

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 classical 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. However, this figure has been declining over the last 50 years. There are also significant geographical 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

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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. 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

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 Henoche-Schonlein purpura or Coeliac disease

Box 1

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.

The most common anatomical site for intussusception is ileocolic. This accounts for more than 80% of idiopathic intussusceptions. 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 only 20–60% of cases.

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

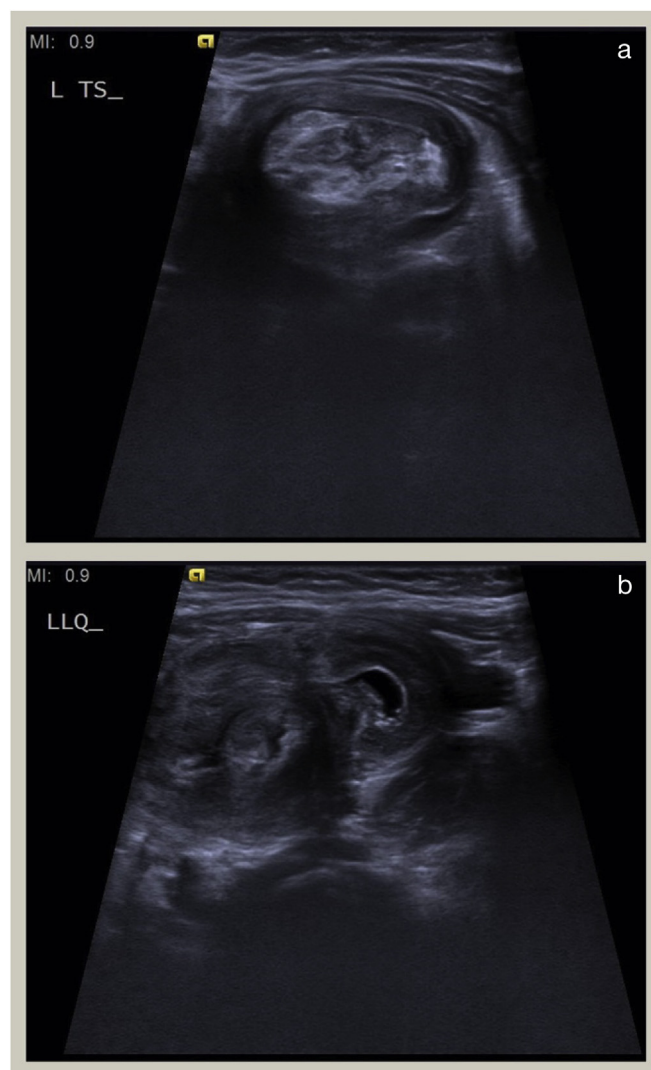


Figure 1 Ultrasound features of intussusception. (a) Target sign; a transverse section through the intussusception with a loop of oedematous bowel sitting within another bowel loop. (b) Pseudo-kidney sign; a longitudinal section through the intussusception with the intussusception seen passing through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

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