

Index of Suspicion in the Nursery

3 Abdominal Distention in a Preterm Infant

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PRESENTATION

A 5-day-old, preterm monochorionic-monoamniotic twin girl is brought to the NICU for large-volume, bilious gastric residuals. The infant was born via scheduled cesarean section to a 25-year-old gravida 2, para 1 woman at 32 weeks' gestation because of evidence of cord entanglement on prenatal ultrasonography. The infant weighed 1,390 g at birth and received nasal continuous airway pressure in the delivery room for respiratory distress. The infant starts enteral feedings of 10 mL/kg per day on postnatal day zero and advances per protocol to 70 mL/kg per day on day 5. She was weaned to room air by day 4. Because the mother had difficulty producing breast milk, the infant primarily receives formula. She has negative findings on infectious disease evaluation, with an age-appropriate complete blood cell count and C-reactive protein values in the first 48 hours after birth. The infant has unremarkable physical examination findings until day 5 when the abdominal girth increased by 4 cm (Fig 1). With onset of abdominal distention and new-onset bilious gastric residuals, feedings are stopped, and a gastric tube is placed via suction to decompress the intestine. Blood and urine culture specimens are obtained, and the infant is started on broad-spectrum antibiotics. Serial abdominal radiographs obtained over the subsequent 12 hours demonstrate distended bowel loops and air-fluid levels followed by evidence of free abdominal air but no pneumatosis intestinalis or portal venous gas (Fig 2). The patient develops hypotension, tachycardia, oliguria, and progressive lactic acidosis. She undergoes intubation and fluid resuscitation and is brought to the operating room for exploratory laparotomy.

DISCUSSION

Differential Diagnosis

Abdominal distention accompanied by bilious gastric residuals can represent myriad pathologic diagnoses in the preterm infant, including necrotizing enterocolitis (NEC), spontaneous/focal intestinal perforation (SIP), septic ileus, and malrotation with volvulus. The classic presentation of malrotation with midgut volvulus in a term infant is bilious emesis; however, the presentation is less well-defined in preterm infants.

Patient Course

Intraoperatively, the patient was found to have intestinal malrotation with midgut volvulus with perforation of the midportion of the small intestine. She had

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Figure 1. Clinical presentation with abdominal distention, postnatal day 5.

complete necrosis of the entire midgut, from the duodenum to 2 cm proximal to the ileocecal junction. The abdomen was closed without resection of the devitalized intestine; the infant returned to the NICU for comfort care and died the following day.

The Condition

During normal embryonic development the bowel undergoes sequential patterned rotation resulting in the duodenojejunal junction positioned to the left of midline and fixed posteriorly. Anomalies of intestinal fixation and rotation, described as malrotation, set the stage for volvulus later in life.

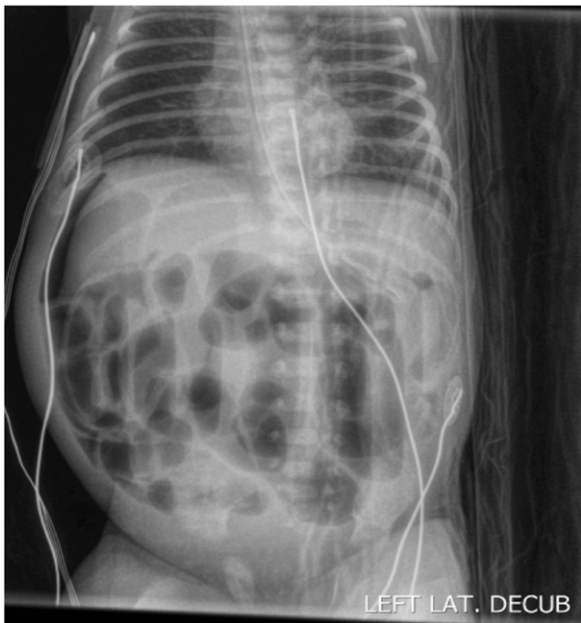


Figure 2. Abdominal radiograph on postnatal day 5, showing free intra-abdominal air.

The classic presentation of malrotation with volvulus in a term infant is bilious emesis; however, the clinical signs and radiographic findings in preterm infants are often nonspecific. A recent retrospective review of preterm infants with malrotation and midgut volvulus listed nonspecific signs including gastric retention, apnea, and abdominal distention in contrast to term infants who presented uniformly with bilious emesis. (1) Preterm infants are at higher risk for developing complex midgut volvulus defined by volvulus requiring bowel resection, intestinal failure, prolonged parenteral nutrition or death, compared with term infants. (2) Furthermore, among the preterm population, it remains difficult to differentiate malrotation with volvulus from other more common gastrointestinal emergencies including SIP and NEC. (1) A single-center retrospective review evaluating the factors leading to diagnosis of malrotation with volvulus versus NEC found bilious emesis or bilious residuals were the only clinical factor significantly associated with volvulus, whereas NEC presented more commonly with systemic illness, grossly bloody stools, thrombocytopenia, and abdominal tenderness. (3) However, clinical signs, laboratory findings, and radiographs cannot reliably distinguish between the 2 pathologies. At one center, a clinical score based on the combination of prematurity (<36 weeks' gestation) plus a base excess below -1.70 in a child with clinical illness reliably predicted midgut volvulus associated with high morbidity and mortality that warranted urgent surgical consultation. (2)

Time is a critical factor in the management of malrotation with volvulus, because a delay in diagnosis can lead to extensive loss of ischemic bowel. Upper gastrointestinal radiographic studies are the gold standard for diagnosis of malrotation with midgut volvulus. (4) However, ultrasonography with Doppler has emerged as an important diagnostic tool that can delineate inversion of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) seen in malrotation. The "whirlpool sign" is a classic sign of malrotation with volvulus and describes the appearance of the mesentery wrapped around the SMA/SMV. (5) A range of abdominal radiographic findings have been described as characteristic of, but not specific for, midgut volvulus, including the "tubular" appearance of bowel secondary to ischemia or a paucity of bowel gas. (6)(7)(8)

Lessons for the Clinician

- Neonatal volvulus in preterm infants is a rare, yet often lethal condition and requires a high index of suspicion. Early diagnosis is crucial to prevent intestinal necrosis and to improve chances of survival.

- Malrotation with midgut volvulus should be included in the differential diagnosis of acute abdominal distention in preterm infants.
- The classic presentation of bilious emesis in the term infant may be more subtle in preterm infants.
- In addition to abdominal radiography, ultrasonography with Doppler can identify compromised flow of the superior mesentery vasculature.
- Metabolic (lactic) acidosis in a preterm infant may be an early diagnostic indicator of intestinal ischemia resulting from malrotation with volvulus. (2)

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