INTRODUCTION — Intussusception, the invagination of a part of the intestine into itself, is the most common abdominal emergency in early childhood, particularly in children younger than 2 years of age, and the second most common cause of intestinal obstruction after pyloric stenosis [1]. Intussusception is unusual in adults, and the diagnosis is commonly overlooked. In the majority of cases in adults, a pathologic cause is identified [2]. In contrast, the majority of cases in children are idiopathic.

EPIDEMIOLOGY — Intussusception is the most common cause of intestinal obstruction in infants between 6 and 36 months of age. Approximately 60 percent of children are younger than 1 year old, and 80 percent are younger than 2. Intussusception is rare before 3 months and after 6 years of age. In a population-wide survey in Switzerland, the yearly mean incidence of intussusception was 38, 31, and 26 cases per 100,000 live births in the first, second, and third year of life, respectively, then fell to less than half that rate in older age groups [3]. Most episodes occur in otherwise healthy and well-nourished children.

Intussusception appears to have a slight male predominance, with a male:female ratio of approximately 3:2.

PATHOGENESIS — Intussusception occurs most often near the ileocecal junction (ileo-colic), although ileo-ileo-colic, jejuno-jejunal, jejuno-ileal, or colo-colic intussusception also have been described. The proximal segment of bowel telescopes into the distal segment, dragging the associated mesentery with it. This leads to the development of venous and lymphatic congestion with resulting intestinal edema, which can ultimately lead to ischemia, perforation and peritonitis.

A lead point can be identified in up to 25% of children with ileo-colic intussusception. The remaining 75% are considered to be idiopathic, although an increasing body of evidence suggests that viral triggers may play a role in some cases:

- The incidence of intussusception has a seasonal variation, with peaks coinciding with seasonal viral gastroenteritis in some populations [3,4].
- Intussusception has been associated with some forms of rotavirus vaccine. An early form of the vaccine (RRV-TV: Rotashield™) was removed from the
market because of a 22-fold increase in intussusception among vaccinated infants. The vaccine currently used in the United States (PRV: Rotateq™) appears to have rates of intussusception that are similar to expected background rates. However, there may be additional cases of intussusception that have not been reported. Providers should be alert for cases of intussusception that may be associated with rotavirus vaccine, and report all suspected cases to the Vaccine Adverse event Reporting System (VAERS).

- Approximately 30% of patients experience viral illness (upper respiratory tract infection, otitis media, flu-like symptoms) before the onset of intussusception.
- A strong association with adenovirus infection has been shown in a variety of populations. In 30 to 40% of cases, there is evidence of recent infection with enteric and non-enteric species of adenovirus [5-9]. In a prospective case-control study examining a variety of potential infectious triggers for intussusception in Vietnam and Australia, infection with adenovirus, as the strongest predictor of intussusception in both populations [9].

Viral infections, including enteric adenovirus, can accentuate lymphatic tissue in the intestinal tract, resulting in hypertrophy of Peyer patches in the lymphoid-rich terminal ileum, which potentially acts as a lead point for ileocolic intussusception [5]. A role for corticosteroids in the prevention of recurrence has been proposed in light of this possible association with lymphoid hyperplasia [10,11].

A lead point is recognized more commonly in children older than five years [1,4,12]. A variety of pathologic conditions associated with intussusception, including small bowel lymphoma [13-15], Meckel diverticulum [16], duplication cysts [17,18], polyps [19], vascular malformations [20], inverted appendiceal stumps [21,22], parasites (eg, Ascaris lumbricoides) [23,24], Henoch-Schönlein purpura [25], and cystic fibrosis [26], have been described.

The mechanisms leading to intussusception differ depending upon the specific cause. As examples:
- Small bowel intussusception may occur after an episode of Henoch-Schönlein purpura, with a bowel wall hematoma acting as the lead point. In these cases, intussusception typically occurs after resolution of the abdominal pain.
- In patients with cystic fibrosis, thick inspissated stool may act as the lead point [26].
- Patients with celiac disease may develop small bowel intussusception secondary to dysmotility and excessive secretion or bowel wall weakness [27,28].
• Patients with Crohn disease may develop intussusception because of inflammation and stricture formation [29].

**Postoperative:** Bowel intussusception (usually jejuno-jejunal) also has been described in the postoperative setting where it is an uncommon but insidious cause of intestinal obstruction. It usually occurs within two weeks of a laparotomy and is caused by adhesions. The patient typically does well for several days and may even resume oral intake before developing symptoms of mechanical obstruction. The diagnosis can be difficult to establish because it may be confused with postoperative paralytic ileus. The ultrasonography and computed tomography (CT) scanning may be helpful. Diagnostic contrast studies are not as helpful in postoperative intussusception because most cases occur in the small intestine.

**CLINICAL MANIFESTATIONS:** Patients with intussusception typically develop the sudden onset of intermittent, severe, crampy, progressive abdominal pain, accompanied by inconsolable crying and drawing up of the legs toward the abdomen. The episodes usually occur at 15 to 20 minute intervals. They become more frequent and more severe over time. Vomiting may follow episodes of abdominal pain. Initially emesis is non-bilious, but it may become bilious as the obstruction progresses. Between the painful episodes, the child may behave relatively normally and be free of pain. As a result, initial symptoms can be confused with gastroenteritis [30]. As symptoms progress, increasing lethargy develops.

In up to 70% of cases, the stool contains gross or occult blood [31]. The stool may be a mixture of blood and mucous, giving it the appearance of currant jelly. A sausage-shaped abdominal mass may be felt in the right side of abdomen. However, the classically described triad of pain, a palpable sausage shaped abdominal mass, and currant-jelly stool is seen in less than 15 percent of patients at the time of presentation [30,32]. As many as 20% of young infants have no obvious pain. Approximately one-third of patients does not pass blood or mucus or develop an abdominal mass.

Occasionally, the initial presenting sign may be lethargy or altered consciousness alone, without pain, rectal bleeding, or other symptoms that suggest an intra-abdominal process [33-35]. In most cases, this clinical presentation occurred in infants, and was often confused with sepsis. Thus, intussusception should be considered in the evaluation of lethargy or altered consciousness, especially in infants.

**DIAGNOSIS AND NONSURGICAL TREATMENT**—A high index of suspicion coupled with early diagnosis
of intussusception may obviate the need for surgical intervention.

For patients in whom the diagnosis is unclear at presentation, initial workup may include abdominal ultrasound or abdominal plain films, provided that these studies do not significantly delay the definitive diagnosis and treatment of intussusception.

Patients with a typical presentation, in whom there is a high index of suspicion for intussusception, may proceed directly to an enema contrast study. These studies can establish the diagnosis of ileocolic intussusception, and frequently also are therapeutic. Before obtaining a contrast study, the patient should be stabilized and resuscitated with intravenous fluids; the stomach should be decompressed with a nasogastric tube. The patient should be considered fit for surgery, in case surgery is necessary, before he or she undergoes radiographic imaging. The surgical team should be notified before attempted reduction because there is a risk of perforation and because surgical intervention may be necessary if contrast enema fails to reduce the intussusception.

**Abdominal plain films** — Plain radiographs of the abdomen may be helpful because they may show frank intestinal obstruction or massively distended loops of bowel with absence of colonic gas. Although rarely seen, the presence of pneumoperitoneum suggests that bowel perforation has occurred. A target sign, consisting of two concentric radiolucent circles superimposed on the right kidney representing peritoneal fat surrounding and within the intussusception, appeared in 26% of 94 patients in one report [36]. A soft-tissue density projecting into the gas of the large bowel (intussusception image) is called the "crescent sign".

The presence of air in the ascending colon can help to exclude intussusception in cases with a low clinical suspicion of the disease. In a retrospective study from a single center, plain radiographs with three views (supine, lateral, and prone) were used to screen patients with suspected intussusception [37]. The presence of air in the ascending colon on at least two of these views had high sensitivity for excluding intussusception in this patient population with a low clinical suspicion of disease (sensitivity 96.3 percent, 95% CI 89.2-100). However, the sensitivity of plain radiographs may be considerably lower in different clinical settings or when fewer views are analyzed. Therefore, we do not recommend relying on plain radiography to exclude intussusception if there is a significant clinical suspicion of the disease.

**Ultrasonography** — Ultrasonography also can be useful; its sensitivity and specificity approach 100 percent in the
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hands of an experienced ultrasonographer [38-40]. The classic ultrasound image of intussusception is a "bull's eye" or "coiled spring" lesion representing layers of the intestine within the intestine. In addition, a lack of perfusion in the intussusceptum detected with color duplex imaging may indicate the development of ischemia. An advantage of ultrasonography is that it can diagnose the rare ileoileal intussusception and identify the lead point of intussusception in approximately two-thirds of cases in which underlying pathology exists [41].

Ultrasonography may also be useful in predicting which children are most likely to need surgical intervention for small bowel intussusception. In a small retrospective study, cases in which the intussusceptum was less than 3.5 cm were more likely to resolve spontaneously during or shortly after the ultrasonographic examination as compared to cases with a longer intussusceptum [42].

Reduction under ultrasonographic control without fluoroscopy has been described [43,44]. Signs of successful reduction include the disappearance of the intussusception and the retrograde flow of the water-soluble contrast into the small intestine. Ultrasound guided water enema also has been used [45]. Signs of successful reduction with water include the disappearance of the intussusception and the appearance of water and bubbles in the terminal ileum.

**Contrast studies** — The standard procedure for diagnosis and treatment of ileocolonic intussusception is a contrast (air or radiopaque) enema [46]. Broad-spectrum antibiotics may be administered before attempting nonoperative reduction since there is a risk of perforation with these procedures.

As compared with ileocolonic intussusception, small bowel (ileoileal, jeunoileal, or jejunoojejunal) intussusception is less likely to be reducible by contrast enema [47,48]. Intussusceptum length as measured by ultrasonography may be helpful in determining which patients may be managed expectantly, and which are likely to require surgical intervention.

**Barium and water-soluble contrast** — Traditionally, barium has been the preferred contrast agent in most North American and European centers [49-51]. Before instilling barium, a water-soluble contrast enema should be considered, particularly if there is a high risk of perforation. Water-soluble agents reduce the risk of electrolyte disturbances and peritonitis in patients in whom perforation has occurred.

Barium enema reduction of intussusception can be performed if there is no evidence of perforation. The
standard method of reduction is to place a reservoir of barium 1 meter above the patient so that constant hydrostatic pressure is generated. With experience (and depending upon the clinical status of the patient), a physician may undertake a more aggressive reduction.

In a typical ileo-colic intussusception, the barium column appears as an intraluminal-filling defect. The intussusception can be found in any part of the large bowel, even the rectum. Occasionally, some barium may coat the outer surface of the intussusciptens, resulting in a coiled spring pattern.

Successful reduction is indicated by the free flow of contrast into the small bowel. Reduction is complete only when a good portion of the distal ileum is filled with barium, thus excluding ileo-ileal intussusception. Other indications of successful reduction include relief of symptoms and disappearance of the abdominal mass. A characteristic sound also may be appreciated with auscultation. In occasional patients, the contrast material does not reflux freely into the small bowel even with a complete reduction, but no filling defect in the cecum is present apart from the ileocecal valve and symptoms and signs of these patients resolve.

A post-reduction filling defect in the cecum commonly is seen, probably the result of residual edema in the ileocecal valve. However, benign and malignant lesions cannot be distinguished by radiologic examination alone. As a result, a repeat study or even laparotomy may indicate if any concern of a benign or malignant lead point exists [53].

After the successful reduction of an ileo-colic intussusception, a temperature higher than 38ºC (100.4ºF) often is noted because of the release of endotoxin, cytokines, or bacterial translocation. The patient should be admitted to the hospital for 12 to 24 hours for observation. Nasogastric suction usually is maintained until bowel function has returned and the patient has had passage of a bowel movement. Feedings then are advanced as tolerated.

**Air contrast** — Air reduction techniques have gained popularity as an alternative to the contrast hydrostatic methods [54-57]. The technique begins with insertion of a Foley catheter into the rectum. Fluoroscopy is used to assess the presence of bowel gas in the abdomen. Air is then instilled, initially by hand pump, until the diagnosis is established and the intussusceptum is pushed back gently [49,50].

An electric pump is connected if the intussusceptum stops moving despite the use of the hand pump. Both of these pumps must have a pressure
release system so that the pressure remains between 80 and 120 mmHg. Carbon dioxide can be used instead of air. It has the advantage of being absorbed rapidly from the gut, is associated with less discomfort, and is less dangerous than air, which potentially could cause an air embolism (although air embolisms have not been reported). Reflux of air into the terminal ileum and the disappearance of the mass at the ileocecal valve indicate reduction.

An advantage of this method is the ease with which the intussusception can be reduced with less radiation exposure and cost, and the relatively harmless nature of air in the peritoneal cavity, compared with that of other contrast materials.

**Risk and complications** — The main risk of hydrostatic or pneumatic reduction is perforation of the bowel, which occurs in 1% or fewer patients [58-60]. The perforation usually occurs on the distal side of the intussusception, often in the transverse colon, and commonly where the intussusception was first demonstrated by radiographic studies [61,62]. The risk of perforation is greatest in infants younger than 6 months of age who have had symptoms for three days or longer and in patients with evidence of small bowel obstruction [63]. Nonoperative reduction should not be attempted in patients with prolonged symptoms or any signs of peritoneal irritation or free peritoneal air.

**Success rate** — Nonoperative reduction using barium or air contrast techniques is successful in approximately in 60 to 90% of patients with ileo-colic intussusception [3,64-66]. Success is more likely to be achieved in patients with idiopathic intussusception (ie, no identifiable lead point), although it also can be accomplished in patients with a recognized lead point [53]. Successful radiographic reduction is more likely if, during reduction, the small bowel is visualized before the appendix [67].

Ileo-ileo-colic intussusception may be more difficult to reduce because the barium often percolates along the loops of small bowel in the colon, reducing the effective pressure of the enema.

In addition, success is less likely to be achieved in infants younger than 1 year of age (particularly younger than 3 months), in children older than 5 years of age, and when plain films show signs of intestinal obstruction [30,58,64]. Although some authors have noted a reduced likelihood of reduction when symptoms have been present for longer than 48 hours [30,58,66,68], others have found no such correlation [65]. In these high-risk cases, the barium study should still be performed to confirm the diagnosis and attempt therapy, but a pediatric surgeon should be readily available in the imaging department.
Recurrence after successful nonoperative reduction is close to 10% [30,69,70]. Recurrence is not necessarily an indication for surgery. Each recurrence should be handled as if it were the first episode, provided that each is successfully reduced [71].

**SURGERY** — Surgery is indicated when nonoperative reduction is incomplete or when a persistent filling defect, indicating a mass lesion, is noted [71]. As noted above, a residual filling defect may be seen because of edema of the ileocecal valve. Thus, when a filling defect appears to be associated with edema in the area, an ultrasonographic or repeat contrast study should be performed provided that symptoms have resolved.

Broad-spectrum intravenous antibiotics should be given before surgery. Manual reduction at operation is attempted in most cases, but resection with primary anastomosis needs to be performed if manual reduction is not possible or if a lead point is seen.

**REFERENCES**