although MRI is lacking in radiation exposure, it is not cost-effective or readily available in many center. As mentioned earlier, CT scans may be useful for identification of lead points in patients with multiple episodes of intussusception.

**Treatment**

**Antibiotics**

Before any treatment of intussusception, our center prefers to administer 1 dose of cefoxitin (40 mg/kg). Although perforation is rare, this provides antibiotic coverage should complications arise. In patients with gross peritonitis, coverage with broad-spectrum antibiotics is necessary and length of treatment should be determined once the degree of contamination is determined.
**Therapeutic enemas**

Therapeutic enemas have become the mainstay in treatment of intussusception. Contraindications to therapeutic enema reduction include evidence of peritonitis, perforation, or necrotic bowel. Both fluoroscopic and US-guided reduction have been advocated with the use of different solutions, including barium, Gastrografin, saline, or pneumatic reduction. Although some centers still use Gastrografin or barium, pneumatic reduction has been shown to have a higher rate of success (>90%) without risk of peritoneal exposure to contrast with perforation. This higher rate of reduction with pneumatic reduction may be secondary to the level of experience in those who use pneumatic reduction, as well as the higher intraluminal pressure used with pneumatic reduction. Although Gastrografin is favored more than barium by some because of its water-soluble state and because it does not hold stool in suspension, which causes intra-abdominal abscesses on perforation, it is less favored than pneumatic reduction secondary to the osmotic shifts of fluid and electrolytes that occur with perforation.\(^58\) Fluoroscopic reduction provides visualization of reduction in most cases, but does expose the patient to radiation. Although classically the fluid or air must be visualized refluxing into the ileum, some studies have shown that nonoperative management may be used in patients without reflux into the ileum if there is symptom resolution.\(^59\) US-guided reduction is limited by the operator’s experience.

Therapeutic enemas can be performed even with delayed presentation with similar outcome in reduction.\(^60\) Some centers advocate repeated attempts at reduction before operative intervention if there is suspicion of partial reduction.\(^61\) Studies have also shown the benefit of repeating reduction at a tertiary care center after failed attempts at an outlying facility.\(^52\) This aggressive approach with therapeutic enemas is prompted by the vast improvement in patient outcome for length of stay and return to oral intake. For this reason, it is also advocated that recurrent intussusception be treated with repeat therapeutic enema.

**Operative intervention**

Indications for operative intervention include perforation, peritonitis, and failed nonoperative reduction of intussusception. Both open and laparoscopic approaches can be used for this procedure.

**Open** The open approach consists of performing a right lower quadrant incision similar to that used in appendectomy. Once the abdomen is entered, gentle pressure is placed on the intussuscipiens, gently milking it away from the intussusceptum. Pulling the 2 ends apart is classically avoided secondary to the friability of the bowel and possible perforation. It is also important to examine the bowel following reduction to rule out a lead point as well as to determine the viability of the bowel. If there is ischemia of the bowel, resection is necessary. Most surgeons also perform an appendectomy at the time of reduction because of the location of the incision.

**Laparoscopic** Laparoscopic reduction of intussusception can either be performed using a single-trocar or 3-trocar approach. Although traditionally avoided in open approach, laparoscopic reduction has shown that gentle tension on the intussusceptum while applying gentle pressure to the edge of the intussuscipiens is safe and effective (Fig. 3). Although some centers previously advocated ileopexy, it has not been shown to be beneficial in reducing recurrences.\(^63\) If there is failure of reduction using this technique, bowel resection can proceed transumbilically or via conversion to a right lower quadrant incision. Although initial success rates with laparoscopic reduction were poor, many centers now achieve approximately an 85% success rates with reduction.\(^64\) Laparoscopic reduction has a small but significant decrease in hospital
stay, as well as in time to diet resumption. Whether this is of clinical significance remains controversial.

**Postreduction Management**

Most patients who undergo enema reduction can be started immediately on a liquid diet and advanced as tolerated. Some centers advocate reduction and immediate discharge to home. When patients undergo surgical reduction, most can also be rapidly advanced. However, many require a more gradual advancement to general diet. Judgment must be used in patients with significant edema of the bowel and after resection of the bowels.

**SUMMARY**

1. A patient presenting with intussusception is usually less than 2 years old. The classic triad of abdominal pain, emesis, and bloody stool is only present in 25% of the population.
2. In older patients (>3 years) with irreducible intussusception or recurrent intussusception, thought should be given to a possible pathologic lead point.
3. After history and physical examination, most patients should undergo US for diagnosis.
4. Therapeutic enema is currently the initial treatment of choice for intussusception and can be used in delayed presentation, recurrent intussusception, and in transferred patients who have had failed reduction attempts at outlying facilities.
5. Patients should be taken to the operating room for gross peritonitis, perforation, or failure of nonoperative reduction techniques. Laparoscopic reduction has been shown to be safe and effective, with reduction in length of stay and return to oral intake.

**MECKEL DIVERTICULUM**

Meckel diverticulum is the most common anomaly of the gastrointestinal tract with prevalence from 1% to 4%. Presentation can include gastrointestinal bleed, obstruction, diverticulitis, perforation, and volvulus. Because of this wide range of clinical scenarios, it is important for a clinician to have a high index of suspicion to prevent significant complications.

**Embryology**

Meckel diverticula arise through a failure of the omphalomesenteric duct to involute. The omphalomesenteric duct is a connection between the yolk sac and the primitive...
gut and typically recedes between the fifth and ninth week of gestation. Failure of this obliteration can result in several anomalies including a persistent omphalomesenteric fistula, umbilical cyst, vitelline duct remnant, fibrous bands from the umbilicus to the small bowel, mesodiverticular bands, and Meckel diverticula.\(^{67}\) Because of the nature of formation, Meckel diverticula are true diverticula involving all layers of the bowel.

**Diagnosis**

**Epidemiology**

Meckel diverticulum is often referred to by the rule of twos\(^{68}\):
- Occurs in 2% of the population (1%–4%)
- Has a 2:1 male/female ratio
- Located within 2 ft (60 cm) of the ileocecal valve on the antimesenteric border
- Commonly 2 cm in diameter
- Commonly 2 in (5 cm) in length
- Can contain 2 types of ectopic tissue (pancreatic and gastric)
- More common before 2 years of age.

**Signs, symptoms, and differential**

Meckel diverticulum can present with a wide range of clinical scenarios and can pose a diagnostic challenge. The most common presentations of Meckel diverticula are obstruction and gastrointestinal bleed.

**Gastrointestinal bleed** Heterotopic gastric tissue can be present in up to 50% of symptomatic Meckel diverticula.\(^{69}\) Because of this active tissue, ulcerations can form at the edge of diverticulum or in the adjoining ileum. These patients present with bloody stool, fatigue, irritability, and abdominal pain. Physical examination is often unremarkable. The bleeding can occur intermittently, causing significant delay in diagnosis.\(^{70}\) The differential diagnosis includes infectious causes (*Clostridium difficile*, *E. coli*), angiodysplasia, malignancy, or upper gastrointestinal bleed.

**Obstruction** Some patients present with an obstructive picture with symptoms including abdominal pain, distention, nausea, and vomiting. Physical examination can include distention, tenderness with palpation, hypoactive bowel sounds, mass, or even peritonitis. Obstruction is often secondary to intussusception of the Meckel into the ileum; however, some Meckel diverticula are attached to the umbilicus by bands of tissue that can lead to internal hernia or volvulus.\(^{71,72}\) Differential diagnosis includes intussusception, obstruction secondary to adhesions, ileus, gastroenteritis, or tumor with mass effect.

**Diverticulitis/perforation** In some patients, the symptoms that lead to diagnosis of a Meckel diverticulum are similar to those of appendicitis, and can include fever, right lower quadrant abdominal pain, nausea, and vomiting. Physical examination may be indistinguishable from appendicitis, with tenderness to palpation, guarding, and rebound tenderness. The mechanism is thought to be similar to diverticulitis in the colon with obstruction of the lumen leading to inflammation and eventual perforation.\(^{73}\) The differential diagnosis includes appendicitis, gastroenteritis, mesenteric adenitis, and gynecologic issue (eg, ovarian torsion, pelvic inflammatory disease). When encountering signs of inflammation intra-abdominally with a normal appendix, an exploration for Meckel diverticulitis should be performed.
Other Presentations

A patent vitelline duct presents as drainage from the umbilicus. Littre hernia is an inguinal hernia containing the Meckel diverticulum and is indistinguishable from any other herniation until operation. Other less common presentations include tumors that form in the Meckel diverticulum. Many of these present at late stages.

Laboratories

The laboratory testing that should be undertaken with a Meckel diverticulum depends on the presentation. Basic metabolic panel and CBC are a starting point, allowing evaluation for dehydration and anemia. In the case of diverticulitis and/or perforation, a WBC count is also important.

Imaging

Abdominal radiographs

Plain films are of limited use in diagnosing a Meckel diverticulum, but can be used to evaluate for pneumoperitoneum and obstruction.

Contrast studies

Both upper gastrointestinal series and enteroclysis have been used to detect Meckel diverticulum. However, these tests have a low sensitivity and are difficult to interpret. The classic finding is a single diverticulum arising from the antimesenteric border of the distal ileum. In instances of intussusception, these studies may also reveal a filling defect with possible obstruction.\(^\text{74}\)

US

US is often used in evaluation of pediatric patients with abdominal pain. Just as Meckel diverticulitis mimics appendicitis clinically, it may also do so on US with the appearance of a long tubular structure with thickened walls. A Meckel diverticulum may also appear similar to a duplication cyst. Intussusception of a Meckel diverticulum may appear as a double-target sign with a targetlike mass with a central area of hyperechogenicity.\(^\text{75}\)

CT/MRI

Both CT and MRI are of limited diagnostic value when evaluating for Meckel diverticulum. They may reveal nonspecific signs such as inflammation, calcifications, obstruction, pneumoperitoneum, and free fluid. CT and MRI enteroclysis have been shown to be more effective when identifying Meckel disorders. However, both tests are expensive, with the added risk of contrast and radiation exposure, and should not be used in the diagnosis of Meckel diverticulum.\(^\text{76,77}\)

Nuclear medicine

For a bleeding diverticulum, the current test of choice is a technetium-99 pertechnetate scan (Meckel scan). Studies have revealed approximately 65% to 85% sensitivity in the pediatric population. This sensitivity can be increased using an H2 antagonist, pentagastrin, and glucagon. However, the NPV is only 74%. Different reasons have been proposed regarding this low value, including rapid loss of blood leading to loss of the tracer, previous inflammation, and postsurgical changes.\(^\text{78}\)

Other tests

Mesenteric angiography and tagged red blood cell scanning may also be used to evaluate for Meckel diverticulum. These imaging studies allow the identification of bleeding, but require a 0.1 to 0.5 mL per hour blood loss for tagged red blood cell scans or a 1 mL per hour blood loss for mesenteric angiography. Both can be useful
to localize the site of bleeding to a Meckel diverticulum that has not been identified on nuclear medicine imaging. Using mesenteric angiography, a persistent vitellointestinal artery may be shown in individuals who are no longer actively bleeding. Endoscopy, both upper and lower, may be used to evaluate for other disorders, but are not directly useful in identifying Meckel diverticula. Capsule endoscopy has been useful in identifying a Meckel diverticulum in several cases, but is not regularly used for this purpose. Although the studies evaluating these methodologies have low power and have not assessed for specificity or sensitivity, these procedures may be used when all other tests have failed to identify the source of occult bleeding.

**Treatment**

**Incidental diverticulum**

There continues to be significant debate surrounding whether an asymptomatic Meckel diverticulum should be excised. In the adult population, most literature agrees that excision is not required unless there is a palpable mass raising concern for cancer. In the pediatric population, the picture is less clear. Many studies conclude that the risk of serious complication exceeds that of the operative risk in children less than 8 years old. A recent study from the University of Pittsburgh urges resection of Meckel diverticulum at all ages because of the risk of cancer as well as the late stage of cancer presentation in most patients. Another study evaluated the increased morbidity with incidental Meckel diverticulum excision compared with the lifetime risk of complication. They concluded that the risk of excision was not worth the increased rate of complications. Because of this debate within the literature, the excision of incidentally noted Meckel diverticula is still highly controversial.

**Symptomatic diverticulum**

The treatment of symptomatic diverticulum is resection performed either laparoscopically or open. In a recent survey, more than 75% of resections were still performed open. The traditional open approach is performed through a right lower quadrant incision similarly to an appendectomy. For this reason, most surgeons perform an appendectomy at the same time as diverticulectomy.

Laparoscopic diverticulectomies can be performed via a transumbilical approach or through a 3-port laparoscopic approach similarly to laparoscopic appendectomy. Many studies have shown that a laparoscopic approach is as safe and effective as the open approach. They have also shown a decreased time to oral intake and to discharge. The disadvantage of a total laparoscopic approach is the loss of tactile sensation, preventing palpation of the base of the Meckel diverticulum to ensure complete resection and lack of masses. For this reason, some surgeons prefer a transumbilical approach, which provides the cosmetic and postoperative advantages of laparoscopic surgery but also enables a surgeon to palpate the specimen. For bleeding diverticula, some centers have reported isolated resection of the diverticula. However, our center still prefers resection of the diverticula as well as the adjoining segment of ileum because of the presence of ulceration in the small bowel.

**SUMMARY**

1. A Meckel diverticulum is an embryologic remnant of the omphalomesenteric duct.
2. Rule of twos: 2% of the population, within 2 ft (60 cm) of the ileocecal valve, 2:1 predominance in males/females, before 2 years of age, and 2 types of tissue (gastric and pancreatic).
3. Meckel diverticula can present in multiple ways, but most commonly present with obstruction or gastrointestinal bleeding.
4. Diagnostic testing depends on the presentation.
5. Resection of asymptomatic diverticula remains controversial.
6. Resection can be performed safely either open or laparoscopically. We recommend a laparoscopic approach for its benefits for postoperative recovery.

REFERENCES


