Pediatric intussusception: A Case Series and Literature Review
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Citation

Abstract
Background: Intussusception is the major cause of acute intestinal obstruction in infants. The classic clinical triad of intussusception is colicky abdominal pain, vomiting and bloody stools. However, only 20% of patients present with this triad.
Objectives: The objective of this article with a series of cases of intussusception is to review the spectrum of clinical presentation of this disease entity.
Methods and Results: We describe 3 patients with intussusception that presented to our Emergency Department (ED) with different complaints. Also this article reviews diagnosis and management strategies of intussusception.
Conclusions: Clinical suspicion is the key for diagnosis. This article highlights the importance of suspecting intussusception by the ED physician in any child with abdominal pain, gastrointestinal symptoms or altered mental status.

INTRODUCTION
Intussusception, a common gastrointestinal (GI) emergency in children, develops when a proximal segment of the gastrointestinal tract (intussusceptum) telescopes into a distal portion (intussuscipiens), most commonly located near the ileocecal valve. Intussusception occurs in approximately 56 children per 100,000 per year in the United States (US). Of these, 60% of cases occur before 1 year of age and 80% before 2 years. Its incidence is highest between 5 and 9 months of age, being uncommon in neonates. There is a male predominance of 3:2.

The majority of pediatric cases involve the ileocolic (or ileocecal) portion of the intestine. The most common location of idiopathic intussusception is at the hepatic flexure. When the intussusceptum invaginates and pulls along the mesentery, there is compromise of venous return. This is followed by engorgement of the intussusceptum, edema and bleeding from the mucosa which lead to currant jelly stools. Finally the arterial blood supply of the intestine gets compromised leading to necrosis, perforation and/or shock.

Children less than 3 months and greater than 2 years old are most likely to have a “lead point” or specific identified cause, such as (in decreasing order of incidence): meckel diverticulum, duplications, polyposis and lymphomas. Hyperplasia of the intestinal Peyer’s patches have been identified as the leading point in infant cases. Most cases are reported during summer and spring which favor a not fully understood infectious mechanism. Peyer’s patches hyperplasia can occur secondary to viruses such as adenovirus, enterovirus, echovirus and human herpes virus 6. The most common surgical complication of patients with Henoch Schönlein Purpura is intussusception. Its incidence is also increased in the postoperative period probably due to edema or adhesions. In 1999, the Rotashield rotavirus vaccine was removed from the US market due to its increased association with intussusception, especially between 3-14 days post-placement of the vaccine. Subsequently vaccines such as RotaTeq and Rotarix have been developed.

CASE REPORTS
We present 3 patients that presented to our pediatric emergency department (ED), in order to review the spectrum of presentation, diagnosis and management of this disease entity.

CASE 1
A 6-month-old, previously healthy boy was brought to the ED for clear emesis of 1 day. No history of fever, diarrhea, irritability or trauma. On exam the child appeared well, with normal vital signs and with a benign physical exam. Abdomen was soft to palpation with normal bowel sounds.
The patient was treated in the ED as a viral gastritis. He tolerated oral fluids well and was discharged home. Parents returned because emesis continued. On his second visit, a rectal exam revealed occult blood in stools. He was taken for abdominal x-rays which showed a questionable mass on the right lower quadrant (RLQ) suggestive of intussusception. Barium enema failed to reduce the mass and the child was taken to the OR with uneventful course.

CASE 2
A 12-month-old male presented with altered state of consciousness (ASC) for 2 hours. Prior to the ED presentation, there was no fever, no vomiting, no diarrhea, no irritability and no trauma. The child was previously healthy. On exam, he was well developed, well nourished but responded to painful stimuli only. Vital signs were normal and physical exam was benign. Workup for ASC was initiated and en route to CT scan of the head he passed currant jelly stools. The patient was taken to the radiology suite where an air enema was performed with successful reduction of the mass.

CASE 3
A 7-year-old healthy male presented to the ED with abdominal pain for a few hours prior to arrival. He presented with diffuse abdominal pain that came and went. No vomiting, no fever and no diarrhea were present. At the ED, the child appeared playful and was running around. Vital signs were normal and physical exam was benign. The plan in the ED was to observe the patient. During the ED stay he developed intense abdominal pain that lasted few minutes after which he was back to his usual self. Abdominal x-rays demonstrated a RLQ mass. The child was taken for barium enema which successfully reduced the intussusception.

DISCUSSION

CLINICAL PRESENTATION
The typical triad of severe intermittent abdominal pain, currant jelly stools and vomiting is seen in less than 20% of cases.

There may be bouts of abdominal pain. In ileocolic intussusception palpation of the abdomen may reveal a sausage-shaped mass which is located in the RUQ. A wide spectrum of symptoms may range from painless intussusception to constipation, dehydration, diarrhea, intestinal prolapse, rectal bleeding, sepsis, shock, syncope, vomiting and altered mental status (lethargy and irritability). Lethargy is seen most frequent in infants and young children with or without history of gastrointestinal symptoms. Altered mental status has been hypothesized to be secondary to a combination of factors such as dehydration, electrolyte imbalance, and endorphins or toxic metabolic products released from the ischemic bowel which can affect the brain.

Recurrent intussusception is present in only 5-8% of children and is most common after hydrostatic versus surgical reduction. Fifty percent of recurrent intussusception cases occur within 48 hours of a prior episode (but have been reported up to 18 months later). Most postoperative intussusception cases are located in the small bowel.

DIAGNOSTIC MODALITIES

ABDOMINAL RADIOGRAPHS
Plain abdominal films (supine frontal and left lateral decubitus) are the first imaging studies used in the ED. However, they lack sensitivity (45%) and may give many false negatives. Classic intussusception radiological signs are:

- Absence of air in the ascending colon (RUQ and RLQ).

(Figure 1)

Figure 1: Absence of air in the ascending colon.
Soft tissue density in the upper abdomen (up to 60% of patients).

Small bowel obstruction signs: small bowel dilation, air fluid levels. (Figure 2)

**Figure 2**
Figure 2: Small bowel obstruction signs: left: small bowel dilation (smooth bowel walls) and right: few air fluid levels.

Target sign: two concentric lines seen in the right upper quadrant which correspond with mesenteric fat of the intussusceptum. (Figure 3 and 4)

**Figure 3**
Figure 3: target sign and crescent sign.

Target sign: 2 concentric lines seen in RUQ which corresponds with mesenteric fat of intussusceptum. Crescent Sign: Semilunar lucency seen within course of colon and corresponds to air trapping between 2 intestinal surfaces.

**Figure 4**
Figure 4: Target sign (left) and crescent (right) sign marked.

Crescent sign: Semilunar lucency within the course of the colon (especially the transverse colon) beyond the hepatic flexure, which is wider than normal bowel in diameter and represent intestinal gas trapping between the two intestinal surfaces caused by stretching of the bowel wall. (Figure 3)(Figure 4)

**ULTRASOUND**

Ultrasound is a fast, non-invasive and simple reproducible test. It’s sensitivity (98-100%) and specificity (88-100%) is high, but is clearly operator dependant. At some centers it has displaced radiographs as the initial imaging test of choice. Other indications for its use are: suspected cases of intussusception where abdominal x-rays are non-conclusive, evaluation for reducibility, presence of a lead point mass, potential incomplete reduction after enema and intussusception limited to small bowel. Classic findings include: the target lesion or doughnut sign on transverse imaging and the pseudokidney sign on longitudinal imaging. (Figure 5) (Figure 6)
COMPUTED TOMOGRAPHY

Computed tomography (CT) scan is usually not indicated in children, since the diagnosis is generally confirmed by ultrasound or enema. The CT scan involves high costs, radiation exposure and sedation risk which is overall less convenient for this population group.

DIAGNOSTICS AND THERAPEUTICS

BARIUM AND AIR ENEMA

Contrast enemas (barium, water-soluble and air) are diagnostic and therapeutic techniques, with reduction rates of 70 to 90%. Barium enema is no longer considered the gold standard for non-surgical treatment, though its use is still extensive. Risks with barium enema such as shock and bowel perforation (secondary peritonitis, infection and adhesions) should never be overlooked. This test should be considered only after stabilizing the child, adequate hydration, and consultation with a pediatric surgeon. The only current absolute contraindication for barium enema is full bowel necrosis.

Air enema’s use has increased due to its lower perforation risk, less radiation exposure, faster and better reduction rate.
One study compared air vs. contrast enema reporting success rates of 76% and 63% respectively. Intussusception recurrence rates for air versus liquid enema are reported to be similar, approximately 10%. (Figure 7)

Figure 7
Figure 7: Air enema with intussusception near the hepatic flexure. Coiled spring appearance which corresponds with barium in lumen of intussusceptum.

Air enema is now considered the gold standard treatment of intussusception in children. Surgical intervention is needed only in unstable patients with non-operative reduction contraindications or in prior unsuccessful reduction attempts. Barium enemas should be avoided in children with evidence of peritonitis, severe shock, sepsis, perforation or extreme ages.

Enema reduction should be attempted 3 times at the most before considering it unsuccessful and submitting the patient to surgery. Poor enema reduction rate is determined by (in descending order): symptoms longer than 24 hours, age less than 3 months, dehydration and intussusception of the rectum. Repeat enema is both safe and effective in recurrent intussusception. Spontaneous reduction occurs in approximately 10-17% of patients, being most common in the small bowel.

CONCLUSIONS/SUMMARY
This article highlights the diverse signs and symptoms pediatric patients with intussusception can present with. Only 20% of children will present with the triad (abdominal pain, currant jelly stools and vomiting). Clinical suspicion is the key factor in making a diagnosis of intussusception and should be considered by the ED physician even in absence of typical signs and symptoms. Delay in diagnosis increases risk for bowel obstruction, perforation, necrosis and death.

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References

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