Abstract:

A 2-year-old previously healthy girl presented to the emergency department with sudden onset of nonbilious emesis and severe abdominal pain. Her clinical examination and point-of-care ultrasound were consistent with a small bowel obstruction. She had sonographic evidence of free fluid in the abdomen and subsequently developed hemodynamic instability. Further imaging was suggestive for a congenital anomaly causing her obstruction. She was ultimately diagnosed with an omphalomesenteric duct remnant, confirmed during an exploratory laparotomy.

Keywords:

small bowel obstruction; closed-loop obstruction; point-of-care-ultrasound (POCUS); omphalomesenteric duct remnant

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You Bowel Believe It!

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A 2-year-old girl presented to her local emergency department (ED) with emesis and abdominal pain. Her parents reported that she awoke suddenly in the middle of the night screaming in pain. She had multiple episodes of emesis, initially mucous-like in character but progressed to blood stained. Her emesis remained nonbilious. She did not have diarrhea or rectal bleeding. A review of systems was negative for fever, weight loss, easy bleeding, or bruising. Her family denied any preceding trauma or witnessing foreign body ingestion. She was a full-term baby and had no significant medical or surgical history. She took no regular medications and had no known drug allergies. She lived at home with her parents and a 2-week-old sibling.

On presentation at the local ED, she had a heart rate of 191 beats per minute and a blood pressure of 61/36 mm Hg. Her oxygen saturation was 100% on room air. She appeared pale and mottled. Her abdomen was soft and nondistended. Preliminary laboratory studies showed a white blood cell count of 31 K/μL and hemoglobin of 13.9 g/dL and were otherwise notable for a blood urea nitrogen of 41 mg/dL, creatinine of 0.7 mg/dL, and glucose of 231 mg/dL. Preliminary radiographic studies showed mildly distended loops of bowel and soft tissue fullness in the right lower quadrant (Figure 1). No free air was demonstrated.

Initial management included fluid resuscitation with 800 mL of normal saline and a dose of ampicillin-sulbactam. A nasogastric tube was placed with minimal output. She continued to deteriorate and developed repetitive episodes of coffee ground emesis. Her blood pressure showed only transient improvement with fluid resuscitation, and she was started on a dopamine infusion at 10 μg/kg/min. She was flown to the nearest tertiary referral center and seen immediately in the resuscitation bay by pediatric emergency medicine physicians.

On arrival, she had a pulse of 202 beats per minute, blood pressure of 78/57 mm Hg, a respiratory rate of 24 breaths per

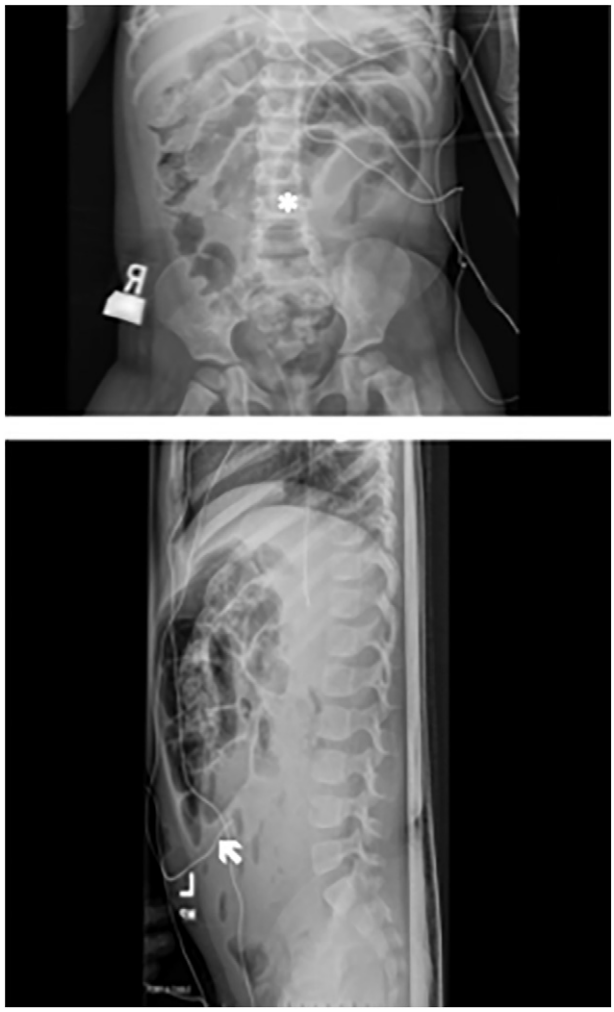

Figure 1 Posterior-anterior and cross-table lateral radiographs of the abdomen demonstrating mildly distended loops of bowel (*) and air-fluid levels (white arrow).

minute, and oxygen saturation of 100% on a nonrebreather mask. She had weak peripheral pulses with a capillary refill time of 4 seconds. She had no adventitial respiratory sounds. Her abdomen was soft and nondistended but globally moderately tender on palpation. There was no appreciable abdominal mass. She had hypoactive bowel sounds. On complete cutaneous examination, she had no signs of bruising. There was no notable lymphadenopathy. Her initial venous blood gas showed a pH of 7.35, pCO_2 of 34 mm Hg, pO_2 of 88 mm Hg, bicarbonate 18.5 mmol/L, and lactate of 1.74 mmol/L. Her point-of-care hemoglobin was 9.7 g/dL. A point-of-care ultrasound (POCUS) showed a significant amount of free fluid in the abdomen (Figure 2).

Her dopamine infusion was titrated to 20 μg/kg/min, and she received low-dose boluses of epinephrine. In light of her continued hemodynamic deterioration, she was intubated. She received doses of etomidate and succinylcholine to facilitate intubation. With her persistent hypotension, POCUS findings, and drop in hemoglobin from the outside hospital, she received a packed red blood cell transfusion. A computed tomography (CT) study of the abdomen was obtained, which suggested the ultimate diagnosis.

DIFFERENTIAL DIAGNOSIS

The differential for patients presenting with acute abdominal pain and vomiting is broad, including both common self-limited conditions such as gastroenteritis and constipation but also life-threatening medical and surgical emergencies that require prompt intervention. Observational case series have shown that of children evaluated in outpatient clinics or EDs with abdominal pain, 22% have a diagnosis that requires either surgical

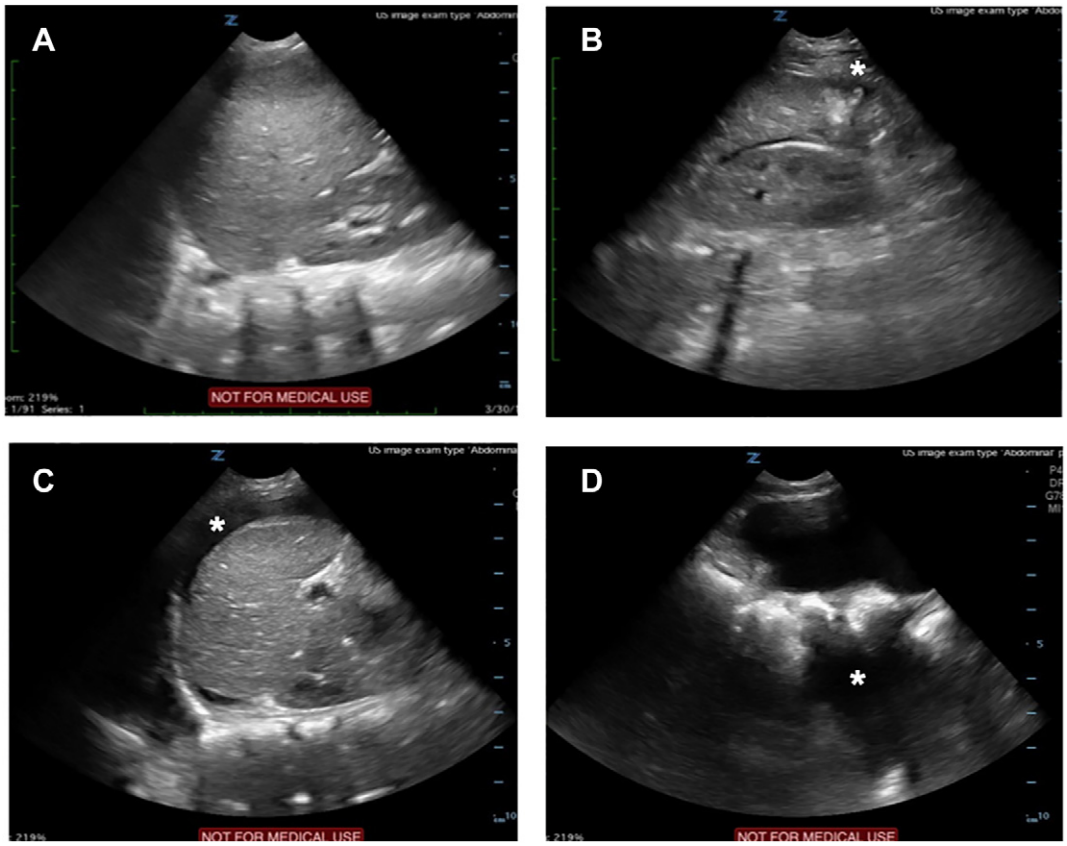

Figure 2 Point-of-care ultrasound images of the abdomen. A, Right upper quadrant (RUQ) no significant free fluid in first image of RUQ (limited view of Morison's pouch, liver tip not visualized). B, RUQ at liver edge showing accumulation of free fluid at tip of liver and in Morison's pouch. C, Left upper quadrant free fluid in a circumferential pattern around the spleen. D, Pelvic free fluid adjacent to the bladder.

intervention or antibiotics.[1] A comprehensive list of differential diagnoses can usually be tailored using epidemiological factors (such as the patient's age and sex), medical history, the pattern and quality of pain, presence of associated symptoms, and repeated careful physical examinations.[2]

The medical history of a patient may help increase suspicion for particular diagnoses. Previous abdominal surgery increases the risk for developing adhesions that can predispose to obstruction. Chronic conditions such as inflammatory bowel disease, choledocolithiasis, nephrotic syndrome, constipation, gastroesophageal reflux, and pancreatitis may all present with exacerbations associated with abdominal pain. In addition, children with Hirschsprung disease can present with obstruction and fulminant colitis.

The quality and location of pain are important considerations for providers as they consider a differential diagnosis. One caveat is that very young children will not be able to describe pain in these terms. Pain that localizes may help narrow the differential, although younger children are typically unable to localize pain. Right upper quadrant pain can be associated with hepatitis, choledocolithiasis, cholecystitis, or Fitz-Hugh-Curtis syndrome. Right lower quadrant pain can be associated with appendicitis (especially with a history of pain that migrates from the periumbilical region). Epigastric pain can be associated with pancreatitis, gastritis, and gastric ulcers. Pain in either the right or left lower quadrant and pelvis in female patients may be associated with ovarian pathologies, pelvic inflammatory disease, and complications from pregnancy. The quality and time course of the pain may also be informative. Intermittent colicky pain can be suggestive for intussusception. Sharp flank pain that radiates to the groin is suggestive for renal calculi, although younger children are less likely to present with localizing pain.[3]

Trauma (both accidental and inflicted) is a common cause of abdominal pain in children. A history of trauma will usually be provided, although in cases of deliberately inflicted or nonaccidental

trauma, this history may be withheld. Specific pathologies to consider include solid organ injuries (splenic and hepatic laceration), perforated viscous, and bleeding with associated retroperitoneal or duodenal hematoma. Although most of these injuries will be symptomatic immediately, some are associated with a delayed presentation. A duodenal hematoma, for example, can gradually accumulate and cause small bowel obstruction (SBO).

Infectious conditions to consider include gastroenteritis (with viral, bacterial, and parasitic pathogens), in addition to nongastrointestinal infections (ie, pneumonia, streptococcal pharyngitis, or urinary tract infections), or adynamic ileus associated with sepsis. A history of particular exposure or infectious contacts may help increase suspicion for infectious gastroenteritis. Although many infections are brief, mild, and self-resolving, some hosts may have comorbid medical conditions that make them more vulnerable to complications such as dehydration or electrolyte abnormalities. In addition, some infections have known serious sequelae including *Clostridium difficile*–associated toxic megacolon, and *Ascaris lumbricoides*–associated bowel obstruction (usually in resource-poor settings).[4]

There is a range of specific gastrointestinal pathologies to consider in a child presenting with vomiting and abdominal pain. In neonates, especially those born preterm, one must be alert for the development of necrotizing enterocolitis. Inguinal hernias can present at any age but are often detected during infancy and are more common in premature infants. These hernias have a high risk of incarceration and require surgical intervention once detected. Malrotation with associated volvulus can present at any age starting with newborns, typically with bilious emesis. Infants and toddlers can present with intussusception as one part of the bowel invaginates into another, causing episodic pain or listlessness. Complications from remnants of the omphalomesenteric duct also typically present in pre–school-aged children. These can include painless rectal bleeding from Meckel diverticulitis and also herniation beneath a fibrous band remnant. Foreign body ingestion (although often asymptomatic) can lead to obstruction and associated pain in young children. In older children, appendicitis becomes a leading differential. Less common differentials to consider at any age include superior mesenteric artery syndrome, bezoar, mesenteric ischemia, intestinal duplication cysts, and mesenteric and omental cysts.[5]

It is important to consider nongastrointestinal pathology as a possible cause of abdominal pain including streptococcal pharyngitis, lobar pneumonia, and diabetic ketoacidosis. In boys, testicular pathology including torsion can present with abdominal pain, making a genitourinary examination mandatory in a boy presenting with abdominal pain, as the history of testicular pain may not be forthcoming.

Finally, there is a range of tumors that can present with pain and distension from an abdominal mass.[6] These can include Wilms tumor, neuroblastoma, rhabdomyosarcoma, and hepatosplenomegaly with leukemia and lymphoma.

CASE PROGRESSION AND DIAGNOSIS

The patient was transported to the Pediatric Intensive Care Unit pending the results of the CT study. A radial artery catheter and an internal jugular venous catheter were placed. Discussion with the radiologist regarding the CT revealed multiple loops of fluid-filled dilated small bowel compatible with a high-grade SBO (Figure 3). In the right upper quadrant, a focal fluid-filled dilated loop of small bowel demonstrated peripheral foci of air, concerning for possible pneumatosis. There was a swirled appearance of the adjacent mesentery, concerning for volvulus. There was mild-moderate ascites but no free air or portal venous gas. No soft tissue mass was visualized. The liver, gallbladder, pancreas, spleen, adrenal glands, and kidneys were all normal. The overall findings were concerning for volvulus or closed-loop obstruction. A specific transition point was not identified.

In light of the patient's CT findings and clinical deterioration, she was taken immediately to the operating room for an exploratory laparotomy. In the operating room, she was confirmed to have a distal ileal volvulus secondary to an internal hernia through a persistent omphalomesenteric duct remnant/band (Figures 4 and 5). She underwent resection of a gangrenous portion of the distal ileum and creation of an ileostomy with 240 cm of small bowel remaining. Her diet was slowly advanced. She had difficulties with large volume ostomy output and was discharged home receiving intravenous fluid replacement through a peripherally inserted central catheter. Approximately 1 month after her initial presentation, she underwent successful takedown of her ileostomy. She continues to be followed in the gastroenterology clinic for complications secondary to short bowel syndrome.

DISCUSSION

Small bowel obstruction may be suspected on clinical examination with emesis, lack of stool output,

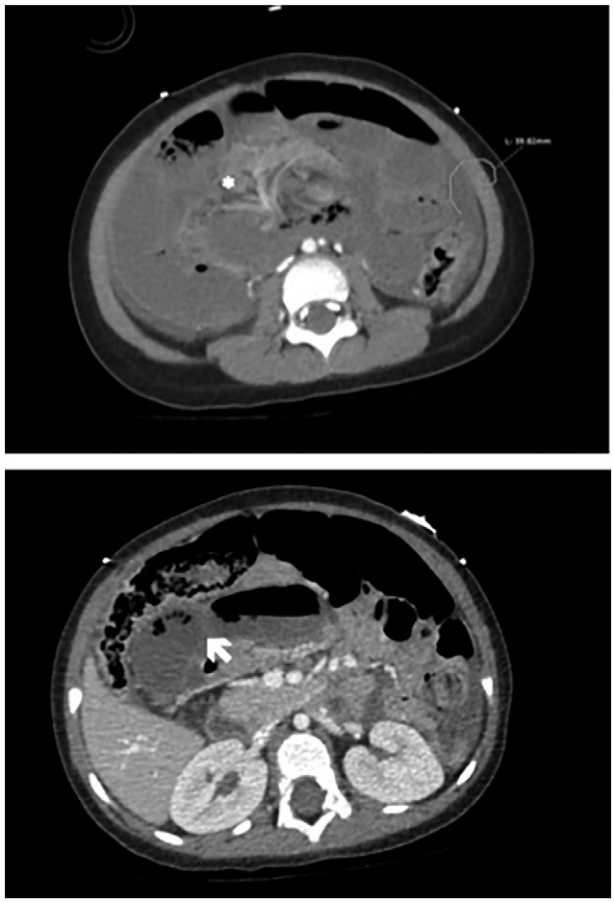

Figure 3 Axial sections of CT study (with contrast) of the abdomen showing the swirling of the mesentery (*) and a fluid-filled loop of bowel with peripheral foci of air concerning for pneumatosis (white arrow).

abdominal distension, and hypoactive bowel sounds. The nature of the emesis (bilious or not) and degree of abdominal distension will vary based on the location of the obstruction in the gastrointestinal tract. Plain radiographs may further offer support for this diagnosis with typical features including distended loops of bowel and air-fluid levels. The most common causes of SBO in pediatric patients are adhesions from previous surgery, inflammation, or trauma.[7] In children without a surgical history, appendicitis is the leading cause of SBO. In our case, the diagnosis of obstruction was more subtle due to a distal closed-loop obstruction. Closed-loop obstruction occurs when the bowel lumen is blocked at 2 contiguous points at a single site, forming an isolated segment of bowel with no outlet either proximally or distally. Closed-loop obstruction can develop because of adhesions, a twist in the mesentery, or internal herniation. The closed loop can then quickly develop strangulation, ischemia, and necrosis.[8] Patients with closed-loop obstruction can have a more benign abdomen until late in the course and lack typical features such as abdominal distension. In addition, closed-loop SBO is more difficult to diagnose on plain radiographs, as there can be a lack of discrepancy in the caliber of proximal and distal bowel loops.[9] The limitations of abdominal radiography for diagnosing SBO are well documented, with a false-negative rate for identifying partial low-grade obstructions of up to 20% and a false-positive rate of 42%.[10] In addition, CT imaging may not allow for these closed loops to be distinguished, and the compromised loop of bowel is not perfused normally initially, leading to a lack of significant acidosis or changes in lactate. Radiographic clues suggestive for a closed-loop obstruction can include a whirl sign (Figure 3) or a beak sign with fusiform tapering of the afferent and efferent limbs of the bowel leading to the obstructed loop. Identification of closed loop SBO is important, as there is a high associated risk of bowel infarction and mortality ranging from 10 to 40%.[11]

Figure 4 Surgical specimen of necrotic bowel.

The presence of a closed loop will often indicate the need for surgical intervention, whereas only 18% of all-comers with SBO otherwise require surgery.[12]

The omphalomesenteric duct is the embryonic structure connecting the primary yolk sack to the embryonic midgut that normally becomes a thin fibrous band and eventually disintegrates and is absorbed spontaneously between 5 and 10 weeks' gestation.[13] If this duct fails to completely atrophy, it may continue to develop. The failure to close and absorb can result in various complications with unique clinical presentations (requiring distinct radiographic diagnostic studies):[14] Meckel, diverticulum with diverticulitis and hemorrhage;[15] bowel

Figure 5 Operative photograph showing internal herniation through a persistent omphalomesenteric duct remnant/band.

obstruction from volvulus around or herniation beneath a fibrous band remnant;[16] leakage of gastrointestinal contents through a patent omphalomesenteric duct tract, and an omphalomesenteric tract cyst presenting as a swelling beneath the umbilicus.[17] The overall prevalence of omphalomesenteric duct remnants is 2% with a 1:1 occurrence in males and females, although symptomatic presentation is more common in males. Of these patients, 60% will remain asymptomatic.[18] Of those who present symptomatically, according to 1 case series, most present before 4 years of age with the mean age of presentation of 2.4 years. Because of the high rate of symptomatic presentations, many surgeons advocate for preemptively resecting remnants if they are discovered incidentally during surgery for other conditions.

There has been increasing utilization of POCUS in pediatric emergency medicine, with an expanding evidence base regarding specific indications for using emergency POCUS.[19] Rapid Ultrasound for Shock and Hypotension (RUSH) is a composite protocol first proposed in 2010 for use in adult patients presenting with undifferentiated hypotension.[20] The RUSH protocol uses a phased array and linear transducer to assess the pump (looking for presence of pericardial effusion, changes in left ventricular contractility, and the relative size of the left and right ventricles), tank (assessing inferior vena cava respiratory dynamics, lungs for fluid, pneumothorax and B-lines, and abdomen for fluid), and pipes (assessment for rupture or obstruction of the abdominal and thoracic aorta, and presence of femoral or popliteal deep vein thrombosis) to look for causes of hypotension. The RUSH protocol has been assessed for interprovider reliability, sensitivity, and specificity in adult patients presenting in uncompensated shock. Some of the disease entities being considered in the RUSH protocol are uncommon in pediatric patients (abdominal aortic aneurysm, for example), and in addition, hypotension is a late presenting symptom in pediatric patients with shock. Pediatric emergency medicine POCUS leaders have noted that more research is needed to develop an evidence-based sonographic assessment tool for pediatric patients presenting with undifferentiated tachycardia and shock.[21]

SUMMARY

Vomiting and abdominal pain are common chief complaints in children presenting for emergency care, and providers must pay careful attention to vital signs and examination findings to determine which children might have a more serious diagnosis than infectious enteritis. Our patient did not have typical findings of SBO but developed uncompensated hypovolemic shock. This prompted a more extensive workup, including utilization of POCUS. As POCUS is being more widely used in pediatric emergency medicine, providers should be aware of the evidence base behind specific studies including the RUSH protocol for evaluation of shock and hypotension. Finally, there is a wide range of potential diagnoses for SBO, especially in children without a history of abdominal surgery, including most commonly appendicitis, hernias, and foreign bodies. Pediatric emergency providers should be aware of the complications relating to persistence of omphalomesenteric duct remnants, including most commonly a Meckel, diverticulum but also bowel obstruction secondary to herniation beneath fibrous band remnants (as presented here), omphalomesenteric duct cysts, and sinus tracts.

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