

CLINICAL VIGNETTE

A Case of Intussusception in a Patient with Congenital Adrenal Hyperplasia

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Case Report

A 4-month-old female with a past medical history of a salt-wasting form of congenital adrenal hyperplasia (CAH) presented to the emergency department with acute onset of non-bilious vomiting. Her family denied crying episodes at home. Symptoms started 3 hours prior to presentation and she has had 5-6 episodes before her emergency department visit. Vomiting started 30 minutes after being fed milk and her medications. There were no bowel movements, fevers, abdominal pain or crying. Her mother reported recovery from an upper respiratory infection 1 week earlier. On physical exam, patient had slightly dry mucus membranes, but was alert, reactive and non-toxic appearing. The abdominal exam was benign with no tenderness, guarding, masses or distension. The genitourinary exam was significant for cliteromegaly due to CAH. She was diagnosed with CAH shortly after birth and has been treated with daily hydrocortisone, fludrocortisone and sodium chloride salts.

In the emergency department, the patient was treated with intravenous fluids, ondansetron and stress dose hydrocortisone. She had two more episodes of vomiting despite anti-emetics and her exam remained unchanged between vomiting episodes. Two hours after admission, she had a large bloody bowel movement described as currant jelly stool. See figure 1.



Figure 1: Diaper after bloody movement in the emergency department.

Pediatric general surgery was consulted and a barium enema was diagnostic for intussusception but not therapeutic after three attempts. See figures 2 and 3.



Figure 2: A filling defect is noted at the mid transverse colon, consistent with an intussusception



Figure 3: After a third attempt, the transverse colon opacified, but the intussusception was pushed into the hepatic flexure/ascending colon region without reduction of the intussusception.

Laparoscopy showed ileocolic intussusception with gangrenous bowel and she underwent an exploratory laparotomy with ileocectomy successful recovery.

Intussusception is the invagination of one part of the bowel into an immediately adjacent distal portion. This results in venous congestion and bowel wall edema¹. The most common type of intussusception is ileo-colic invagination. This

medical emergency most commonly occurs between the ages of 3 months to 5 years². For all age groups, the cause of intussusception is idiopathic but it has been associated with polyps, malignancies such as lymphoma and Merkel's diverticulum in older children³.

The classic triad of colicky abdominal pain with intermittent crying, vomiting and bloody stools is observed in less than 40% of patients^{4,5}. In our patient, vomiting and later bloody stools were the only presenting signs. Some parents will describe episodes of a child crying for periods of 1-5 minutes and then 10-15 minutes of a well-appearing and playful child⁵. However, symptoms may progress to more frequent crying, vomiting, fevers, lethargy and later bloody stools. The abdominal exam may be tender, guarded and distended. A mass may be palpated in the right upper or lower quadrants in some patients⁶; however, the abdominal exam can be benign and unrevealing. Rectal exam may reveal occult blood or frank bloody stools described as currant jelly stools. This finding is not always present early in the course⁷.

Differential diagnoses to consider in suspected cases of intussusception include testicular and ovarian torsion, appendicitis, incarcerated hernia and gastroenteritis. These diagnoses may be excluded with a careful history and physical exam.

Several modalities have been used in the diagnoses of intussusception. Plain radiographs are not sensitive and usually normal but on occasion may show air-fluid levels, dilated bowel loops or a visible abdominal mass as the disease progresses⁷. Abdominal ultrasound has been used by many institutions and may show a single, hypoechoic ring with a hyperechoic center, representing edematous bowel wall⁸. The gold standard for diagnoses and treatment of intussusception has been barium enema, however, air and saline enemas have also been successful⁹. When reduction is unsuccessful, patients should undergo laparoscopy or laparotomy. In addition, patients who present with evidence of perforation, peritonitis or shock in whom use of enemas are contraindicated will require immediate surgical intervention. CT scan of the abdomen enables the physician to rule out other potential causes of acute abdomen pain and may provide a definitive diagnosis of intussusception¹⁰.

Patients who are treated either by radiology or surgery need to be admitted for a period of

observation. This is because of recurrence rates of about 5% in radiology treated and 2% in surgically treated in the first few hours to days¹¹. A more recent study reports recurrence rates of 14% at 6 months¹².

Intussusception can lead to devastating consequences. However, the diagnosis cannot be entirely excluded with certainty by history, physical examination, laboratory studies or plain X-rays. If the diagnosis is considered, evaluation with either an ultrasound or the gold standard contrast enema should be performed expeditiously.

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