Intussusception in Children

Rajendra K Ghrilaharey*
Department of Pediatric Surgery, Gandhi Medical College and Associated, Kamla Nehru and Hamidia Hospitals, India.

*Corresponding author: Rajendra K Ghrilaharey, Department of Pediatric Surgery, Gandhi Medical College and Associated, Kamla Nehru and Hamidia Hospitals, Bhopal, Madhya Pradesh 462001, India, Email: drrajendrak1@rediffmail.com

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ABSTRACT

The word “intussusception” is derived from the Latin words “intus” (within) and “suscipere” (to receive). Intussusception is the invagination of one part of the intestine into another. Intussusception remains a common cause of bowel obstruction in infants and young children and results in significant morbidity and mortality, if not promptly treated. Most commonly it occurs in infants and young children, but it also occurs in adults. More than 75% of intussusceptions occur within the first two years of life, and 40 - 90% of them are seen within 12 months of the age. The classic quadrant of symptoms (abdominal pain and vomiting), and signs (abdominal mass and rectal bleeding) are seen in 30-70% of the cases, while the classic triad of vomiting, passage of blood through rectum and abdominal pain are seen in 30-60% of the cases of childhood intussusception, and more than 85% of intussusceptions are of ileocolic. The primary type (idiopathic) intussusceptions are reported to occur in 95% of the cases, and mostly seen during infancy. Secondary intussusceptions are reported to occur in 1.5 to 14% of the cases and mostly reported in older children and tends to increase with the age. Ultrasonography of the abdomen is a reliable screening tool for the intussusception in infants and children with accuracy for the diagnosis of up-to 100%, and at many centres it is a primary investigation of choice. Currently, children with intussusception are managed by attempted non-surgical enema reduction under sonographic or fluoroscopic control, and laparotomy with reduction or resection is reserved for those in which peritonitis or when radiological reduction (hydrostatic or pneumatic) has
failed. Non-surgical, radiological reduction of intussusception is successful in up to 90%, but this technique is employed mostly in developed countries. In the developing countries, due to delayed presentation and presence of bowel gangrene, laparotomy is employed in majority as the primary option for managing infants and children with intussusception.

**Keywords:** Children; Infants: Intussusceptions; Intestinal obstruction; Laparotomy; Meckel’s diverticulum; Non-surgical reduction; Pathological lead points; Surgery; Ultrasonography

**BRIEF HISTORY**

Intussusception is derived from the Latin words “intus” (within) and “suscipere” (to receive). Intussusception is the invagination of one part of the intestine into another [1]. Hippocrates, 5th century B.C. (460–377 BC), Greek physician and surgeon, the father of medicine, is often credited with the first recorded description of intussusception, but it is uncertain whether he identified this condition as a distinct clinical entity from other forms of ‘ileus’ [2]. Intestinal invaginations or intussusception was first recorded by Paul Barbette of Amsterdam in 1676, who also suggested the possibility of operative reduction of intussusception [2]. Johann Peyer’s also described about intussusception and this can be found in Johann Peyer’s monograph of 1677, within which he differentiated intussusception from volvulus of the small intestine and also described the lymphoid follicles of the terminal ileum which bear his name [2]. Cornelius Velse first successfully operated in adult for intussusception, which he reported in 1742 [2]. John Hunter (1728-1793) clearly described the pathological features of intussusception, while suggesting possible mechanisms by which the disease occurs, and he also observed that intussusception occur most frequently in the first fifteen years of life, not occurring so frequently in older people. In 1789, Hunter presented a case of intussusception in a 9-month-old child who subsequently died of his diseases; the report of this account was published in 1793 [2]. In 1836, Samuel Mitchell reported the first cases of successful air enema reduction in childhood intussusception, which was published in 1838. Wilson in 1831 first operatively reduced an intussusception in an adult, and 40 years later in 1871, Hutchinson reported it in an infant [1]. On November 11, 1873, Sir Jonathan Hutchinson read a paper before the Royal Medical and Chirurgical Society, reporting the first successful laparotomy and reduction of an ileocolic intussusception performed under chloroform anaesthesia in 1871 on a 2-years-old girl at the London Hospital, after failed attempts at hydrostatic enema reduction, and the same was published in 1874 [2-4]. In 1876, Harald Hirschsprung’s published the first series on hydrostatic reduction of intussusception, in which he described his experience with controlled hydrostatic pressure reduction for intussusception. By 1905, Hirschsprung’s had collected 107 cases of intussusception with a mortality of only 35%, establishing the enema treatment of intussusception used in the current time [1,2,4]. In 1913, Ladd first reported the use of diagnostic imaging with bismuth enemas and he published the first photographs of roentgenologic pictures of an intussusception, but he didn’t recognize its therapeutic values that time [1,4]. By the 1958, non-operative reduction of intussusception up to 75% was achieved with a mortality rate close to zero [1]. Currently, children with intussusception
are managed by attempted enema reduction (hydrostatic or pneumatic), under sonography or fluoroscopic control, and laparotomy with reduction or resection is reserved for those in which peritonitis or when radiological reduction (hydrostatic or pneumatic) has failed [1,2,4].

**DEMOGRAPHICS OF INTUSSUSCEPTION**

Intussusception remains a common cause of bowel obstruction in infants and young children. Intussusception has been reported to occur in 1-4 in 2000 infants and children. Most of the series on intussusception reported its occurrence more in males than in females, usually at a ratio of 1.5 to 3.6:1. Approximately 75% of intussusceptions occur within the first two years of life, and 40-90% of them are seen within 12 months of the age. Intussusception occurring in utero may lead to intestinal atresia, most commonly ileal atresia and rarely jejunal atresia. Intussusception has also been reported in families and relatives. Intussusception is reported to occur in greater numbers in Caucasian infants and children. The incidence of intussusception tends to vary by season and from one study to another study, and even within the same study, but this seasonal variation may not occur always. Seasonal variation associated with intussusceptions usually correlates with viral infections (respiratory, gastrointestinal, or both), with most of the cases seen in May, June, and July. Takeuchi, et al (2012) in a largest survey of childhood intussusception in Asia concluded that there were an estimated 2,000 cases of infantile intussusception occurring annually in Japan, with an incidence of 180-190 cases per 100,000 infants. They further highlighted that this incidence of intussusception in Japanese infants is higher than those reported by other countries [1,5-18].

**CAUSES OF INTUSSUSCEPTION**

Intussusceptions may be categorised as primary type (idiopathic) where there is no pathological lead points (PLPs) and secondary type where PLPs are present.

**Primary (Idiopathic) Intussusception**

More than 95% of childhood intussusceptions are idiopathic where there is no identifiable cause for it. In facts, in idiopathic intussusceptions also there is thickening of the Peyer’s patches and non-specific mesenteric lymphadenitis (Figure 1a) are observed in majority of the cases, and could be a possible causative factor for the occurrence of primary (idiopathic) intussusception [1,9-11,16].
Secondary Intussusception

Secondary intussusceptions in which pathological lead points are present, and reported to occur in 1.5 to 14% of the cases and mostly reported in older children and tend to increase with the age. Secondary intussusception caused by PLP is mostly reported as case reports in the literature. The PLPs leading to intussusceptions in children are either intraluminal or intramural pathologies and are Meckel’s diverticulum (Figure 1b), ileal polyp (Figure 1c, Figure 1d), appendicular stump, tumors, Henoch-Schonlein purpura, duplication of gut, etc [1,5,7-9,11,16,19-21].

Figure 1a: Idiopathic intussusception showing enlarged Peyer’s patches and mesenteric lymph nodes.

Figure 1b: Meckel’s diverticulum as a pathological lead point for intussusception (operatively reduced).
Post-Operative Intussusception

Post-operative intussusception may present as an acute intestinal obstruction and it occurs mostly following laparotomy for intestinal conditions, but may also occur following surgery done outside the abdomen. Post-operative intussusception may be single or multiple, and mostly involves small intestine, and rarely involves a colon as colocolic intussusception [1].

Rotavirus Vaccination and Intussusception

Rotavirus vaccination has been shown to small increase in the occurrence of intussusception
in infants. Studies revealed that infants with a history of rotavirus vaccination are at some more risk for intussusception [1,12,14,22,23].

**Anatomical Types of Intussusception**

Intussusception may be categorised depending upon the part of intestine that involved in the intussusceptions, and in order of frequency are; ileocolic (85%), ileoileocolic (10%), colocolic (2.5%), jejunojejunal / ileoileal (2.5%). This above incidence varies from series to series, but majority of the series observed ileocolic (Figure 2a) is the commonest type of intussusception, followed by colocolic and jejunojejunal or ileoileal (Figure 2b) [1,8-10].

![Figure 2a: Operative photograph showing ileocolic intussusception.](image)

![Figure 2b: Operative photograph showing ileoileal intussusception.](image)
Other Types of Intussusception

The recurrence of intussusceptions (recurrent intussusception) may occur following the enema reduction or following the operative manual reduction of intussusception. Retrograde intussusception and multiple intussusceptions have also been reported in child [1,5,9].

CLINICAL PRESENTATION

Children with intussusception have been described as being generally healthy, sturdy, well developed, and well nourished, supporting Hirschsprung’s classic statement, that “I never saw a malnourished child with an intussusception.

Abdominal Pain

The most common classic symptom of intussusception in children is a sudden onset of severe, colicky, and intermittent abdominal pain. This abdominal pain last only for few minutes and reported to occur in up to 85% or more, but absence of abdominal pain does not rule out intussusception [1,5,7,9,12,13].

Vomiting

The vomiting is the second common symptoms of childhood intussusception. In some series vomiting is more frequent while in other series abdominal pain is more frequently observed symptoms. Vomiting is reported to occur in 40-90% of the cases, but its absence does not rule out intussusception [1,5,8,10,12,13].

Abdominal Lump

In intussusception, abdominal mass is due to the invagination of one part of the intestine to another and this lump is mostly palpable in the right hypochondrium and epigastric region and reported to occur in 40-75% of the cases. In some delayed cases, abdominal lump at right hypochondrium may not be there (Signe de Dance), and may be palpable on rectal examination, and seen in 5-7.5% of the cases. In the presence of other classic symptoms and signs of intussusception; absence of palpable abdominal lump does not rule out intussusception [1,7,9,10,12].

Rectal Bleeding

Rectal bleeding is also an important sign of infantile intussusception and reported to occur in 40-95% of the cases. This rectal bleeding in intussusception is characterised as “currant jelly appearance”, but its absence does not rule out intussusception. This currant jelly appearance of blood may also be observed during rectal digital examination. In some of the delayed cases, the intussusception may also prolapse through the rectum or palpable on rectal digital examination [1,8-10,12,13].

The classic quadrant; all four of classic symptoms (abdominal pain and vomiting) and signs (abdominal mass and rectal bleeding) are seen in 30-70% of the cases. The classic triads of
vomiting, passage of blood through rectum and abdominal pain are seen in 30-60% of the cases of intussusception. Presence of diarrhoea and absence of vomiting, abdominal pain, abdominal lump or rectal bleeding does not rule out intussusception. Delayed cases may present with features of shock, and septicaemia due to gangrenous changes in bowel, and mostly observed in developing countries [1,7,9,11].

**DIAGNOSIS AND INVESTIGATION**

Most of the time, the diagnosis of intussusception in infants and children is usually suspected clinically, but the accuracy of clinical diagnosis is only in 50% of the cases, and therefore the clinical diagnosis is needing confirmation on radiological investigations with x-ray and USG of the abdomen.

**Plain Roentgenogram of the Abdomen**

The radiographic signs of intussusception are target sign, crescent sign, absent liver edge sign (also called absence of the subhepatic angle), and features of bowel obstruction. It is possible to suspect intussusception on plain radiography of the abdomen in approximately 50% of the cases. Target sign is a soft tissues mass effect locating at right side of the upper abdomen, and absences of caecal gas may also be there. Sometimes does not have a target appearance and just resembles a solid mass. It is sometime also called as pseudo-kidney sign, because it may resemble the shape of a kidney mass in the right upper abdomen. The crescent sign is caused by the intussusceptum protruding into a gas filled pocket / bowel, which often results in a crescent shaped gas pocket, but this crescent shape is not always present or identified. Plain x-ray of the abdomen in standing position may also provide the multiple air fluid levels, the features of intestinal obstruction. In delayed cases of intussusception, which had bowel ischemia and perforation and the plain x-ray of the abdomen only show the gas under the dome of the diaphragm, and the other features are masked [1,9,11-13,24,25].

**Ultrasonography of the Abdomen**

At many centres a primary investigation for childhood intussusception is USG of the abdomen, and it is a reliable screening tool for the diagnosis of intussusception with accuracy up-to 100%. Ultrasound signs for intussusceptions include, target sign (also known as the doughnut sign), pseudokidney sign, and crescent in a doughnut sign. (a) Target sign (doughnut sign) on USG: When bowel telescopes into itself, concentric rings are seen creating the target sign. The characteristic appearance is not only due to the three bowel walls paralleling each other, but is also due to each wall itself demonstrating the normal striation of echogenic mucosa, hypoechoic submucosa and echogenic muscularis. This is mostly seen as 3-5 cm mass, mostly in the right upper quadrant of the abdomen. (b) Crescent sign: If a large portion of mesenteric fat becomes incorporated within the intussusception then an additional echogenic crescent may be seen creating the crescent within a doughnut sign. In transverse section concentric rings of tissue representing components of the bowel wall and mesenteric fat are seen, sometimes referred to as the doughnut
or target sign. (c) Pseudokidney sign: the combination of intussusception and adjacent enlarged mesenteric lymph nodes may simulate the appearance of a kidney creating the pseudokidney sign. In longitudinal section the mass is roughly ovoid in shape, with different tissues appearing layered longitudinally. USG of the abdomen also helps in documentation of PLPs, if present. On USG, if a complex mass is present in the centre of the intussusception in both transverse and longitudinal sections, there is a possibility of a presence of a lead point / secondary cause for intussusception. Doppler sonography may be able to interpret the viability of the bowel and help in the planning of the management of intussusception. The sonography has added advantage of that it is portable, non-invasive technique, no radiation, and can be repeated without any hazards, and the procedures can also be performed at bedside [1,11-13,24,26].

**Enema Studies**

Barium enema of the colon was considered as a gold standard for the diagnosis and exclusion of the intussusception in children, and it was the choice of investigations till 1980, prior to the use of the USG. Contrast enema has 90-100% accuracy and also has therapeutic value. By doing enema studies, a diagnosis and treatment of the intussusception can be done in one procedure. Its only disadvantage is that is an invasive procedure, require sedation and also involve radiation and may miss the PLPs. Clinical signs of shock, features of peritonitis or bowel perforation are the absolute contra-indications for doing air or contrast enema, not only for diagnostic, but also for therapeutic use [1,5,6,8,26].

Other investigations like small bowel follow through, computed tomography, magnetic resonance imaging are rarely used for making the diagnosis of intussusception in clinical practice.

**MANAGEMENT**

In general, the treatment options for the intussusception are non-surgical radiological reduction (hydrostatic, or pneumatic reduction), exploratory laparotomy (manual reduction, segmental bowel resection for gangrene and anastomosis, hemicolecotomy, creation of stoma), and laparoscopic procedures.

**Non-Surgical Reduction of Intussusceptions**

A major advancement that occurred for the management of intussusception is more and more use of non-operative (non-surgical reduction) radiological reduction. Non-surgical reduction involves an initial confirmation of the diagnosis of intussusception, either with USG or barium enema (excluding peritonitis and gangrene of the intestine) and followed by the radiological reduction under image guidance. Radiological reduction includes; hydrostatic reduction (barium enema, saline enema, other enema) or pneumatic (air) reduction and can be done under fluoroscopic or sonographic guidance. Radiological reduction can be done as either of one: (1) hydrostatic reduction with fluoroscopic guidance, (2) hydrostatic reduction with USG guidance, (3) pneumatic reduction with fluoroscopic guidance, and (4) pneumatic reduction with
USG guidance. It is always needed to select the cases for radiological reduction, which do not need urgent surgical intervention. Contra-indication for radiological reduction are; presence of evidence of peritonitis, features suggestive / presence of bowel gangrene, intestinal perforation and all above warrants an urgent surgical intervention. Non-operative management has been shown to shorten hospitalisation and reduce morbidity and mortality with a success rate of up to 90% or more. This radiological reduction of intussusception is more frequently employed in developed countries, while in developing countries more and more cases needs laparotomy as primary option for treating infants and children of intussusception, due to delayed presentation and presence of bowel gangrene [1,5,6,8,10,12,16,24,27-31].

**Hydrostatic Reduction of Intussusception**

Hydrostatic reduction under the guidance of fluoroscope or USG using barium enema / saline enema or other contrast enema. Barium enema reduction was the gold standard for the diagnosis and radiological reduction of intussusception in the past. This technique is simple, safe and familiar to every radiologist. The success rate of barium reduction of intussusception is reported to occur in 46-93%. The disadvantage is that it requires sedation and radiation, and if bowel perforation occurs during the procedure, it may produce secondary peritonitis [1,5,6,8,16,24,27]. In a survey conducted by Stein-Waxler R, et al (2014) revealed that amongst 456 pediatric radiologists reported 3834 attempted radiological reduction in the preceding 12 months, and concluded that 96% used fluoroscopy and only 4% used sonography for guiding the reduction of intussusception. They further concluded that 78% used air, 20% preferred fluid for reduction, and 64% of them also repeated the reduction after a waiting of 2 hrs or more with a success rate of 20% [28].

**Pneumatic Reduction of Intussusception**

Pneumatic reduction using air for intussusception may be done under fluoroscopic or sonographic guidance. USG guided reduction of intussusception has an added advantage of non-radiation and easy to perform and can be done with equal results, and if any bowel perforation occur during the procedure it is with less of peritonitis. Many of authors also reports that trans-abdominal manipulation of intussusception improve the reduction rate [1,8,12,27-31]. An initial location of intussusception in the descending colon / rectum, presence of peritoneal fluid, paracentasis containing fluids, and bloody stools are the most important risk factors for failure of the USG guided reduction [31].

**Repeat Non-Surgical Reduction**

Repeat or delayed repeat reduction of intussusception is a usual practice. Repeating the non-operative radiological reduction procedures after a wait of 2 hrs or more after first attempt and reported a success rate of 20-60% [1,8,10,28].
Post Reduction Management

The non-surgical radiological reduction is preferably done as an indoor procedure. The post reduction management consist of nil orally, I/V fluids, naso-gastric aspiration, injectable antibiotics, etc and all depends upon pre reduction condition of a child. A repeat abdominal sonography is also advisable, if any doubt exist.

SURGICAL MANAGEMENT OF INTUSSUSCEPTION

In the developed countries more and more of the cases of intussusception in infants and children are managed with non-surgical radiological reduction, and only 10% of the cases require surgical intervention. In the developing countries due to various reasons, the primary option for managing the intussusception is exploratory laparotomy, and required in 50-90% of the cases. The indication of surgical intervention for infantile and childhood intussusception are; failed non-surgical reduction, presence of bowel gangrene (Figure 3a, and Figure 3b), bowel perforation (Figure 3c), peritonitis, presence of PLP for intussusception, and bowel perforation following radiological reduction. Surgical management consists of exploratory laparotomy and either of one; operative manual reduction, operative manual reduction and repair of serosal tear (Figure 3d), repair of bowel perforation occurred following manual reduction or following non-surgical reduction, segmental bowel resection for gangrene and ileoileal anastomosis, hemicolecotomy, and resection of gangrenous bowel (Figure 3e) and creation of stoma. It is always better to check for the viability of bowel following operative reduction of intussusception, and if the viability of bowel is in doubt, resects it. If a PLP (mostly inverted Meckel’s diverticulum or ileal polyp) detected during investigation on USG or barium enema or during laparotomy for intussusception, it also need to deal accordingly. The interval between the onset of symptoms and institution of the treatment is of paramount importance. It is therefore very important for the treating medical personnel to diagnose the case of intussusception in its early phase and should institute an effective therapy (medical/surgical) to reduce the morbidity and mortality. The delay in the diagnosis of intussusception leads to increased chances of bowel gangrene and the need for bowel resection and thereby increasing the morbidity and mortality as well. Twenty five to 80% cases require bowel resection for bowel gangrene associated with intussusception in children in the developing countries [1,5-12,14,19,20,29,32].
Figure 3a: Operative photograph, showing gangrenous bowel.

Figure 3b: Operative photograph, showing ileocolic intussusception with gangrenous ileal.

Figure 3c: Operative photograph, showing ileocolic intussusception with ileal perforation.
Figure 3d: Operative photograph, showing serosal tear of bowel after manual reduction of intussusception.

Figure 3e: Operative photograph, showing resected gangrenous bowel, needed an ileostomy.

LAPAROSCOPIC MANAGEMENT OF INTUSSUSCEPTION

Intussusception can also be safely and effectively treated laparoscopically as an alternative to laparotomy. Laparoscopic procedures for childhood intussusceptions is not only safe and an effective, but also have an added advantage of minimal access surgery as fewer scars, less post-operative pain and shorter length of hospital stay. During the laparoscopic procedures for intussusception, conversion to open surgery is also requiring for various reasons. A systematic review (conducted by Apelt N, et al in 2013) of all publications on the laparoscopic treatment of pediatric intussusceptions from Jan 1990 to April 2012 was performed and found 276 cases of laparoscopically reduced intussusception. They concluded that the success rate of above procedures was 71%, one had intra-operative iatrogenic intestinal perforation, 2.9% post-operative complications with as recurrence of 3.6% [33-36].
COMPLICATIONS FOLLOWING NON-SURGICAL REDUCTION

Major complication following non-surgical reduction of intussusception is a bowel perforation and reported to occur in less than 1% [1].

COMPLICATIONS FOLLOWING LAPAROTOMY AND LAPAROSCOPIC PROCEDURES

Complications are known to occur with exploratory laparotomy done in children for intussusception and are more in the cases needed bowel resection and with gangrene and peritonitis. Most of the complications are wound dehiscence, anastomotic leak, faecal fistula, burst abdomen, peritoneal abscess, post-operative bowel obstruction, incision hernia [1,5,7-9,11,32-34,36].

ADULT INTUSSUSCEPTION

Although, intussusception is a leading cause of intestinal obstruction in infants and children, but it also rarely occurs in adults. Adult intussusception is differ from childhood intussusception that it is rare in adults accounting only in 1-5% of all the cases of intestinal obstructions, and more than 90% of them are secondary to the PLPs. The most commonly seen PLPs (intramural, intraluminal or extramural) that causing intussusception in adults are polyps, Meckel's diverticulum, buried appendicectomy stump, benign and malignant tumors. The clinical presentation in adult intussusception is with acute, chronic or sub-acute intestinal obstruction and majority may not have classic quadrant or classic triad of intussusception that are seen in childhood intussusceptions. Adult intussusceptions are investigated with USG of the abdomen and also requiring CT scan of the abdomen in many of the cases. The accuracy of USG of the abdomen for diagnosing the intussusception is inferior to that of childhood intussusceptions. CT scan of the abdomen provides better results for detecting the intussusception in adults. Majority of the adult intussusception require surgical management due to the presence of PLPs, and executed either through laparotomy or with laparoscopic procedures [37-40].

MORTALITY

There is study decline in the mortality relating to the intussusception in infants and children. This decline in mortality is more obvious and mostly in the developed countries and is directly attributing to the early diagnosis and more use of non-surgical radiological reduction. In the developed countries; the mortality relating to non-surgical reduction of intussusception is reported in less than 1%, and the mortality following surgical treatment of childhood intussusception is also reported in less than 1%. In the developing countries at many centres, more than 50% of the infants and children presenting late and with bowel gangrene and requiring exploratory laparotomy as a primary option for managing these cases and also needs bowel resection in more than half of the cases. Due to above reasons, in developing countries at many centres the mortality relating to intussusception are occurring in 1.5-15% [1,7-9,11,41-43].
SUMMARY

Intussusception is one of the most common causes of bowel obstruction in infants and children, but also rarely occurs in adults. Approximately 75% of the childhood intussusception occurs within two-years of the age. More than 95% of intussusception in infants and children are idiopathic and secondary intussusception are reported to occur in 1.5 -14% of the cases. It is clinically suspected on the presence of classic symptoms and signs. It is possible to document and diagnoses childhood intussusception with accuracy of up-to 100%. In developed countries it is managed with non-surgical radiological reduction with a success of 90% and only 10% needing surgical intervention. In the developing countries due to delayed presentation and presence of bowel gangrene, more than 50% childhood intussusceptions needing laparotomy as a primary option for management and also require more of bowel resection. Due to above reason the mortality relating to intussusceptions is less than 1% in developed countries while it is 1-15% in developing countries.

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