

Intussusception

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Abstract

Intussusception is the most common cause of intestinal obstruction in infancy and early childhood. It occurs when one segment of bowel (the intussusceptum) invaginates into an adjacent distal segment of bowel (the intussuscepien). The classic presentation is with intermittent abdominal pain, vomiting and redcurrant jelly-like stool. Diagnosis can be accurately confirmed with an ultrasound scan. Initial management is with fluid resuscitation and antibiotics. Following adequate resuscitation, treatment is usually with a non-operative air enema reduction under fluoroscopic guidance. If this fails to completely reduce the intussusception, the air enema may be repeated in patients that are clinically stable. The main risks associated with an air enema are bowel perforation, failed reduction and recurrence. Surgical intervention is indicated in patients presenting with perforation, those that are clinically unstable or where multiple air enemas have failed to reduce the intussusception. Surgery can be performed open or laparoscopic and involves attempted manual reduction of the intussusception and may require bowel resection and anastomosis.

Keywords Air enema reduction; bowel obstruction; bowel resection; intussusception; perforation; recurrent jelly stool

Introduction

Intussusception is defined as the invagination of one segment of bowel (the intussusceptum) into an adjacent distal segment of bowel (the intussuscepien).

This subsequently leads to intestinal obstruction and intussusception is the most common cause of bowel obstruction in early childhood.

The incidence of intussusception in the UK is approximately 25 per 100,000 live births.¹ However, this figure has been declining over the last 50 years. There are also significant geographic variations in incidence. Intussusception most commonly affects children between 3 months and 3 years of age, with a peak incidence at 5–6 months. Male children are more likely to be affected with a ratio of 2:1.

Pathogenesis

Intussusception occurs when peristalsis projects a proximal segment of bowel into an adjacent distal segment. With ongoing antegrade peristalsis of the bowel this intussusceptum can be

pushed further and further into the distal bowel. This results in compression of the mesenteric vessels and lymphatics causing venous congestion and tissue oedema. It is this that leads to secretion of mucus and bleeding from the bowel giving the characteristic 'redcurrant jelly-like' stool. Ultimately this will lead to bowel ischaemia and necrosis, usually affecting the intussusceptum first, followed by the intussuscepiens at a later stage. Alongside this, the oedematous intussusceptum results in intraluminal bowel obstruction. As the bowel becomes compromised, bacterial translocation occurs and significant third space fluid losses, culminating in sepsis and hypovolaemia. Consequently these patients may present in septic and hypovolaemic shock.

The aetiology of intussusception is unknown in the majority of cases and is, therefore, labelled primary idiopathic intussusception. The widely accepted hypothesis is that the intussusception is triggered by inflammation of the lymphoid tissue in the bowel wall (the Peyer's patches) and enlargement of the local mesenteric lymph nodes, and that these act as the 'lead point' of the intussusceptum. Many common viral infections are associated with intussusception, including adenovirus, rotavirus and human herpes simplex virus. Viral infection in children can induce significant lymphoid hyperplasia.

More recently, intussusception has been linked with the rotavirus vaccination. The rotavirus vaccination, Rotarix[®], was added to the UK national childhood immunization programme in July 2013 following the WHO recommendation for global implementation after analysis of the risks and benefits associated with the vaccine. It is estimated that Rotarix[®] results in 35 additional cases of intussusception each year. However, it will prevent 13,000 rotavirus admissions and many deaths from rotavirus gastroenteritis. The vaccination should be given before 3 months of age to avoid the peak age for intussusception.² It is important that clinicians are aware of this small increased risk in the first 7 days post-vaccination and refer infants promptly for assessment should suggestive symptoms arise.

Secondary intussusception occurs when there is the presence of a pathological 'lead point'. These are lesions within the bowel lumen or attached to the bowel that project into the intestinal lumen and can therefore act as a 'lead point' for the intussusceptum. The reported incidence of a pathological lead point in intussusception is between 2% and 10%. There are a variety of structures and conditions that can act as pathological 'lead points' (Box 1). These are usually of gastrointestinal origin but can be related to a systemic disease process. The presence of an underlying condition may predispose patients to having intussusception resulting from a pathological lead point, such as Peutz–Jeghers Syndrome. Patients outside the typical idiopathic intussusception age range, and those with recurrent intussusceptions should raise suspicion of the presence of a pathological 'lead point' and may require further investigation. The incidence of a pathological lead point increases with age and in children over 4 years old, 57% of intussusceptions will have a pathological lead point. Although less likely, a typical presentation of intussusception with successful air enema reduction does not exclude the presence of a pathological lead point. Indeed, about 5% of spontaneously reduced cases will still have a pathological lead point.

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Pathological lead points

- Meckel's diverticulum
- Appendix
- Intestinal polyps (e.g. Peutz–Jeghers syndrome)
- Duplication cyst
- Neoplasm (e.g. carcinoid tumour, lymphoma)
- Foreign bodies including feeding tubes
- Intra-luminal inspissated stool in cystic fibrosis
- Intra-mural haematoma in Henoch–Schonlein purpura
- Coeliac disease

Box 1

The most common anatomical site for intussusception is ileocolic. This accounts for more than 80% of idiopathic intussusception. The ileum advances into the colon and usually reaches the ascending or transverse colon. However, it can sometimes pass all the way through to the anus. A pathological lead point is uncommon in ileo-colic intussusception regardless of age. Intussusception can also occur at other sites; ileo-ileal, ileo-ileo-colic, and colo-colic, and these sites are more commonly associated with a pathological lead point.

Ileo-ileal intussusception can be a transient phenomenon that resolves spontaneously. Indeed, sometimes when one is observing a lengthy laparotomy, self-resolving intussusceptions are observed. They can also be observed during ultrasound scans (USS). Approximately half of these patients will have symptoms while the intussusception is present.

Clinical presentation

The classical presentation of an infant with intussusception is with intermittent episodes of colicky abdominal pain associated with drawing up of legs, crying and inconsolable. The episode usually last a few minutes and occur in 15–20 minute intervals. In between episodes the infant is often pain free and quiet. Vomiting is common and initially this is non-bilious. Subsequently, the child can become listless and lethargic. The vomiting progresses to bilious vomiting with abdominal distension. Blood and mucus can be passed in the stools and this is classically described as 'redcurrant jelly' stools. In the history there is often a recent preceding upper respiratory tract infection or gastroenteritis.

On examination, these children are often pale and profoundly dehydrated. They can be febrile with signs of shock. On palpation of the abdomen a sausage-shaped mass may be felt anywhere, although most commonly in the right upper quadrant, when the child is relaxed. The right lower quadrant may be unusually flat, known as Dance's sign. This occurs as the caecum is collapsed distal to the obstruction. The abdomen may be distended and signs of peritonism, such as involuntary guarding, may suggest bowel perforation. Rectal examination may reveal blood stained stool. Occasionally a rectal mass may be palpable or even visible protruding through the anus.

The classical triad of episodes of drawing up legs and inconsolable crying due to abdominal colic, along with redcurrant jelly stool and a palpable mass on abdominal examination is reported in 20–60% of cases.³

Diagnosis

Differential diagnoses to consider include other causes of intestinal obstruction, such as irreducible inguinal hernia, volvulus or adhesional small bowel obstruction, peritonitis, acute appendicitis and other medical conditions including gastroenteritis, meningitis and sepsis.

Classical presenting features may be absent and therefore a high index of suspicion is needed to identify the diagnosis of intussusception. Abdominal ultrasound (USS) is the first-line investigation in order to confirm intussusception. When performed by a trained practitioner it has high sensitivity (>90%) and specificity (>90%).³ Features on USS include the 'target sign', which represents the intussusception visualized on transverse section and the 'pseudo kidney sign', when it is seen on longitudinal section (Figure 1). A pathological lead may be identified but can often be missed on USS.

Contrast enema under fluoroscopic imaging guidance used to be the gold standard for diagnosis of intussusception. However,

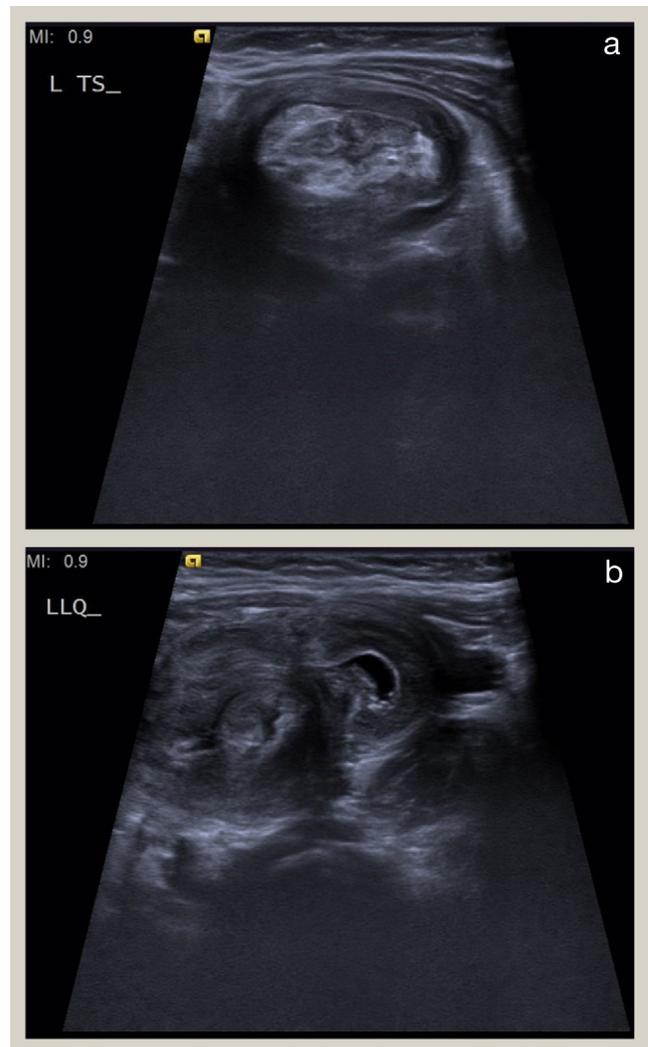


Figure 1 Ultrasound features of intussusception. (a) Target sign; a transverse section through the intussusception with a loop of oedematous bowel sitting within another bowel loop. (b) Pseudo-kidney sign; a longitudinal section through the intussusception with the intussusceptum seen passing through the distal segment of bowel.

with the advances in high-resolution ultrasound, it is rarely used for diagnosis unless the USS is inconclusive.

Cross-sectional imaging, such as CT and MRI, can diagnose intussusception but, again due to the accuracy of USS, are rarely necessary in the acute setting. However, these may be useful for subsequent investigation for a pathological lead point in atypical or recurrent cases.

A plain abdominal radiograph is not sensitive for diagnosing intussusception and especially in early cases it can be normal. It may be useful to assess features of intestinal obstruction or perforation. Findings seen on an abdominal radiograph in intussusception include paucity of bowel gas in the right iliac fossa; a soft tissue mass representing the intussusceptum; dilated bowel loops suggestive of bowel obstruction. Occasionally the tip of the intussusceptum is outlined by a gas shadow giving a crescent shaped lucency known as the meniscus sign. Free air may be identified, which indicates bowel perforation.

Initial blood tests should be taken, to include full blood count, urea, creatinine and electrolytes, CRP and blood cultures. A sample for group and save for blood transfusion is also usually taken concurrently in case surgical intervention is required later.

Management

In the UK, over the last few decades, treatment of children with intussusception has moved to specialized tertiary centres with availability of paediatric surgeons and paediatric radiologists. If a child is suspected of having an intussusception by local paediatricians or emergency physicians, then referral to a paediatric surgical centre is required. Initial resuscitation is often initiated and the child stabilized prior to transfer.

Spontaneous reduction of an intussusception without treatment can occur in approximately 20% of cases.

Resuscitation

Initial resuscitation is of paramount importance. Intravenous cannulation is required and blood samples should be taken. Fluids resuscitation is with a minimum of 20 ml/kg bolus of normal saline. A nasogastric tube should be inserted to decompress the stomach and prevent further vomiting. Sometimes these children can present with shock and require resuscitation using the advanced paediatric life support algorithms, including supplemental oxygen and more rarely intubation and ventilation if needed, as well as significant fluid resuscitation. These cases should be stabilized in a critical care setting such as a paediatric high dependency unit or intensive care unit prior to further radiological or operative intervention.

Intravenous broad-spectrum antibiotics, such as co-amoxiclav, should be given prior to further intervention. Analgesia should be given; morphine may be required and can be used with appropriate precautions.

Adequate resuscitation and stabilization is essential prior to transferring the child for a definitive intervention.

Non-operative intervention (Figure 2)

In the UK, non-operative intervention with air enema reduction is the first-line treatment of choice for uncomplicated intussusception. The procedure involves the insufflation of air under

pressure into the large bowel via the anus to push back the intussusceptum. Fluoroscopic imaging is used to visualize the process and confirm its success.

It is contraindicated in patients with significant haemodynamic instability, peritonitis or perforation. Cases with a delayed presentation (i.e. symptoms for more than 48 hours) or evidence of small bowel obstruction are less likely to be successful. Older children (aged over 2 years) can be uncooperative with the procedure and therefore it may be more challenging. Younger infants (under 3 months of age) may not tolerate the procedure well and caution must be taken in these cases. Patients, in whom a pathological lead point has already been identified should proceed straight to operative intervention.

Informed consent must be taken from a parent. Complications of the procedure include incomplete reduction, recurrence and perforation. Due to the pressure at which the bowel is being inflated, if a perforation occurs during the procedure it will result in a tension pneumoperitoneum. This requires immediate needle decompression by insertion of a large bore (18G) cannula into the abdomen, usually in the right iliac fossa.

The procedure is usually performed after confirmation of intussusception on an USS in the radiology department. Although children undergoing air enema should have undergone adequate resuscitation before being taken to radiology for the procedure, essential equipment should be available, including nasogastric tube, cannulation equipment, intravenous fluids, analgesia and access to a paediatric resuscitation trolley. A paediatric radiologist with adequate experience in the technique should carry out the procedure. Additionally, there should be a radiographer, experienced paediatric nurse and paediatric surgeon present during the procedure. It has been shown that the presence of a paediatric surgeon during the air enema reduction increased the rate of successful reduction. The surgeon is able to aid decision-making and also manage the complication of a perforation should this occur. The fluoroscopy room should be adequately warmed prior to starting. The child needs continuous monitoring of their heart rate and oxygen saturations throughout the procedure.

A large catheter (ideally 24 Fr) is inserted into the rectum. The buttocks are then taped or held together to create a seal and prevent air leaking out the anus to allow the pressure to be maintained. A hand-held sphygmomanometer or a pressurized air delivery system with a pressure control valve may be used. Recommended insufflation pressures start at 60–80 mmHg and can be increased up to 120 mmHg. Each insufflation attempt should not last more than 3 minutes at a time but can be repeated, usually up to three times. The majority of successful reductions occur within the first 1–2 minutes of the procedure. The fluoroscopic imaging is used to monitor the progress of reduction. Success is confirmed when reflux of air is seen into the ileum and air fills multiple small bowel loops centrally. This is often associated with an immediate improvement in the child's clinical picture and they may fall asleep. At this stage, especially if morphine has been used for analgesia, close monitoring of the child respiratory status is essential to detect apnoea or desaturation.

Occasionally it can be difficult to determine success fluoroscopically and a repeat USS may be needed to confirm reduction. Observation following the procedure for 24 hours is usual policy

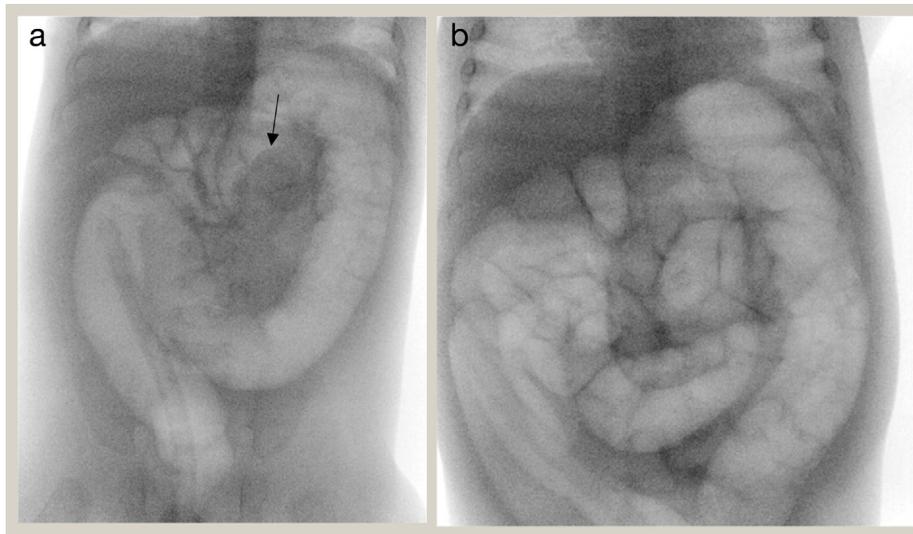


Figure 2 Fluoroscopic images taken during the air enema reduction of intussusception. (a) The air is seen filling the distal colon pushing and outlining the tip of the intussusceptum (marked with an arrow), the rectal catheter can also be seen sitting in the rectum. (b) Air is now seen filling the whole colon and refluxing into multiple central small bowel loops.

to ensure that symptoms have fully resolved and there is no recurrence. Once the child is clinical improved they can be allowed to eat and drink and be discharged home.

If the reduction is not completely successful, it is possible to repeat the air enema 2–6 hours later, as long as the child remains clinically stable. Delayed repeat air enemas have increased the success rate of non-operative intervention and avoids the need for surgery in some children, reducing length of stay and reducing the numbers of case in which bowel resection is needed. Repeat air enemas can also be used for recurrent intussusception however, the possibility of a pathological lead point must be considered and investigated for as appropriate.

Success rates for air enema reduction reported are variable, but a recent UK study showed a median success rate of 70%⁴ and recurrence rates in the following 48 hours after a successful air enema reduction is around 8.5%.⁵

An alternative technique to the air enema reduction is a hydrostatic enema reduction under USS guidance, which is used in some countries. This was originally described using barium but this has been replaced with saline. The disadvantage of barium is the significant peritoneal contamination should a perforation occur.

Operative intervention

Indications for surgery in intussusception include bowel perforation, peritonitis, haemodynamic instability, known pathological lead point and failed attempts with non-operative reduction. Surgery involves manual reduction of the intussusception and can be performed with either an open or laparoscopic approach. Partial non-operative reduction does benefit subsequent surgical manual reduction by making this process easier.

Open procedure: open reduction of intussusception is performed via a right-sided infra-umbilical transverse muscle-cutting laparotomy incision. The intussusception mass is manipulated out of the wound. Reduction is attempted by applying gentle and constant pressure at the distal end to push the intussusceptum

proximally. Continuous pressure on the mass with warm saline-soaked swabs may help to reduce bowel wall oedema thus helping with the reduction. Serosal tears may occur during this process, and as long as they are not full thickness, are usually inconsequential, although they should be avoided if possible. Reduction through the ileocecal valve is often the most difficult stage and requires patience and persistence.

Once the bowel is fully reduced it should be inspected for adequate viability. If the appearance of the bowel is dusky and viability is uncertain, the bowel can be wrapped in warm saline swabs or returned to the abdominal cavity for 5 minutes then re-assessed for return of adequate perfusion. If areas of necrosis are apparent, the bowel is not viable, there is a significant perforation or complete reduction is not possible then resection of the affected segment should be performed. This usually consists of a limited right hemicolectomy with primary end-to-end anastomosis. Around 40% of those cases requiring surgical management will need bowel resection. Resection is more likely in those with a prolonged history. Stoma formation is usually unnecessary in the majority of cases, although may be required if there has been a perforation with significant contamination. The bowel should also be assessed for the presence of a pathological lead point and, if present, this can be managed with surgical excision as appropriate.

Laparoscopic procedure: reduction of the intussusception can also be performed laparoscopically and this approach is becoming increasingly popular. An open technique at the umbilicus is used to insert an initial 5 mm port and create the pneumoperitoneum. Further laparoscopic ports can then be placed under direct vision, usually using a 30 laparoscope, commonly in the left iliac fossa and epigastrium to allow access to the right side of the abdomen, in particular the right iliac fossa. Gentle traction is applied by pulling on the proximal limb of bowel and distal pressure on the intussusceptum to push and pull the intussusceptum back.

Conversion to open surgery is required in approximately 30% of cases. It is more likely to be required in cases where there has been a late diagnosis, those with a perforation, or when the proximal bowel is significantly dilated. If open conversion is required, the umbilical incision can be extended and the bowel mobilized and delivered through the umbilical wound.

One main disadvantage is the lack of tactile feedback mean that a pathological lead point such as a gastrointestinal polyp may be easily missed than during an open procedure. However, limited evidence does suggest that laparoscopic reduction may be associate with a shorter length of stay.^{6,7}

Further management

Following reduction of the intussusception, oral fluids can be resumed and can be built up as tolerated. Most patients are observed for 24–48 hours until they are clinically well and can be discharged. Within the first 24–48 hours, spikes of temperature can be common and do not need further treatment. If bowel resection is required during surgical intervention then the patient may need to be nil by mouth for a period of time post-operatively, may need further antibiotics and require a longer hospital stay.

Follow-up after discharge is not usually required unless a pathological lead point is suspected and requires further investigation or management.

Complications

Perforation: iatrogenic perforation can occur during air enema reduction and the incidence is approximately at 1%. It is more common in patient under 3 months of age. It requires immediate decompression of the pneumoperitoneum using a large bore needle and definitive management with surgery.

Recurrence following air enema reduction is quoted at 10–15%, while with surgical reduction it is 4–5%.^{4,5}

Mortality: death related to intussusception is now very rare in the UK. Deaths tend to be related to delayed diagnosis, inadequate resuscitation and surgical postoperative complications.⁸ ♦

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