Intussusception in children

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INTRODUCTION

Intussusception refers to the invagination (telescoping) of a part of the intestine into itself. It is the most common abdominal emergency in early childhood, particularly in children younger than two years of age [1]. The majority of cases in children are idiopathic, and pathologic lead points are identified in only 25 percent of cases involving children [2]. Intussusception is unusual in adults, and the diagnosis is commonly overlooked. In the majority of cases in adults, a pathologic cause is identified [3].

The first successful surgical correction of intussusception in an infant was described in 1871 by Hutchinson. In 1876, Hirschsprung reported his experience with the treatment of intussusception by enema. This technique was associated with approximately 35 percent mortality, considerably better than the mortality rates after surgery. Reduction of intussusception by fluoroscopy-guided enema was described as early as 1927, and was soon incorporated by radiologists as part of their expertise. The technique has further evolved to include ultrasound as an additional imaging option. Reduction was traditionally performed using barium or other liquid contrast agents (hydrostatic enema), but can also be performed using air or carbon dioxide (pneumatic enema) [4].

The clinical manifestations, diagnosis, and management of intussusception in infants and children are discussed below. Intussusception in adults is presented separately. (See "Management of small bowel obstruction in adults".)

EPIDEMIOLOGY

Intussusception typically presents between 6 and 36 months of age, and is the most common cause of intestinal obstruction in this age group. Approximately 60 percent of children with intussusception are younger than one year old, and 80 to 90 percent are younger than two years [5]. 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 [6]. After the third year of life, the incidence fell to less than one-half of these rates.

Although intussusception is most common in infants and toddlers, it is important to consider this diagnosis in children outside this age range. Approximately 10 percent of cases occur in children over five years, 3 to 4 percent in those over 10 years, and 1 percent in infants younger than 3 months [6,7]. When intussusception does occur outside of the typical age range, it is likely to be associated with a pathologic lead point, which may include reactive lymphoid hyperplasia. (See 'Lead point' below.)

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 [5].

TYPES AND TERMINOLOGY

Intussusception refers to the invagination (telescoping) of a part of the intestine into a more distal segment. The proximal segment is known as the **intussusceptum**, and the distal segment into which it telescopes is known as the **intussuscipiens**.

The intussusception is classified by the location (figure 1):

- **Ileocolic** intussusception involves the ileocecal junction, and accounts for 90 percent of all cases [5].
- **Ileo-ileal**, **ileo-ileo-colic**, **jejuno-jejunal**, **jejuno-ileal**, or **colo-colic** intussusception also have been described. Ileo-ileo-colic intussusception refers to an ileo-ileal intussusception that telescopes further through the ileocecal valve into the right colon.

The intussusception is considered **idiopathic** if there is no identifiable mass in the intussusceptum (no "lead point") (see 'Lead point' below). The intussusception can be idiopathic even if there was a likely or possible triggering event, such as viral or other enteric infection. (See 'Idiopathic' below.)

PATHOGENESIS

As the intussusception develops, the mesentery is dragged into the bowel. This leads to the development of venous and lymphatic congestion with resulting intestinal edema. If untreated, the process can ultimately lead to ischemia, perforation, and peritonitis.

Idiopathic — Approximately 75 percent of cases of childhood intussusception are considered to be idiopathic because there is no clear disease trigger or pathologic lead point. Idiopathic intussusception is most common in children between three months and five years of age [2].

Influence of viral factors — An increasing body of evidence suggests that viral triggers may play a role in some cases, as illustrated by the following observations:

- The incidence of intussusception has a seasonal variation, with peaks coinciding with seasonal viral gastroenteritis in some populations [6,8].
- 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. 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). The risk of intussusception associated with currently licensed vaccines is discussed in a separate topic review. (See "Rotavirus vaccines for infants", section on 'Intussusception'.)
- Approximately 30 percent 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 percent of cases, there is evidence of recent infection with enteric and nonenteric species of adenovirus [9-14]. In a prospective case-control

study examining a variety of possible infectious triggers for intussusception in Vietnam and Australia, infection with adenovirus, species C emerged as the strongest predictor of intussusception in both populations [12]. In these populations, rotavirus infection and poliovirus vaccine administration were not associated with intussusception. Another study found an association with human herpes virus 6 [13].

Viral infections, including enteric adenovirus, can stimulate lymphatic tissue in the intestinal tract, resulting in hypertrophy of Peyer patches in the lymphoid-rich terminal ileum, which may act as a lead point for ileocolic intussusception (picture 1) [9,15]. Because of this putative association with lymphoid hyperplasia, treatment with glucocorticoids has been suggested to prevent recurrence, but this approach is not generally recommended. (See 'Recurrence' below.)

Other enteric infections — Bacterial enteritis is also associated with intussusception. In a series of 1412 cases of bacterial enteritis seen at military treatment facilities, intussusception ensued in 37 patients (comprising 12.6 percent of all intussusceptions seen at these facilities) [16]. This association was noted for infection with *Salmonella*, *Escherichia coli*, *Shigella*, or *Campylobacter*. Most cases of intussusception occurred within the first month after the bacterial enteritis.

Lead point — A lead point is a lesion or variation in the intestine that is trapped by peristalsis and dragged into a distal segment of the intestine, causing intussusception (figure 1). A Meckel diverticulum, polyp, duplication cyst, tumor, hematoma, or vascular malformation can act as a lead point for intussusception [17].

Underlying disorders — In approximately 25 percent of cases, an underlying disease causes a pathologic lead point for the intussusception, which may be focal or diffuse. Such triggers account for a greater proportion of cases of intussusception in children younger than three months or older than five years [1,18,19]. Nonetheless, it is important to be vigilant for pathologic lead points in children of any age.

A variety of conditions have been associated with intussusception, including Meckel diverticulum [20], polyps [21], small bowel lymphoma [22-24], duplication cysts [25,26], vascular malformations [27], inverted appendiceal stumps [28,29], parasites (eg, *Ascaris lumbricoides*) [30,31], immunoglobulin A vasculitis (IgAV; Henoch-Schönlein purpura [HSP]) [32], cystic fibrosis [33], and hemolytic-uremic syndrome [34]. Meckel diverticulum is the most common pathologic lead point in most case series in children, followed by polyps,

and then either duplication cysts or IgAV (HSP) [19]. (Refer to appropriate UpToDate topic reviews.)

The mechanisms leading to intussusception depend upon the specific cause. As examples:

- Meckel diverticulum, polyps, duplication cysts, lymphomas, areas of reactive lymphoid hyperplasia, or other focal abnormalities of the intestinal tract act as lead points for peristalsis advancing the intestine into a distal segment of intestine. (See "Meckel's diverticulum".)

- In patients with IgAV (HSP), a small bowel wall hematoma acts as the lead point. Intussusception typically occurs after resolution of the HSP-associated abdominal pain. (See "IgA vasculitis (Henoch-Schönlein purpura): Clinical manifestations and diagnosis".)

- Similarly, in patients with hereditary or acquired coagulopathy (eg, von Willebrand disease or anticoagulant therapy), a bowel wall hematoma may act as a lead point [17]. In other cases, a bowel wall hematoma may mimic the symptoms and ultrasound findings of intussusception [35,36].

- In patients with cystic fibrosis, thick inspissated stool may act as the lead point [33]. (See "Cystic fibrosis: Overview of gastrointestinal disease".)

- Celiac disease appears to be associated with a modestly increased risk for intussusception, as suggested by a large study in Sweden [37]. The proposed mechanism is that celiac disease may promote small bowel intussusception because of dysmotility and excessive secretions or bowel wall weakness [38,39]. Subclinical intussusception has been reported in approximately 25 percent of children with newly diagnosed celiac disease and typically resolves spontaneously during the first few days on a gluten-free diet, without radiologic or operative intervention [40]. Conversely, the Swedish study found no association between intussusception and future celiac disease, arguing against the need for routine antibody testing for children with intussusception [37]. (See "Epidemiology, pathogenesis, and clinical manifestations of celiac disease in children".)

- Patients with Crohn disease may develop intussusception because of inflammation and stricture formation [41]. (See "Clinical manifestations and complications of

inflammatory bowel disease in children and adolescents".)

Postoperative — Small bowel intussusception (usually jejuno-jejunal or ileo-ileal) has been described in the postoperative setting where it is an uncommon but insidious cause of intestinal obstruction [42-45]. Most cases occur after abdominal surgery (especially open procedures), but there is also a modest increased risk after nonabdominal procedures. The intussusception is thought to be caused by uncoordinated peristaltic activity and/or traction from sutures or devices such as a gastrojejunal feeding tube [46]. Affected patients typically do well for several days and may even resume oral intake before developing symptoms of mechanical obstruction. In many cases, this is an incidental finding and/or spontaneously resolves. (See 'Spontaneous reduction of intussusception' below.)

The diagnosis can be difficult to establish because intussusception may be confused with postoperative paralytic ileus. Evaluation with ultrasonography or computed tomography (CT) scanning can establish the diagnosis, monitor for spontaneous reduction, and help to predict which children are likely to need surgical reduction. Because most cases of postoperative intussusception occur in the small intestine, contrast enemas do not usually contribute to the diagnosis. (See 'Small bowel intussusception' below.)

CLINICAL MANIFESTATIONS

- **Typical presentation** – The classic presentation of intussusception is an infant or toddler with the sudden onset of intermittent, severe, crampy, progressive abdominal pain, accompanied by inconsolable crying and drawing up of the legs toward the abdomen, often with pallor [5,47]. The episodes usually occur at 15- to 20-minute intervals. They become more frequent and more severe over time. Vomiting is often a prominent symptom, often starting shortly after the first episodes of abdominal pain. Initially, emesis may be nonbilious, but it often becomes bilious as the obstruction progresses. A sausage-shaped abdominal mass may be felt in the right side of the abdomen. The stool is grossly bloody in up to 50 percent of cases, and an additional 25 percent have occult blood [48]. In some cases, the stool may be a mixture of blood and mucous, giving it the appearance of currant jelly, but this is a late finding and seen in a minority of patients. (See "Causes of acute abdominal pain in children and adolescents".)

Between the painful episodes, the child may behave relatively normally and be free of pain. As a result, initial presentation can be confused with that of gastroenteritis [49]. As symptoms progress, increasing lethargy often develops, which can be mistaken for meningoencephalitis. Some authors have hypothesized that the lethargy is caused by increased levels of endogenous opioids, suggested by cases with pupillary miosis and reversal with naloxone [50].

- **Atypical presentation** – 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 [49,51]. Up to 20 percent of young infants have no obvious pain, and approximately one-third of patients do not pass blood or mucus, nor do they develop an abdominal mass. Many older children have pain alone without other signs or symptoms.

  Occasionally, the initial presenting sign is lethargy or altered consciousness alone, without pain, rectal bleeding, or other symptoms that suggest an intraabdominal process [52-56]. This clinical presentation primarily occurs in infants and is often confused with sepsis. Thus, intussusception should be considered in the evaluation of otherwise unexplained lethargy or altered consciousness, especially in infants.

- **Incidental finding** – An intussusception is sometimes discovered incidentally during an imaging study performed for other reasons or for nonspecific symptoms. If these intussusceptions are short in duration and the patient is asymptomatic, they can be managed with observation alone. Patients with minimal symptoms may also not require intervention. (See 'Spontaneous reduction of intussusception' below.)

EVALUATION

History and physical examination — Because the presentation of intussusception is variable, a high index of suspicion for intussusception is important, particularly in children between three months and five years of age, which is the peak age range for idiopathic intussusception, or those with other risk factors. However, it is important to consider the possibility of intussusception in children who are younger or older than this age range. (See 'Epidemiology' above.)

- **History** – The history is directed to identifying features suspicious for

intussusception, which are detailed above (see 'Clinical manifestations' above). Although most patients have several of these features, intussusception should be considered if any of these are present:

- Intermittent, severe, crampy, progressive abdominal pain
- Vomiting
- Rectal bleeding (gross or occult)
- Lethargy (often episodic)

The history should also solicit information that would suggest a different cause of the symptoms, including (see 'Differential diagnosis' below):

- Fever (suggests gastroenteritis, appendicitis, or other infection but also may be a presenting symptom in children with intussusception [57])
- Ill contacts (suggests gastroenteritis)
- Exposure to potential toxins (medications, alcohol, or poisons)

- **Physical examination** – On physical examination, features suspicious of intussusception include (see 'Clinical manifestations' above):

  - No abdominal tenderness, or only focal tenderness (especially in the right mid or upper abdomen)
  - Abdomen not distended
  - Lethargy or altered consciousness (often episodic)

  Features that are more specific for intussusception, but are present in a minority of patients, include:

  - Right lower quadrant that is scaphoid (empty; Dance's sign)
  - Palpable "sausage-shaped" mass in the right mid or upper abdomen

Differential diagnosis — The differential diagnosis of intussusception depends on the presenting symptoms:

- Rectal bleeding and vomiting:

  - Meckel diverticulum
  - Bacterial or amoebic colitis

- Malrotation with midgut volvulus

These and other causes of rectal bleeding are summarized in separate topic reviews. (See "Lower gastrointestinal bleeding in children: Causes and diagnostic approach" and "Meckel's diverticulum" and "Intestinal malrotation in children".)

- Acute onset of crampy abdominal pain:

  - Gastroenteritis
  - Appendicitis
  - Mesenteric ischemia
  - Ovarian torsion
  - Malrotation with volvulus
  - Incarcerated hernia
  - Peritonitis

  These and other causes of acute abdominal pain are discussed separately. (See "Causes of acute abdominal pain in children and adolescents".)

- Lethargy and coma:

  - Trauma
  - Infections, including sepsis
  - Metabolic derangements
  - Intoxications

  These and other causes of altered consciousness are discussed separately. (See "Evaluation of stupor and coma in children".)

Approach to diagnostic testing — The optimal strategy for diagnosis and treatment depends on the clinical suspicion for intussusception (typical or atypical presentation), and on the preference and experience of the consulting radiologists [58].

Typical presentation — Patients with a typical presentation (eg, infant or toddler with sudden onset of intermittent severe abdominal pain with or without rectal bleeding) or characteristic findings on radiography or ultrasound, may proceed directly to nonoperative reduction using hydrostatic (contrast or saline) or pneumatic (air) enema, performed under either sonographic or fluoroscopic guidance. In these cases, the procedure is both

diagnostic and therapeutic. (See 'Nonoperative reduction' below.)

Atypical presentation — For many other patients, the diagnosis is unclear at presentation, especially in children who are younger or older than the typical age group for intussusception. In this case, initial workup may include abdominal ultrasound or abdominal radiographs, provided that these studies do not significantly delay the definitive treatment of intussusception. If the ultrasound supports the diagnosis of intussusception, nonoperative reduction is then performed, provided that the child has normal vital signs and no signs of peritonitis [4]. (See 'Nonoperative reduction' below.)

Ultrasonography — Ultrasonography is the method of choice to detect intussusception in many institutions [59]. The sensitivity and specificity of this technique approach is 100 percent in the hands of an experienced ultrasonographer [60]. Negative predictive value is also nearly 100 percent, so a negative study by an experienced sonographer can definitively rule out intussusception [61,62]. Ultrasound is better able to detect pathologic lead points than fluoroscopic techniques, can be used to monitor the success of a reduction procedure, and does not expose the patient to radiation [19,61,63-65]. Ultrasound can also evaluate for several alternative causes for the child's symptoms, such as appendicitis or hydronephrosis. In other institutions, fluoroscopy is used as the primary diagnostic and therapeutic procedure for intussusception. (See 'Fluoroscopic or sonographic guidance' below.)

The classic manifestation of intussusception on ultrasound is a "target sign" (also known termed "bull's eye" or "coiled spring"), representing layers of the intestine within the intestine (image 1). For ileocolic intussusception, which is the most common type, the "target sign" usually is in the right lower quadrant. Color duplex imaging may reveal a lack of perfusion in the intussusceptum, indicating the development of ischemia. An advantage of ultrasonography over fluoroscopy is that it can diagnose the rare ileo-ileal intussusception; ultrasound also can identify the lead point of intussusception in approximately two-thirds of cases in which underlying pathology exists [66].

The possibility of small bowel intussusception (eg, jejuno-jejunal or jejuno-ileal, rather than ileocolic intussusception) is suggested by location of the intussusception outside of the right lower quadrant (eg, in the paraumbilical or left abdominal region), and/or lesion size ≤3 cm [67]. If small bowel intussusception is suspected and the child's symptoms are mild, the first step is to repeat the ultrasound to see if the finding persists because most small

bowel intussusceptions will spontaneously reduce. If the finding persists, evaluation with a computed tomography (CT) scan may help to confirm the location of the intussusception and whether there is a lead point. In small bowel intussusceptions, the length of the intussusceptum, as measured by ultrasound or CT, and the patient's symptoms help to determine prognosis and management. (See 'Small bowel intussusception' below.)

Abdominal plain film — In patients with suspected intussusception, the initial evaluation usually should include two-view plain abdominal radiographs. The main purpose of the radiograph is to exclude perforation, which, if present, requires operative management rather than nonoperative reduction [68,69]. They are also useful to screen for other causes of abdominal symptoms that might be detected by radiography.

Plain radiographs are not sufficiently sensitive or specific to diagnose intussusception, but may support the diagnosis if one or more of the following findings are present:

- Signs of intestinal obstruction, which may include massively distended loops of bowel with absence of colonic gas (image 2).
- A target sign, consisting of two concentric radiolucent circles superimposed on the right kidney, represents peritoneal fat surrounding and within the intussusception. In one report, this finding was present in 26 percent of patients with intussusception [70].
- A crescent sign, which is a soft tissue density (representing the intussusceptum) projecting into the gas of the large bowel.
- An obscured liver margin [69].
- Lack of air in the cecum, which prevents its visualization [69].
- Pneumoperitoneum, which suggests that bowel perforation has occurred; this is rarely seen.
- Intracolonic soft tissue mass (image 3) [71].

Plain radiographs should not be used to exclude intussusception, particularly in patients with a high clinical suspicion. The sensitivity for abdominal radiographs to diagnose intussusception is less than 48 percent while the specificity is 21 percent [4]. In one study,

more than 20 percent of patients with intussusception had negative plain films [72]. In patients with a low clinical suspicion of intussusception, the presence of air in the cecum or terminal ileum helps to exclude intussusception [73]. However, ultrasound is far superior to radiography both for diagnosing and excluding intussusception.

Computed tomography scan — An intussusception can be recognized on CT, which may also identify the cause (image 4). However, CT cannot be used to reduce the intussusception, can be time-consuming in children who may require sedation, and also exposes the child to substantial radiation. Thus, CT generally is reserved for patients in whom the other imaging modalities are unrevealing, or to characterize pathologic lead points for intussusception detected by ultrasound [19].

DIAGNOSIS

The diagnosis of intussusception can be made in any of the following ways:

- By diagnostic ultrasound – This is the usual procedure where diagnostic ultrasound is readily available (eg, in the emergency department), or for patients with atypical presenting features. Once the diagnosis is established, the patient should proceed promptly to treatment (typically nonoperative reduction under ultrasonographic or fluoroscopic guidance). (See 'Nonoperative reduction' below.)

- By ultrasound or fluoroscopy, as part of an attempt at nonoperative reduction – This is the usual procedure for patients with typical presenting features. In this case, the procedure is both diagnostic and therapeutic. (See 'Nonoperative reduction' below.)

- Incidental finding on ultrasound or computed tomography (CT) – In this case, next steps depend upon the patient's symptoms since incidentally discovered intussusception can be asymptomatic and often resolves spontaneously. (See 'Spontaneous reduction of intussusception' below.)

TREATMENT

The approach to treatment of intussusception depends upon patient characteristics:

- Most patients – Patients with a high clinical suspicion and/or imaging evidence of

ileocolic intussusception, normal vital signs, and no evidence of bowel perforation should be treated with nonoperative reduction as described below. It is important that nonoperative reduction be performed at an institution with extensive experience in the technique and by providers with experience in managing potential complications of nonoperative reduction, such as tension pneumoperitoneum [74]. Patients presenting to an institution without this experience should be transferred if this can be done promptly. (See 'Nonoperative reduction' below.)

- Acutely ill or with perforation – Surgical treatment is indicated as a primary intervention for patients with suspected intussusception who are acutely ill or have evidence of perforation. Surgery may also be appropriate when the patient is treated in a location where the radiographic facilities and expertise to perform nonoperative reduction are not readily available and if timely patient transfer would not be feasible. Operative intervention is also indicated for patients in whom nonoperative reduction is unsuccessful, or for evaluation or resection of a pathologic lead point. (See 'Surgery' below.)

- Small bowel intussusception – Patients with intussusception limited to the small bowel (ileo-ileal, jejuno-ileal, or jejuno-jejunal) are managed differently. (See 'Small bowel intussusception' below.)

Nonoperative reduction — Nonoperative reduction using hydrostatic or pneumatic pressure by enema is the treatment of choice for an infant or child with ileocolic intussusception who is clinically stable and has no evidence of bowel perforation or shock, when appropriate radiologic facilities are available. Enema reduction has high success rates in children with ileocolic intussusception. In settings in which nonoperative reduction is not available (eg, in resource-limited countries), patients with intussusception usually should be managed with surgical reduction. (See 'Surgery' below.)

Before attempting reduction by enema, the patient should be stabilized and given intravenous fluids if there is evidence of volume depletion. The surgical team should be notified before attempting nonoperative reduction and remain immediately available because there is a risk of perforation during the procedure. Surgical intervention also may be necessary if nonoperative reduction fails to reduce the intussusception. (See 'Surgery' below.)

Most radiologists perform nonoperative reduction of ileocolic intussusception without

sedation or general anesthesia [75]. A few practices routinely use sedation or general anesthesia and continue to demonstrate high success rates, while maintaining low complication rates related to the sedation or anesthesia [76,77]. However, the rate of bowel perforation in sedated patients was slightly higher than in non-sedated patients (3 of 124 patients, versus 0 of 90 patients) [76]. Having an anesthesiologist available emergently may be a logistical limitation for some practices. In our practice we do not routinely utilize sedation or a general anesthetic.

There is no need for prophylactic intravenous antibiotics prior to or during nonoperative reduction, except for children with hemodynamic instability or critical illness. This is consistent with recommendations from the American Pediatric Surgical Association, based on observational evidence summarized in a systematic review [78]. Some institutions give prophylactic antibiotics because of the potential risk of bacteremia and perforation with these procedures. However, there is no evidence that this practice is beneficial, likely because bacteremia and perforation are rare [79]. (See 'Risk and complications' below.)

Fluoroscopic or sonographic guidance — Reduction of intussusception is most commonly performed under fluoroscopic guidance, using either hydrostatic (saline or contrast) or pneumatic (air) enema. Ultrasound guidance is also an increasingly used option, and has the advantage of avoiding ionizing radiation, as well as improved detection of pathologic lead points compared with fluoroscopic technique. As a result, some providers with expertise in this technique prefer to use ultrasound guidance to reduce ileocolic intussusception [80-86]. A disadvantage of ultrasound is that it can only be used for hydrostatic reduction. Fluoroscopy and ultrasound-guided techniques have comparable success rates for reduction of intussusception, ranging from 80 to 95 percent [59,81,82,87,88].

The techniques are performed as follows:

- **Sonographic guidance** – Sonographic guidance requires a hydrostatic technique (saline enema) to provide retrograde pressure because use of air would interfere with ultrasound visualization. Sonographic signs of successful reduction include the disappearance of the intussusception and the appearance of water and bubbles in the terminal ileum.

- **Fluoroscopic guidance** – Under fluoroscopy for a typical ileocolic intussusception, the intussusceptum appears as a filling defect within the bowel lumen. This is seen as

either a low density filling defect when contrast is used for hydrostatic reduction, or a higher density filling defect when air is used as a negative contrast with pneumatic reduction techniques (image 5 and image 6). The intussusception can be found in any part of the large bowel, even the rectum. Occasionally, contrast may coat the outer surface of the intussuscipiens, resulting in a coiled spring pattern.

Successful reduction is indicated by the free flow of contrast or air into the small bowel. Reduction is complete only when a good portion of the distal ileum is filled with contrast or air, thus excluding ileo-ileal intussusception. Other indications of successful reduction include relief of symptoms and disappearance of the abdominal mass. Occasionally, the contrast material does not reflux freely into the small bowel even after a complete reduction [89]; however, a successful reduction is suggested by lack of a filling defect in the cecum (apart from the ileocecal valve), and clinical resolution of symptoms and signs. A post-reduction filling defect in the cecum commonly is seen, probably the result of residual edema in the ileocecal valve. However, this finding cannot be distinguished from a focal lead point by radiologic examination alone. As a result, a repeat study or even laparotomy may be indicated if there is any concern for a focal lead point [90].

Hydrostatic or pneumatic reduction — Either pneumatic (air) or hydrostatic (saline or contrast) technique is acceptable for reduction of intussusception in stable children. The choice of technique depends primarily upon the expertise and comfort level of the radiologist, and availability of the necessary equipment.

In our institution, pneumatic reduction is the method of choice, rather than hydrostatic reduction [91]. This is because pneumatic reduction of intussusception has somewhat higher success rates and no increased risk of perforation. This was shown in a meta-analysis, which reported a success rate of 83 percent for pneumatic reduction, and 70 percent for hydrostatic reduction [92]. There was no difference in rate of perforation (0.39 and 0.43 percent, respectively), or of early recurrence within the first 48 hours [92]. In addition, pneumatic reduction may be advantageous if perforation occurs (see 'Risk and complications' below). Finally, pneumatic reduction typically requires a lower overall radiation exposure dose compared with hydrostatic enema, which is independent of the fluoroscopy time required for the procedure. This is likely because air has a lower density than the contrast that is typically used for hydrostatic reduction, and thus reduces the exposure required for generating the image with fluoroscopy [93].

In other institutions the hydrostatic enema technique is used, either because it is the primary nonoperative method available, or if ultrasound guidance is used.

The techniques are performed as follows:

- **Hydrostatic technique** – The standard method of reduction is to place a reservoir of contrast 1 meter above the patient so that constant hydrostatic pressure is generated. With experience (and depending upon the clinical status of the patient), a clinician may undertake a more aggressive reduction (ie, increase the hydrostatic pressure by raising the reservoir higher above the patient).

  - When hydrostatic reduction is performed under fluoroscopic guidance, a water-soluble contrast enema is preferred because of the risk of perforation before or during the procedure. Water-soluble agents reduce the risk of electrolyte disturbances and peritonitis in patients in whom perforation has occurred [81]. Traditionally, barium was used as the contrast agent in most North American and European centers (image 5) [94-96].

  - When hydrostatic reduction is performed under ultrasonographic guidance, normal saline is used for the enema. (See 'Fluoroscopic or sonographic guidance' above.)

- **Pneumatic technique** – Pneumatic (air reduction) techniques are now generally preferred to the hydrostatic methods if fluoroscopy is used for guidance [91,92]. The pneumatic technique cannot be used with ultrasonography, because the air interferes with ultrasound visualization.

  The technique begins with insertion of a Foley catheter (or equivalent) or rectal tube into the rectum. A tight anal seal is formed around the catheter or tube, typically using tape; a good seal is critical as a means to minimize air leak and maintain adequate pressure for reduction throughout the procedure. If a Foley catheter is utilized, the balloon may be inflated with up to 30 mL of air to further help maintain air retention during reduction; this is done under real-time fluoroscopy to ensure appropriate degree of inflation within the rectal ampulla. Inflation of Foley balloon should not be done in children 10 months or younger, to minimize risk of bowel perforation during the pneumatic reduction procedure [97]. Fluoroscopy is used to monitor the procedure. Air is then instilled until the intussusceptum is pushed back

gently, taking care to avoid excessive pressure [94,95]. A sphygmomanometer can be used to monitor colonic intraluminal pressure (typically not to exceed 120 mmHg) to aid in reduction [98]. Carbon dioxide can also 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).

A successful reduction of the intussusception is indicated by a rush of air reflux into the terminal ileum, a sudden drop in the intraluminal pressure, and the disappearance of the mass at the ileocecal valve (image 6). Water-soluble contrast material can be instilled to confirm the reduction, or the air reduction can be repeated if the completeness of reduction is questioned [99].

Risk and complications — The main risk of hydrostatic or pneumatic reduction is perforation of the bowel, which occurs in 1 percent or fewer patients (image 7) [81,92,100-102]. 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 [103,104]. Risk factors for perforation include age younger than six months, long duration of symptoms (eg, three days or longer), and evidence of small bowel obstruction; use of higher pressures during the reduction is a contributing factor in some patients [81,105]. Nonoperative reduction should not be attempted in patients with any signs of peritoneal irritation or free peritoneal air.

The pneumatic reduction technique provides an advantage if perforation occurs because air is generally less harmful than other contrast materials in the peritoneal cavity [81]. When perforation is noted with air reduction, the colonic wall tears are smaller than those observed with the hydrostatic contrast techniques, and peritoneal pathology tends to be minimal. Needle decompression of the abdomen may be necessary if the excess air in the peritoneal cavity compromises the patient's respiratory status [74].

Success rate — In institutions with extensive experience, nonoperative reduction using hydrostatic or pneumatic techniques is successful in approximately 70 to 85 percent of patients with ileocolic intussusception [6,81,92,106,107]. 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 [90]. The supplemental use of glucagon or atropine to relax colonic smooth muscle had no benefit

in a double-blind study and a subsequent observational study [108,109].

Ileo-ileo-colic intussusception may be more difficult to reduce because the contrast 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 one year of age (particularly younger than three months), and in children older than five years of age (due to increased likelihood of a pathologic lead point), and when plain films show signs of intestinal obstruction [100,110-112]. In a systematic review and meta-analysis (40,133 cases), the probability of successful nonoperative reduction was associated with shorter duration of symptoms (<24 hours), presence of abdominal pain, and intussusception located in the right abdomen by ultrasound [113]. Failure of nonoperative reduction was associated with the presence of fever, rectal bleeding, and vomiting and with the ultrasonographic findings of left-sided intussusception and presence of ascites or trapped fluid.

For patients presenting with prolonged symptoms, nonoperative reduction is often successful, although the overall success rate is somewhat lower than in patients with a more recent onset of symptoms. Studies have reported successful pneumatic reduction of intussusception after prolonged symptoms, as long as 240 hours (and a median time of 18.5 hours), after initial symptoms, suggesting that nonoperative reduction should remain the primary initial treatment method regardless of the length of history in a stable child [114]. In these more complicated cases, a pediatric surgeon should be readily available in case the nonoperative reduction is not successful.

Delayed repeat enema — If the nonoperative reduction is **partly** successful (ie, the intussusception moved but was not completely reduced) and the patient remains stable, it is reasonable to perform a second attempt at nonoperative reduction, often referred to as delayed repeat enema. The time gap or delay between nonoperative enema attempts varies from 30 minutes to a few hours. There is some evidence that this approach is successful and avoids surgery for some patients who would have otherwise required surgical reduction following initial unsuccessful reduction attempts [78,81,115-118].

Delayed repeat enema should **not** be attempted in patients in whom the initial attempt at nonoperative reduction was **completely** unsuccessful (ie, the intussusception did not move), or in those who are unstable. Such patients should proceed promptly to surgery.

Post-procedure management — There are no compelling data or widely used guidelines for clinical care after successful nonoperative reduction of an ileocolic intussusception. Our own practice is as follows:

- **Antibiotics** – We do not routinely give intravenous antibiotics after a successful nonoperative reduction; we give antibiotics only if there are progressive symptoms and signs of sepsis, or if there is a suspicion of intestinal perforation despite successful nonoperative reduction. A low-grade fever (temperature higher than 38 to 39°C [100.4 to 102°F]) is often noted during the first few hours after the procedure, probably because of the systemic inflammatory response associated with transient bowel ischemia.

- **Diet** – We offer clear fluids as soon as the patient is awake and alert, and advance the diet as tolerated [119]. We place a nasogastric tube only if the patient has recurrent vomiting or clinical evidence of bowel obstruction.

- **Disposition** – If patients are well-appearing and tolerating clear fluids within two hours after nonoperative reduction, we typically discharge the patient as long as they are able to return to the hospital promptly for recurrent symptoms.

  At many institutions, it has been common practice to admit the patient for 12 to 24 hours of observation, primarily to monitor for early recurrence. The duration of the observation is guided by monitoring symptoms and vital signs, physical examination, the distance the family lives from the hospital, and tolerance of a diet. However, the utility of routine inpatient observation has been questioned because only approximately 4 percent of patients experience recurrence within 48 hours and recurrence rates of intussusception do not differ between children observed in the hospital and those discharged home [120-122]. A meta-analysis and observational studies have concluded that children who had uncomplicated enema reduction and are afebrile, hemodynamically normal, asymptomatic, and tolerating a diet can be safely discharged home. For children with these characteristics, outpatient management is associated with no increase in the rate of return to the emergency department, recurrence, need for operation, or mortality compared with inpatient observation [123,124].

- **Imaging** – No routine imaging is required for patients that remain asymptomatic.

Should symptoms develop that are suspicious for recurrent intussusception, an ultrasound study is warranted. If there is concern for perforation, the patient should be evaluated with conventional radiographs. (See 'Recurrence' below.)

- **Consultations** – We consult the pediatric surgical service promptly if there is suspected intestinal perforation or incomplete reduction of intussusception.

- **Follow-up** – When the patient is discharged, we give instructions to call promptly if symptoms recur, and plans for follow-up by a clinician within one to two days.

Recurrence — Intussusception recurs in 3.7 to 20 percent of children after successful nonoperative reduction [120,121,125-128]. Approximately one-half of the recurrences are within the first 72 hours after nonoperative reduction (perhaps because of residual bowel edema or inflammation, which may act as a lead point), and the remainder occur weeks or months later [120].

Management of recurrent intussusception depends upon the patient's individual characteristics. Recurrence is not necessarily an indication for surgery. In general, each recurrence should be handled as if it were the first episode, provided that each attempt at nonoperative reduction is successful and the patient remains stable [126,127]. However, imaging studies should be reviewed carefully for the possibility of a pathologic lead point. If a lead point is identified, the patient may still be treated with nonoperative reduction, particularly if the lead point is diffuse (eg, immunoglobulin A vasculitis [IgAV; Henoch-Schönlein purpura (HSP)]) [19]. Surgical exploration should be performed for any patient who is unstable, and should also be considered for those with a focal lead point or multiple recurrences.

The risk for recurrence is associated with patient age older than one year, but not the duration of symptoms at the initial presentation [129,130]. The rate is similar for the different nonoperative techniques of reduction described above [121,125]. Multiple recurrences of intussusception are associated with the presence of a pathologic lead point, but may also occur in those with "idiopathic" intussusception. In one series, 19 percent of children with two or more episodes of intussusception had a pathologic lead point, whereas 4 percent of children without a recurrence had a pathologic lead point [19].

Among children with idiopathic intussusception, lymphoid hyperplasia may act as a lead point. Because of this putative association, treatment with glucocorticoids has been

suggested to prevent recurrence [131,132]. However, this approach has not been sufficiently studied, so until further information is available we do not recommend routine use of glucocorticoids to prevent recurrences [133].

Surgery — Indications for urgent surgical intervention include:

- Unstable patient – In this case, initiate resuscitation, consult surgeon, and stabilize the patient before proceeding to the operating room.
- Peritonitis or intestinal perforation.
- Nonoperative reduction is completely unsuccessful. If the reduction attempt was partially successful, it may be repeated. (See 'Delayed repeat enema' above.)

Surgery is also indicated when imaging reveals a **persistent focal** filling defect, indicating a mass lesion [134]. However, not all filling defects are indications for surgery:

- If the patient has undergone successful nonoperative reduction (as indicated by relief of symptoms) and there is a residual filling defect that is consistent with an edematous ileocecal valve, the patient can be safely observed. However, repeat evaluation with ultrasound or contrast study within 12 to 24 hours is appropriate to confirm successful reduction.
- If the patient has a filling defect that appears diffuse and has a suspected or a known explanation (eg, IgAV [HSP]), the patient can be managed with repeated nonoperative reduction, provided that each attempt is successful. (See 'Recurrence' above.)

Children needing operative management require adequate intravenous fluid resuscitation and prophylactic intravenous antibiotics covering enteric flora prior to skin incision. Patients presenting with clinically significant nausea and emesis may require nasogastric tube decompression until their bowel obstruction resolves.

Contemporary operative management of childhood intussusception by pediatric surgeons uses a minimally invasive approach via laparoscopy in most cases. Laparoscopy allows for accurate diagnosis, as well as reduction of intussusception and, if necessary, bowel resection and anastomosis. Successful laparoscopic approaches to childhood intussusception have been demonstrated in several series worldwide to be safe, effective, and may be associated with a more rapid recovery of intestinal function with less need for

postoperative narcotic analgesia [135-139].

Reduction of intussusception during operation is attempted in most cases, but resection with primary anastomosis is performed if manual reduction is not possible or if a pathologic lead point is seen. The risk of recurrence is approximately 1 percent after manual reduction and virtually nonexistent after surgical resection [140].

Small bowel intussusception — Patients with intussusception limited to the small bowel are managed somewhat differently. Compared with ileocolic intussusception, small bowel intussusception is less likely to respond to nonoperative reduction [19,141,142], and more likely to reduce spontaneously (provided that the intussusceptum is short).

Patients with small bowel intussusception are managed in one of two ways, depending on the clinical circumstances:

- If a short ileo-ileal intussusception is detected incidentally on a radiographic examination in a patient with mild or no symptoms, the intussusception often resolves spontaneously, during the imaging procedure or shortly thereafter. (See 'Spontaneous reduction of intussusception' below.)

- If the patient is symptomatic or if the intussusceptum is long, nonoperative reduction may be attempted, but is rarely successful. If nonoperative reduction is unsuccessful or is not attempted, surgery is often necessary.

Intussusception is more likely to resolve spontaneously if it is limited to the small bowel (eg, ileo-ileal intussusception), and if the intussusceptum is short (<2.3 cm in length) [19]. In one series of patients with isolated small bowel intussusception, all cases in which the intussusceptum was <3.5 cm resolved spontaneously, and the children who required surgery had a mean intussusceptum length of 7.3 cm (95% CI 4.8-9.7 cm) [143]. Similar conclusions were reached by a study in adults [144].

Spontaneous reduction of intussusception — Intussusception that is detected incidentally by imaging in a patient with mild or no symptoms often resolves spontaneously (termed spontaneous reduction of intussusception [SROI]), typically during the imaging procedure or shortly thereafter. In this case, the patient does not require further evaluation or intervention if no lead point is identified [145,146]. Other radiographic criteria suggesting a transient process include normal bowel wall thickness,

no proximal dilatation, and no colonic involvement.

These cases of SROI are increasingly recognized, probably because ultrasound is frequently used for evaluation of patients with nonspecific abdominal symptoms and may detect transient intussusceptions. In one series from a single institution, 17 percent of children with an intussusception experienced SROI and approximately one-half of these were asymptomatic [145].

INFORMATION FOR PATIENTS

UpToDate offers two types of patient education materials, "The Basics" and "Beyond the Basics." The Basics patient education pieces are written in plain language, at the 5th to 6th grade reading level, and they answer the four or five key questions a patient might have about a given condition. These articles are best for patients who want a general overview and who prefer short, easy-to-read materials. Beyond the Basics patient education pieces are longer, more sophisticated, and more detailed. These articles are written at the 10th to 12th grade reading level and are best for patients who want in-depth information and are comfortable with some medical jargon.

Here are the patient education articles that are relevant to this topic. We encourage you to print or e-mail these topics to your patients. (You can also locate patient education articles on a variety of subjects by searching on "patient info" and the keyword(s) of interest.)

- Basics topic (see "Patient education: Intussusception (The Basics)")

SUMMARY AND RECOMMENDATIONS

- **Definition** – Intussusception refers to the invagination (telescoping) of a part of the intestine into itself (figure 1). It is the most common cause of intestinal obstruction in infants between 6 and 36 months of age. Ileocolic intussusception involves the ileocecal junction, and accounts for 90 percent of all cases. (See 'Epidemiology' above and 'Types and terminology' above.)

- **Causes** – Approximately 75 percent of cases of intussusception are considered "idiopathic," although some of these episodes may be triggered by viral infections.

The remaining 25 percent of cases are caused by an underlying disease or condition that creates a pathologic lead point for the intussusception, most commonly a Meckel diverticulum. (See 'Pathogenesis' above.)

- **Clinical presentation** – Intussusception typically presents with the sudden onset of intermittent, severe, crampy, progressive abdominal pain, sometimes with vomiting and grossly bloody stools, alternating with relatively pain-free periods. The classic triad of abdominal pain, palpable mass, and currant jelly stools is found in approximately 15 percent of cases. Initially, emesis is nonbilious, but it may become bilious as the obstruction progresses. As symptoms progress, lethargy often develops and may be profound. In a minority of cases, the initial presenting sign may be lethargy or altered consciousness alone, without apparent abdominal symptoms. (See 'Clinical manifestations' above.)

- **Evaluation** – Ultrasonography is the method of choice to detect intussusception. A "bull's eye" or "coiled spring" lesion is seen, representing layers of the intestine within the intestine (image 1). (See 'Ultrasonography' above.)

- **Management**

  - **Nonoperative reduction** – For symptomatic but stable patients with evidence of intussusception by imaging (ultrasound or radiographs), we recommend nonoperative reduction of the intussusception rather than surgery, unless the patient has specific indications for surgery such as bowel perforation or a strong suspicion of a pathologic lead point, as described below (**Grade 1C**). Nonoperative reduction is successful in approximately 70 percent of patients with intussusception, with somewhat lower success rates in young infants and children >5 years. When performed by expert clinicians, complications (primarily intestinal perforation) are rare. (See 'Nonoperative reduction' above.)

    Nonoperative reduction can be guided by fluoroscopy or ultrasound, and either pneumatic or hydrostatic enemas may be used (image 6). There is a slightly higher success rate with pneumatic reduction under fluoroscopy, but ultrasound-guided approaches may allow for better identification of pathologic lead points and lower exposure to radiation. The choice of technique depends primarily upon the expertise and comfort level of the radiologist. The rates of complications for either of these techniques are similar. (See 'Fluoroscopic or sonographic guidance' above

and 'Hydrostatic or pneumatic reduction' above.)

- **Recurrence** – Intussusception recurs after successful nonoperative reduction in approximately 10 percent of patients. If the patient is stable, we suggest treating recurrences with repeated nonoperative reduction rather than surgery (**Grade 2C**). Patients with one or more recurrences are more likely to have pathologic lead points. (See 'Recurrence' above.)

- **Surgery** – Surgical treatment is indicated as a primary intervention for patients with suspected intussusception who are acutely ill or have evidence of perforation. Surgery also may be appropriate when the patient is treated in a location where the radiographic facilities and expertise to perform nonoperative reduction are not readily available. Surgery is necessary for patients in whom nonoperative reduction is unsuccessful after one or more attempts, or for evaluation or resection of a focal pathologic lead point. The majority of patients may be safely and effectively treated with minimally invasive approaches. (See 'Surgery' above.)

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