You are what you eat: The diagnosis of recurrent intussusception in the emergency department

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INSTRUCTIVE CASE

You are what you eat: The diagnosis of recurrent intussusception in the emergency department

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Ileocolic intussusception is the most common abdominal emergency in early childhood and is usually idiopathic.[1] Diagnosis requires a high index of suspicion as the pathognomonic triad of redcurrant jelly stools, palpable mass and intermittent abdominal pain is present in less than a quarter of cases.[1] It should be differentiated from ileoileal intussusception, which is typically transient and self-limiting.[2] However, if children present to the emergency department (ED) with acute, severe abdominal pain due to recurrent intussusception, an underlying diagnosis should be considered, as in the case reports detailed below.

Case Report

Case 1

A 13-month-old girl presented to the ED with 1 day of vomiting and reduced oral intake, on the background of 6 days of frequent loose stools, and 2 weeks of abdominal distension. She had no recent infective symptoms. She was vaccinated and had no significant past medical or family history.

On examination, she was irritable, afebrile, tachycardic (heart rate 150 bpm) and hypertensive (blood pressure 150/100 mmHg). She had a markedly distended, tender but soft abdomen, with quiet bowel sounds and a reducible umbilical hernia. The main differential diagnosis included acute surgical pathology, including a partial bowel obstruction, or constipation with faecal impaction and overflow diarrhoea.

Key Points

1. Ileocolic intussusception is a surgical emergency and must be differentiated from ileoileal intussusception.
2. Recurrent intussusception can be associated with an underlying diagnosis of coeliac disease (CD).
3. Infants and pre-school-aged children presenting to the emergency department with recurrent intussusception should be investigated for a diagnosis of CD.
4. Recurrent ileoileal intussusceptions due to CD generally resolve with the introduction of a gluten-free diet.

Initial investigations included venous blood gas with a normal pH, lactate of 2.8 mmol/L, glucose of 3.6 mmol/L and ketones of 3.2 mmol/L. Full blood count, electrolytes, liver function tests and CRP were normal, with thyroid function and coeliac serology (anti-TTG IgA titre) pending as secondary causes of constipation. An abdominal X-ray demonstrated a non-specific bowel gas pattern, with no evidence of bowel obstruction.

She proceeded to an abdominal ultrasound scan (USS) which demonstrated a 6-cm irreducible ileocolic intussusception in the right iliac fossa (Fig. 1). She underwent a successful air enema reduction by interventional radiology and was discharged the following day with regular osmotic laxative.

She represented to the ED 2 days later with worsening abdominal pain and distension, mucous in her stool and minimal oral intake. Repeat abdominal USS demonstrated a self-resolving ileoileal intussusception. This did not require any intervention and she was discharged after overnight observation.

She represented 10 days later with further vomiting, inconsolability, abdominal distension and new weight loss. Repeat USS did not demonstrate an intussusception. However, review of her first presentation bloods showed an anti-TTG IgA titre of 710 g/L. Coeliac disease (CD) was confirmed serologically as per ESPGHAN guidelines,[3] with repeat rising anti-TTG titre (>1000 g/L) and positive endomysial antibody. In consultation with paediatric gastroenterology, she was discharged on a gluten-free diet (GFD). Outpatient follow-up 9 days later demonstrated markedly improved symptoms with no further episodes of intussusception to date.

Case 2

A 4-year-old boy was seen in the ED with acute severe central abdominal pain associated with a pre-syncopal episode, inconsolability and dry retching. He had a background of six similar episodes over the previous 2 months, with three USS confirmed ileoileal intussusceptions diagnosed in ED. Additionally, he had iron deficiency anaemia on oral supplementation, poor weight gain and asthma. He had a family history of type 1 diabetes in a first-degree relative.

On this presentation, he was inconsolable, tachycardic and non-compliant with examination. He required intranasal midazolam for an abdominal USS, which demonstrated a further ileoileal intussusception. He was admitted to the ED short stay ward for observation, analgesia and a trial of oral fluids. Serial point-of-care USSs demonstrated the persistence of the ileoileal

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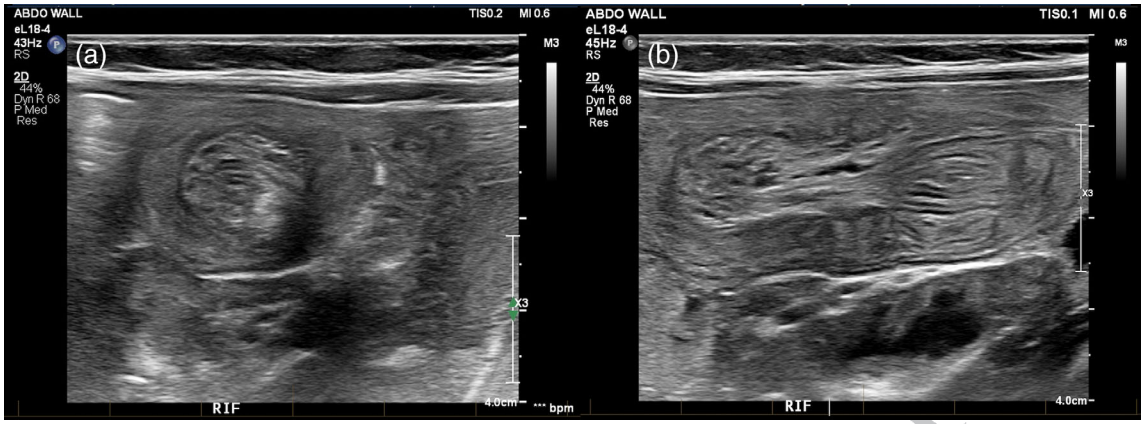

Color Figure - Print and Online

Fig 1 Radiology performed ultrasound images of an ileocolic intussusception in a 13-month-old infant demonstrated in both (a) transverse and (b) longitudinal views.

intussusception. However, he improved symptomatically over this observational period, with no further episodes of severe abdominal pain. During this admission, CD was considered as an underlying diagnosis for his recurrent intussusception, and screening bloods were sent prior to discharge. Five days later, the anti-TTG titre returned at >1000 g/L with positive endomysial antibody. His serological diagnosis of CD was independently confirmed by a paediatric gastroenterologist, and he had complete resolution of symptoms with a GFD.

Discussion

CD affects up to 1% of the population and, despite a rising incidence, remains significantly underdiagnosed due to a wide spectrum of symptomatology.[3] CD presents classically in young children shortly after the introduction of gluten into the diet and is usually associated with chronic diarrhoea, anorexia, abdominal distension and pain, and poor weight gain or weight loss. However, it can also be subclinical with mild symptoms or non-specific extraintestinal features.[3] There have been previous case reports which link the relationship between intussusception and CD in children.[4–6] All of these children had recurrent transient ileoileal intussusception with subsequent diagnosis of CD, and resolution with the commencement of a GFD.

Larger studies have attempted to establish the association of intussusception more definitively by investigating children with untreated CD. One retrospective study examined the medical records of 254 children with biopsy-diagnosed CD and found an incidence of four children (1.2%) having ileoileal intussusception in the preceding 9 months, which was about 17 times higher than the matched population in the study.[7] Another study, assessing both the prevalence and natural history of intussusception in CD, performed serial abdominal USSs on 133 newly diagnosed CD children and found almost 25% had an incidental intussusception, almost exclusively ileoileal, all of which were relatively asymptomatic and conservatively managed, with the vast majority resolving within a month of commencement of a GFD.[8]

Although in retrospect both children we reported had underlying chronic symptomatology of CD, it was the repeated episodes of acute severe abdominal pain due to intussusception that necessitated presentation to the ED, prompting further investigation for an underlying cause. The exact mechanism of CD causing recurrent intussusception remains unclear but is thought to be due to inflammatory changes and altered motility of the small bowel wall.[8] Moreover, it is becoming apparent that many children with untreated CD have occurrences of relatively asymptomatic ileoileal intussusceptions. However, for children who present to the ED with recurrent episodes of acutely symptomatic intussusception, a diagnosis of CD should be strongly considered, as their definitive management usually requires the introduction of a GFD.

Written consent to publish these case reports was obtained from the legal guardian (parent) of each child.

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