# Intussusception

By John W. DiFiore
*Cleveland, Ohio*

**Intussusception is the invagination of one portion of the intestine into another and is the most common form of intestinal obstruction in infants. This report reviews the clinical presentation and diagnostic and treatment options available for intussusception. The etiologies of childhood intussusception are discussed. Details and literature review are provided on the advantages and disadvantages of ultrasonography, barium enema, air contrast enema, and surgery in the diagnosis and treatment of intussusception.**


## HISTORY

ALTHOUGH INTUSSUSCEPTION was accurately described by John Hunter in 1793,[1] Hippocrates (born circa 490 BC) recommended that treatment of "ileus" consist of connecting a bellows to the anus and inflating the bowel with air if hydrostatic reduction, possibly with the use of oil, failed.[2,3] The first successful operative reduction was performed by Jonathan Hutchinson in 1871.[4,5] Hirschsprung first described hydrostatic reduction of intussusception in 1876 and in 1905 described 107 patients successfully treated with this method.[2] In the 1920s Scandinavian radiologists reported the use of fluoroscopically controlled barium enema for reduction of intussusception.[6-8] Ravitch's standardization of techniques with barium helped nonoperative reduction gain more widespread acceptance.[9] In 1959 the use of air reduction was first reported,[10] and, more recently, thousands of patients in China have been treated successfully this way.[11]

## PATHOGENESIS

Intussusception is the invagination of one portion of the intestine into another and is the most common form of intestinal obstruction in infancy. Intussusception begins as a proximal segment of intestine, the intussusceptum, telescopes into a more distal one, the intussuscipiens. The intussusceptum is propagated distally by the action of peristalsis. This leads to compression and angulation of the mesenteric vessels to the invaginated bowel with subsequent obstruction, ischemia, and, eventually, necrosis. Necrosis begins at the apex of the intussusceptum and extends proximally.[12] Secondary pressure on the intussuscipiens may cause ischemic necrosis of its outer layer resulting in free perforation or prolapse of the intussusceptum into the peritoneal cavity.[13,14]

Greater than 80% of intussusception involves the ileocecal region with ileoileal, colocolic, and jejujojenal variations progressively less common. Eighty percent of ileocolic intussusceptions have their apex in the ascending or transverse colon.[14,15] Isolated retrograde small bowel intussusception has been described.[16] Gastroduodenal and duodenogastric invaginations have been reported as complications of gastrostomy tubes[14,17,18] and in association with gastric polyps.[19]

The wall of the terminal ileum of infants is rich in lymphoid tissue, and in approximately 90% of cases there is no obvious cause other than hyperplasia of this lymphoid tissue.[14,20] In the remaining 10%, there is a distinct pathological lead point or other underlying abnormality.[21] Under the age of 2 years, lead points occur in less than 4%; after the age of 2 years, a pathological lead point is found in one third of patients.[22,23] One half of children with recurrent intussusception will have lead points.[24] In contrast to the 10% incidence of lead points in children, adults with intussusception have pathological lead points greater than 90% of the time. Adult intussusceptions are of the small bowel variety over 75% of the time, and are associated with malignant lesions 50% of the time.[25,26]

There are many different pathological lead points for intussusception in children (Table 1). The most common lead point is Meckel's diverticulum, which can cause intussusception throughout childhood. Duplication cysts also seem to be independent of age, whereas intestinal polyps and lymphomas affect older children.[22,48,51] Small bowel tumors are rare in children but most often present with intussusception.[48] Over the age of 3 years, ileal lymphoma or lymphosarcoma should be considered.[14,51,52] In older children with cystic fibrosis, abnormally viscous bowel contents are thought to play a role in subacute ileocolic intussusception.[62] Postoperative intussusception accounts for only 1% to 2% of all cases and is associated with both abdominal and nonabdominal procedures.[30,35,69-71] Intriguing studies suggest hypergastrinemia may play a role in the pathogenesis of intussusception.[72]

There is indirect evidence that intrauterine intussusception may be related to intestinal atresia. Among premature infants, there have been 13 reported cases of perinatal intussusception. Only one of these cases was diagnosed correctly preoperatively, most being explored for bowel obstruction or presumed necrotizing enterocoli-

*From the Department of Pediatric Surgery, The Cleveland Clinic Foundation Children's Hospital, Cleveland, OH.*

*Address reprint requests to John W. DiFiore, MD, The Cleveland Clinic Foundation Children's Hospital, Department of Pediatric Surgery, 9500 Euclid Ave, Cleveland, OH 44195.*


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**Table 1. Pathological Lead Points in Pediatric Intussusception**

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| Structural |
| Meckel's diverticulum[27] |
| Ectopic gastric mucosa[28] |
| Ectopic pancreas[29] |
| Appendix (normal, appendicitis, mucocele, or appendiceal stump)[30-33] |
| Alimentary tract duplications[34] |
| Anastomotic suture line[27,35] |
| Neoplastic |
| Peutz-Jeghers hamartomatous polyps[37,38] |
| Crohn's pseudopolyps[39] |
| Lymphoid polyps[40] |
| Inflammatory fibroid polyps[41] |
| Myoepithelial hamartoma[42] |
| Lymphatic malformation[43] |
| Infantile intestinal leiomyosarcoma[44] |
| Neurofibroma (von Recklinghausen's)[45] |
| Lipoma[46] |
| Hemangioma-vascular malformations[47] |
| Small bowel carcinoid[48] |
| Adenomyoma[49] |
| Leiomyoma[50] |
| Lymphoma-lymphosarcoma[51,52] |
| Fibrosarcoma[22] |
| Metastatic tumors[53] |
| Vascular-Hemorrhagic |
| Henoch-Schonlein purpura[54] |
| Leukemia[53,55] |
| Idiopathic thrombocytopenic purpura[56] |
| Hemophilia[57,58] |
| Miscellaneous |
| Foreign body (bezoars,[59] broken glass[60]) |
| Ascaris lumbricoides[61] |
| Cystic fibrosis[62,63] |
| Celiac disease[64] |
| Oral anticoagulation therapy[65] |
| Eosinophilic granuloma[66] |
| Abdominal trauma[67] |
| Pneumatosis intestinalis[68] |

Adapted with permission.[14]

tis. Pathologically, ileal atresia and stenosis were noted in the areas of intussusception in 6 of the 13 cases. In the absence of premature delivery, these areas of nonreduced intussusception may have persisted throughout gestation, leading to small bowel atresia.[73]

## CLINICAL PRESENTATION

Classically, infants with intussusception are previously healthy and present with intermittent, severe cramping abdominal pain. Episodes are characterized by screaming, drawing up of the legs, and flexing at the waist and last anywhere from several seconds to several minutes. The time between attacks ranges from 5 to 30 minutes during which infants are generally calm with normal appearance and activity. Some infants are somnolent or lethargic between attacks. Vomiting is a very common feature, initially of undigested food, then bilious. Early in the course of the illness, stools appear normal but may eventually become dark red and mucoid resembling "currant jelly," a sign of intestinal ischemia and mucosal sloughing. Although blood in the stool (occult or gross) is quite common, significant or ongoing hemorrhage from intussusception is unusual.

A thorough review of the literature by Stringer et al[14] indicates the following mean occurrence of symptoms: abdominal pain, 83%; vomiting, 85%; blood in stool (occult or gross), 53%; abdominal mass, 63%. The classic triad of colicky, intermittent abdominal pain; vomiting; and currant jelly stool appears in only 10% to 20% of cases. Changing the triad to pain, vomiting, and gross *or* occult blood increases the average incidence of the triad to 32%.

Clinical signs with significant positive predictive value (PPV) for intussusception include right upper quadrant abdominal mass (PPV 94%), gross blood in stool (PPV 80%), blood on rectal examination (PPV 78%), the triad of intermittent abdominal pain, vomiting and right upper quadrant mass (PPV 93%), and the triad with occult or gross blood per rectum (PPV 100%).[74]

## DIAGNOSTIC STUDIES

### *Abdominal Radiograph*

Although rarely definitive, the plain abdominal film can be helpful for the diagnosis. The most specific and sensitive sign is a soft-tissue mass with 2 concentric circles of fat density, the so-called "target sign" (Fig 1).[75] Other findings include the absence of cecal gas and stool, loss of visualization of the tip of the liver, and paucity of bowel of gas, particularly in the right lower quadrant.[76,77] Overall, the plain abdominal radiograph identifies intussusception only about 50% of the time.[77,78]

### *Sonography*

The sonographic appearance of intussusception was first described in the 1980s.[79] Characteristic findings include the target sign seen on transverse section and the pseudokidney sign seen on longitudinal section (Fig 2).[80-82] This appearance makes the sonographic diagnosis or exclusion of intussusception straightforward.

In 1987, Pracos et al[82] reported 100% accuracy of sonography for the diagnosis and exclusion of intussusception in 426 children, including 145 with intussusception. A 97% to 100% accuracy rate has been verified since by others.[74,83-86] In addition to the positive predictive value of ultrasonography, large, prospective studies also have shown a 100% negative predictive value. In these studies, no patients with a negative sonogram had intussusception at enema.[74,86-88] Additional advantages of sonography include the ability to document ileo-ileal intussusception, the absence of ionizing radiation to the patient, and identification of lead points.

Although excellent data are available for the use of

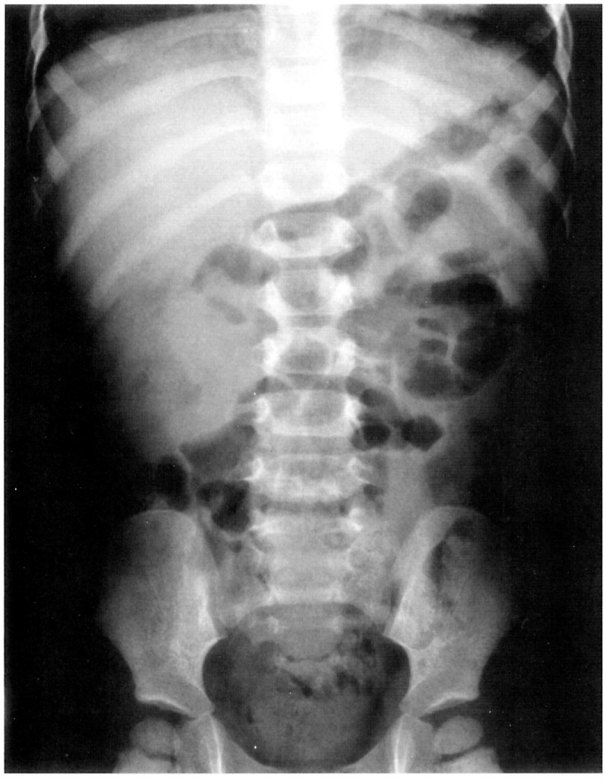

**Fig 1. Plain abdominal radiograph of intussusception. Note the soft tissue mass in the right upper quadrant with two concentric circles of fat density, the so-called "target sign."**

ultrasonography in the diagnosis and exclusion of intussusception, attempts to define sonographic criteria to predict irreducibility have been less successful. Thickness of the outer hypoechoic rim of the intussusception,[82,87] intraperitoneal fluid,[87] fluid present within the intussusception,[89] and color Doppler flow studies all have yielded equivocal results when used to predict reducibility.[83,90]

Despite the fact that the advantages of sonography have been repeatedly documented for the evaluation of patients with known or suspected intussusception,[80-87] its use has been somewhat slowly accepted throughout the world. This reflects the fact that many radiologists caring for such patients are not equally skilled with sonography.[79] There has been much discussion about the operator-dependent nature of ultrasonography and its interpretation. However, Verschelden et al[87] showed that a 100% accuracy rate could be achieved by third- and fourth-year residents who have completed a 3- to 5-month training in adult sonography. Similar accuracy rates have been documented with pediatric radiology fellows.[79] Although there is limited information in the literature confirming this, in recent years, increasing numbers of radiologists are relying solely on ultrasonography for diagnosis or exclusion of intussusception.[79]

*Diagnostic Enema*

Barium study of the colon has been the gold standard in many parts of the world until the mid 1980s when it was recognized that sonography could accurately diagnose intussusception (Fig 3). It was also around this time that the benefits of air contrast enema both for diagnosis and treatment of intussusception became evident. Many institutions still consider air enema the quickest method of excluding or confirming the presence of intussusception. It also can be more cost effective, particularly with an affirmative finding of intussusception in which the condition can be both diagnosed and treated with a single procedure.

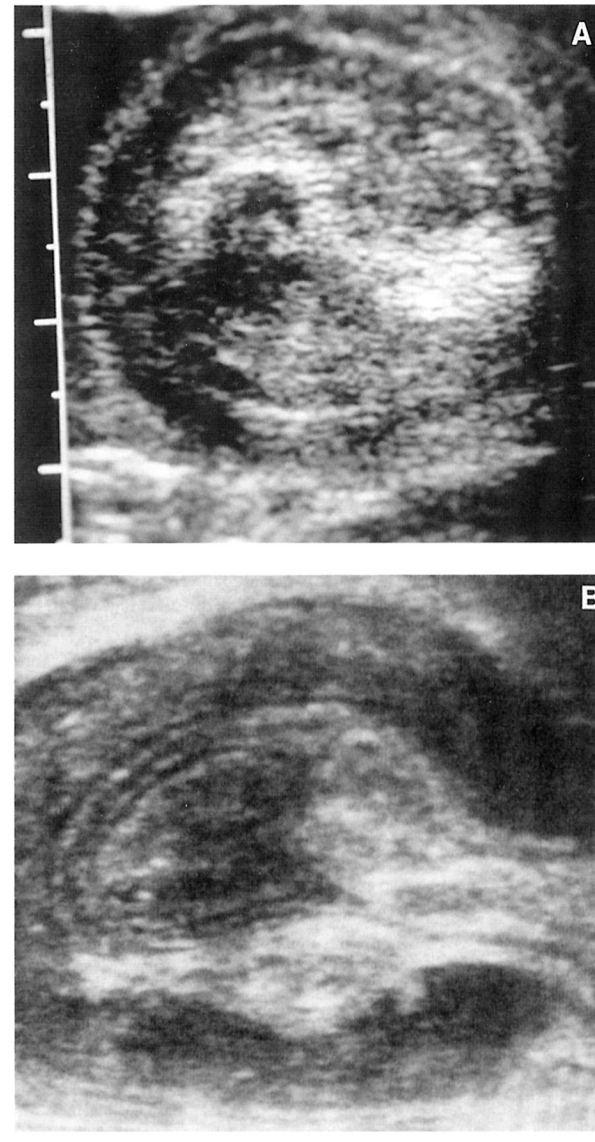


**Fig 2. Typical sonographic appearance of intussusception. (A) Transverse cross-section shows typical "target sign" with echogenic center, hypoechoic outer rim, and multiple layers of concentric rings within the intussusception. (B) Longitudinal section shows typical "pseudokidney sign."**

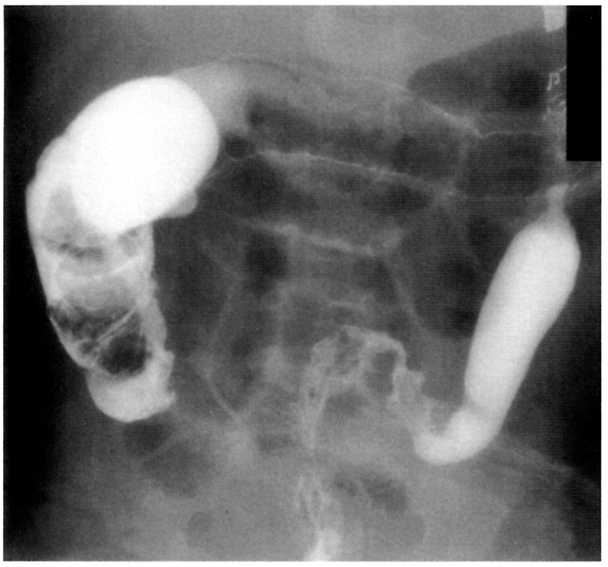

**Fig 3. Barium enema shows intussusception with apex at the hepatic flexure.**

Several reports have shown that over 50% of children suspected clinically of having an intussusception have negative results for intussusception on enema studies.[85,87,91,92] This reflects the overlap of clinical signs and symptoms in children with other abdominal conditions that may mimic intussusception. Institutions using the enema for diagnosis will, therefore, find that more than 50% of enemas on children suspected clinically of having an intussusception are negative.

Surgeons and radiologists need to consider these issues in the context of clinical suspicion for the presence of intussusception and operator skill and experience with ultrasonography versus air or barium enema. Many departments that are just beginning to use ultrasonography as a diagnostic tool for intussusception may find performing both ultrasonography and air contrast enema on all patients an excellent way to become comfortable with techniques. As skills progress, ultrasonography can become the primary diagnostic tool, reserving air contrast enema for intussusception found by ultrasound or as the initial procedure when clinical suspicion is exceptionally high.

## TREATMENT

Successful management of intussusception depends on early diagnosis, adequate resuscitation, and prompt reduction. This is best achieved through close cooperation between surgeon and radiologist.

### *Nonoperative Reduction*

It is now generally accepted that nonoperative reduction of intussusception should be attempted in most patients. Absolute contraindications to nonoperative reduction include peritonitis, perforation, or profound shock. Relative contraindications to nonoperative reduction include chronic and neonatal intussusceptions,[93] history greater than 48 hours,[36,94,95] small bowel obstruction on plain radiograph,[94,96] and children older than 2 years.[95,97] In these situations there is a lower likelihood of success, a higher incidence of pathological lead points, and an increased risk of complications. Nonoperative reduction in these circumstances should be considered in the context of the patient history and clinical condition.

The first reports of barium enema reduction of intussusception under fluoroscopic control occurred in 1927.[6-8] In the 1980s, pneumatic reduction using air as the contrast agent was reported in China with high success rates in series of greater than 6,000 patients.[11] In recent years, pneumatic reduction has become the procedure of choice.

Air contrast enema with fluoroscopic guidance has several advantages over enema reduction with barium or water-soluble contrast. Air enema is quicker, less messy, easier to perform, and therefore delivers less radiation to the patients who appear to be more comfortable during the procedure.[79,91-92,98-100] Although perforation rates for both air and barium are less than 1%, the morbidity in the event of perforation with air versus barium clearly favors pneumatic reduction.[101] The review of Stringer et al[14] of the literature from 1980 to 1991 found that reduction rates with barium or water-soluble contrast were 50% to 78% versus 75% to 94% success with pneumatic reduction. Review of more recent series (1995 through 1998) shows barium reduction success rates at 50% to 85%[102-104] and pneumatic reduction success rates at 81% to 91%.[104-107] Although these data have led to air contrast enema being the current procedure of choice for nonoperative reduction of intussusception, to date there is still no large, prospective study comparing the use and success of pneumatic and barium or water soluble enemas under fluoroscopic guidance.[79]

A newer technique involves the hydrostatic reduction of intussusception using sonographically guided saline enema. In this technique, saline enemas are administered with real-time visualization of the intussusception by ultrasound scan. The sonographic criteria for successful reduction are disappearance of the intussusception and visualization of the passage of fluid and air bubbles from the cecum into the terminal ileum. Recently, success rates from 73% to 91% have been reported in China, Korea, and Germany.[103,108-111] The advantages of this technique are avoidance of radiation exposure and more detailed visualization of the intussusception.

Although standard practice remains immediate surgery if nonoperative reduction is unsuccessful, there are several reports describing delayed repeat enemas when initial reduction attempts are not successful. In 1994, Saxton et al[112] recorded an over 50% success rate in 22

patients in whom a repeat air enema was performed 30 minutes or more after an initial unsuccessful attempt. Gorenstein et al[105] reported an increase in success rate with air contrast enema from 70% to 91% by using several attempts 45 to 60 minutes apart. Although the author does not advocate these approaches, repeat delayed attempts may improve outcome for a select group of patients and may be reasonable under very close surgical observation and examination during the waiting periods. This will require documentation of success in much larger series of children.

*Operative Reduction*

Operative reduction is necessary when radiological techniques are contraindicated or have failed, when a pathological lead point is suspected, or in the case of multiple recurrences (Fig 4). The operative approach is generally through a right-sided transverse incision slightly above or below the umbilicus. Gentle, steady pressure at the apex of the intussusception usually will achieve reduction, after which, bowel is allowed to reperfuse and is then inspected for viability. Necrotic areas, persistently ischemic areas, or intussuscepted areas that cannot be reduced require prompt resection and primary anastomosis.[14]

Many surgeons perform appendectomy to avoid future confusion regarding its presence. With recurrence rates for operative reduction only to 1% to 4%, appendectomy should not pose a significant risk with respect to the need for reduction enema in acute postoperative reintussusception. In the event that acute postoperative intussusception does occur, successful barium enema reduction of intussusception without disruption of the appendiceal stump has been reported as early as 6 days status post open reduction of intussusception with appendectomy.[113]

**Fig 4. Intraoperative view of intussusception.**

## OUTCOME

The overall mortality rate of intussusception is less than 1%.[14] The risk of postoperative adhesive small bowel obstruction after operative reduction is 3% to 6% and has not been reported after nonoperative reduction. Recurrence after nonoperative reduction is 5% to 10% and 1% to 4% after surgical reduction. Most recurrences develop within 1 to 36 months after reduction with a peak incidence during the eighth month. In patients with multiple recurrences, structural abnormalities of the ileocecal area are common, most often florid lymphoid hyperplasia of the terminal ileum. These will occasionally require ileocecal resection.

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