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Fetal malrotation with midgut volvulus: Prenatal diagnosis and planning

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Abstract

Introduction: Malrotation of the intestinal tract is a congenital malformation commonly found either incidentally or after affected individuals develop signs and symptoms of intestinal obstruction. Malrotation is prone to midgut volvulus that can cause intestinal obstruction and lead to ischemia and necrosis requiring emergent surgical intervention. Rare instances of *in utero* midgut volvulus have been reported in the literature and carry a high mortality given the difficulty in establishing a diagnosis prior to development of signs of intestinal ischemia and necrosis. Advancements in imaging have made it possible to diagnose *in utero* malrotation earlier, raising the question of optimal timing of delivery, especially in cases of prenatally diagnosed midgut volvulus. In these cases, the risks of premature birth must be weighed against the risks of fetal intestinal ischemia and potential fetal demise.

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Author contributions

OOO, JH, JG, RMB, MB, SSK, JD, MVM, RD, MAB, AK, PAM, and TCL conceptualized, wrote, and reviewed manuscript content. PAM analyzed imaging included in the manuscript.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Statement of informed consent

Informed consent was obtained from the guardian.

Authorship statement

All authors attest that they meet the current ICMJE criteria for Authorship.

Statement of ethics

This study protocol was reviewed and the need for approval was waived by the Baylor College of Medicine Institutional Review Board.

Case presentation: This case report details an interesting presentation of intestinal malrotation with suspected midgut volvulus found on prenatal imaging at 33 weeks and 4 days' gestation. This prompted delivery of the infant at 34 weeks and 2 days' gestation with urgent operative management, within 3 hours of life, after diagnosis was confirmed postnatally. Intraoperatively, the infant was confirmed to have midgut volvulus without bowel ischemia, the intestines were reduced, and a Ladd procedure was performed without incident. The infant recovered postoperatively without complication, tolerated advancement to full volume feeds and was discharged on day of life 18.

Conclusion: Successful management of fetal malrotation with midgut volvulus may be accomplished by early access to a multi-disciplinary team of professionals, prompt postnatal confirmation of diagnosis, and urgent correction to minimize the risk of complications.

Keywords

Malrotation; Volvulus; Whirlpool sign

1. Introduction

Normal embryonic intestinal development involves the protrusion of bowel through the yolk stalk and counterclockwise 90-degree primary rotation of the bowel about the superior mesenteric artery axis. This is followed by retraction of the bowel into the abdomen and further 180-degree counterclockwise rotation [1]. Intestinal rotation anomalies consist of a spectrum of disorders that range from nonrotation, as seen in some congenital anomalies (such as congenital diaphragmatic hernia and omphalocele), to malrotation. Intestinal malrotation is defined as an error of midgut rotation about the superior mesenteric artery and subsequent return to the peritoneal cavity. Intestinal nonrotation is present in 1 in every 500 births [2]. The exact incidence of malrotation, however, is unknown, as many present in adulthood [3]. Most cases of intestinal malrotation are secondary to incomplete intestinal rotation during development which leads to a narrow distance between the attachments of the midgut mesentery to the posterior abdominal wall and peritoneal band formation. This narrow distance predisposes the neonate to intestinal volvulus [4]. In the neonate, midgut malrotation may present as bilious vomiting secondary to volvulus or duodenal obstruction by peritoneal bands [5]. Malrotation may then be diagnosed via ultrasound, magnetic resonance imaging, upper gastrointestinal series, or by direct visualization during operative management. Midgut malrotation with volvulus detected late in presentation is often associated with bowel necrosis requiring surgical resection [6].

Improvements in imaging have increased our ability to detect midgut volvulus shortly after birth and even prenatally. Intrauterine midgut volvulus has been diagnosed with presence of the sonographic "whirlpool" sign *in utero* [7–9] and is associated with preterm delivery [10] and high rates of *in utero* fetal demise [11–13]. Thus, prenatal diagnosis may help improve outcomes in malrotation with midgut volvulus. This report discusses a case of prenatally diagnosed malrotation with midgut volvulus that led to induction of labor and delivery of a neonate at 34 weeks' gestation. This manuscript was prepared following the CARE guidelines.

2. Case Presentation

A 22-year-old African American G3P2002 with a history of asthma had an uneventful pregnancy until her referral comprehensive anatomy ultrasound at 24 weeks and 4 days by last menstrual period demonstrated a singleton male fetus with mildly increased amniotic fluid index (AFI) of 24.03cm with the largest pocket being 8.92cm. At that time, images were concerning for gastric enlargement and slightly dilated duodenum with no other abnormality. Follow-up ultrasound at 28 weeks and 4 days demonstrated polyhydramnios (AFI:33.53cm, largest pocket 14.17cm) and mild stomach dilation. The patient was then referred to our fetal center for further evaluation.

Fetal ultrasound and magnetic resonance imaging (MRI) evaluation was done at 33 weeks and 4 days, showing resolution of polyhydramnios (AFI: 21cm) with a widely open pylorus, gastric dilation, and dilation of the first and second portions of the duodenum. The third portion of the duodenum traveled abnormally anteriorly and rotated in a clockwise fashion around the superior mesenteric vascular pedicle, with abrupt tapering of the bowel lumen (Figs. 1 and 2). Ultrasound imaging also displayed this duodenal obstruction but was not able to elucidate the T1-hyperintense colonic meconium that terminated in the right upper quadrant seen on MRI, consistent with a right upper quadrant cecum that supports the diagnosis of malrotation (Fig. 3). There was no bowel wall thickening or ascites. Taken together, the fetal MRI findings suggested duodenal obstruction secondary to malrotation with midgut volvulus.

Given the results of the fetal imaging, the patient was admitted with a recommendation for induction of labor at 34 weeks following 2 doses of betamethasone. Pediatric surgery and neonatology teams were made aware at this time, so that they could be present at delivery for potential need for immediate interventions. The neonate was delivered by induced vaginal delivery at 34 weeks and 2 days without complication. The birthweight was 2,530g and 1- and 5-min APGAR scores were 9 and 10, respectively. The infant underwent an unremarkable resuscitation and displayed no evidence of respiratory distress or clinical illness in the delivery room apart from a mildly distended abdomen. An orogastric tube was placed in the delivery room for decompression with 36mL of blood-tinged fluid aspirated from the stomach upon initial placement. The infant was immediately transferred to the neonatal intensive care unit with a plan for fluoroscopic upper gastrointestinal series.

On admission to the neonatal intensive care unit, radiology was notified, and an urgent fluoroscopic upper gastrointestinal series was performed which demonstrated persistent duodenal obstruction with upstream dilation, consistent with the prenatal diagnosis (Fig. 4). Given the pre- and postnatal findings, the patient was intubated and taken immediately to the operating room by 3 hours of life for an exploratory laparotomy. Intraoperatively, malrotation with a narrow mesentery was seen along with a 360° volvulus. There was no bowel ischemia (Fig. 5). The midgut volvulus was reduced in a counterclockwise manner, and a Ladd’s procedure (including appendectomy) was performed.

Neonatal management following surgical correction included close observation and supportive treatment. Close monitoring for sepsis and coagulopathy perioperatively allowed

for early correction prior to the development of complications. The patient was started on total parenteral nutrition (TPN) post-operatively. He was extubated to room air on postoperative day 2. Feeds of donor expressed breast milk were introduced on postoperative day 9 at 20 cc/kg/day and were advanced per unit feeding protocol. TPN was discontinued at day 14, and the infant was tolerating full feeds on day 16. The mother was discharged two days following delivery without complication, and the infant was discharged on the 18th day of life.

3. Discussion

Fetal diagnosis of malrotation with midgut volvulus is rare. Since ultrasound and MRI images were suggestive of bowel obstruction and whirlpool sign was concerning for volvulus, it was felt that the natural history of this patient's malrotation with midgut volvulus may have resulted in significant bowel injury or fetal demise. Imaging at 28 weeks displayed polyhydramnios with gastric and duodenal dilation. The patient was referred to our fetal center for repeat ultrasound paired with fetal MRI at 33 weeks, and though the polyhydramnios had improved, the proximal duodenum was noted to be dilated and a "whirlpool" sign was noted about the superior mesenteric vascular pedicle compatible with midgut volvulus. Currently, midgut volvulus cannot be surgically confirmed *in utero* but represents a surgical emergency when diagnosed postnatally. Thus, our team was faced with the decision of whether to proceed with delivery or continue prenatal monitoring.

Risks of premature delivery at 33 weeks include but are not limited to temperature instability, hypoglycemia, feeding intolerance, respiratory distress secondary to surfactant deficiency, and adverse neurodevelopmental outcomes. However, this was weighed against previous reports of fetal midgut volvulus complicated by meconium peritonitis [14,15], hemorrhagic ascites [16], and fetal demise secondary to midgut volvulus [17,18], along with our knowledge that the presence of a whirlpool (barber pole) sign [15,19–22] is associated with bowel ischemia and increases accuracy of diagnosis of midgut volvulus.

The decision to deliver the infant at 34 weeks following completion of antenatal steroids was multidisciplinary, involving the maternal-fetal medicine, pediatric surgery, radiology, and neonatology teams at a large, tertiary care institution and was discussed by all involved to minimize the most harm to the infant and mother. Fortunately, the fetus was diagnosed at a later gestational age, improving anticipated perinatal complications. However, the care teams were faced with the dilemma that, though prenatal imaging demonstrated midgut volvulus, the polyhydramnios had improved, suggesting intermittent midgut volvulus or non-obstructive malrotation with midgut volvulus. If there was spontaneous resolution of midgut volvulus, labor would have been induced in vain and a child that would have been term could be harmed by exposure to risks of prematurity. Care was made to employ ethical principles of beneficence, non-maleficence, autonomy, and informed consent to ensure that no harm was done to either the mother or the fetus. Informative discussions were had with the mother ensuring full understanding of imaging results, benefits, and risks of preterm induction of labor.

At the time the decision was made to deliver, all teams involved were prepared for the role they would play during and following delivery with cohesive vertical integration of care. The maternal-fetal medicine team determined induction of labor to facilitate vaginal delivery with the pediatric surgery and neonatology teams standing by would be appropriate. This cohesive preparation allowed for diagnosis with upper gastrointestinal series and operative management within 3 h of life. This report is not without limitations in that we report just one case of management of prenatal diagnosis and management of malrotation with midgut volvulus. Furthermore, this report, viewed in conjunction with the current body of literature, provides an additional perspective and multidisciplinary, team-based approach that may benefit future management of this pathology.

As imaging modalities and awareness of this diagnosis improve, we anticipate increased detection of rare congenital anomalies earlier in gestation, which will make decisions surrounding preterm delivery versus expectant management more common. However, with these decisions comes the opportunity to allow fetuses a chance for survival and improved quality of life in cases where prenatal complications lead to high rates of fetal morbidity or demise.

4. Conclusion

Documenting and describing outcomes following preterm delivery for prenatally diagnosed surgical conditions will allow for more nuanced discussions surrounding these decisions amongst providers and parents. In this case of prenatally diagnosed midgut volvulus, we describe a favorable outcome that was accomplished by early access to a multi-disciplinary team of professionals, prompt postnatal confirmation of diagnosis, and urgent correction to minimize the risk of complications.

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Data availability statement

No data or other patient health record information was obtained in the formation of this manuscript.

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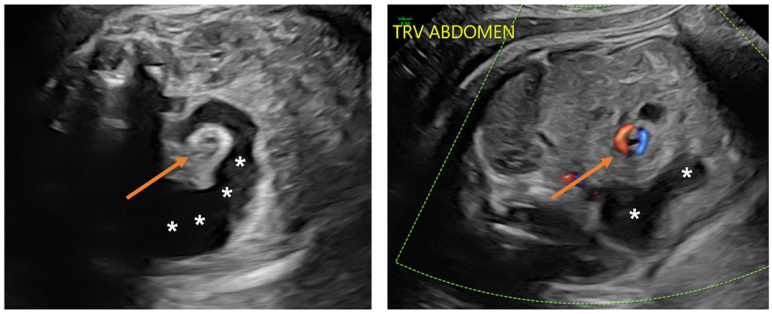

Fig. 1.

Ultrasound images at 33 weeks, 4 days demonstrate dilation of the 2nd and 3rd portions of the duodenum (asterisks), abruptly tapering as they swirl around the superior mesenteric vascular pedicle (arrows).

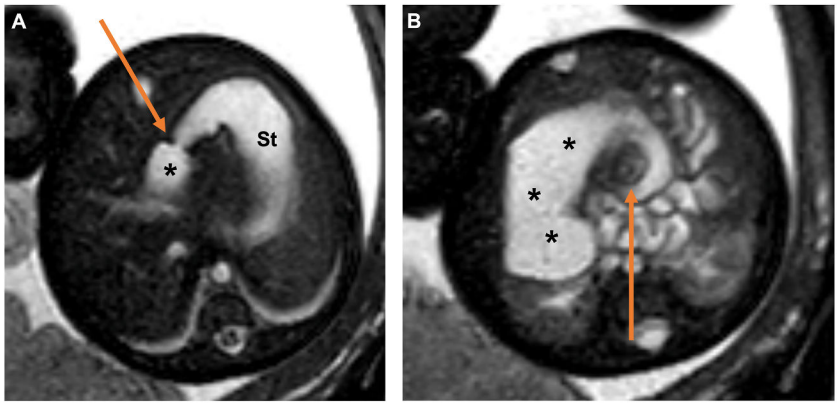

Fig. 2.

Axial balanced turbo field-echo fetal magnetic resonance images at 33 weeks, 4 days demonstrate A) a widely open pylorus (arrow) with dilation of the stomach (St) and B) duodenum (asterisks), abruptly tapering as they swirl around the superior mesenteric vascular pedicle (arrow).

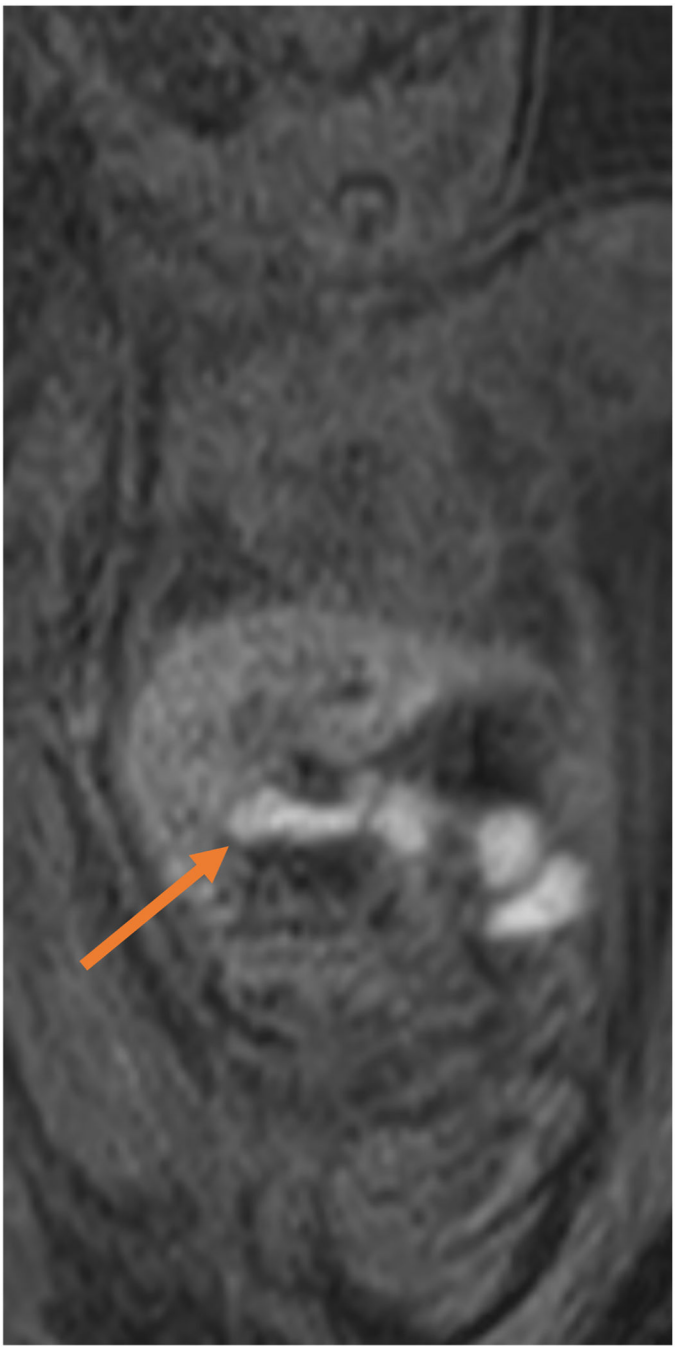

Fig. 3.
Coronal T1 weighted with fat saturation fetal MR image at 33 weeks, 4 days demonstrate a right upper quadrant position of the cecum (arrow).

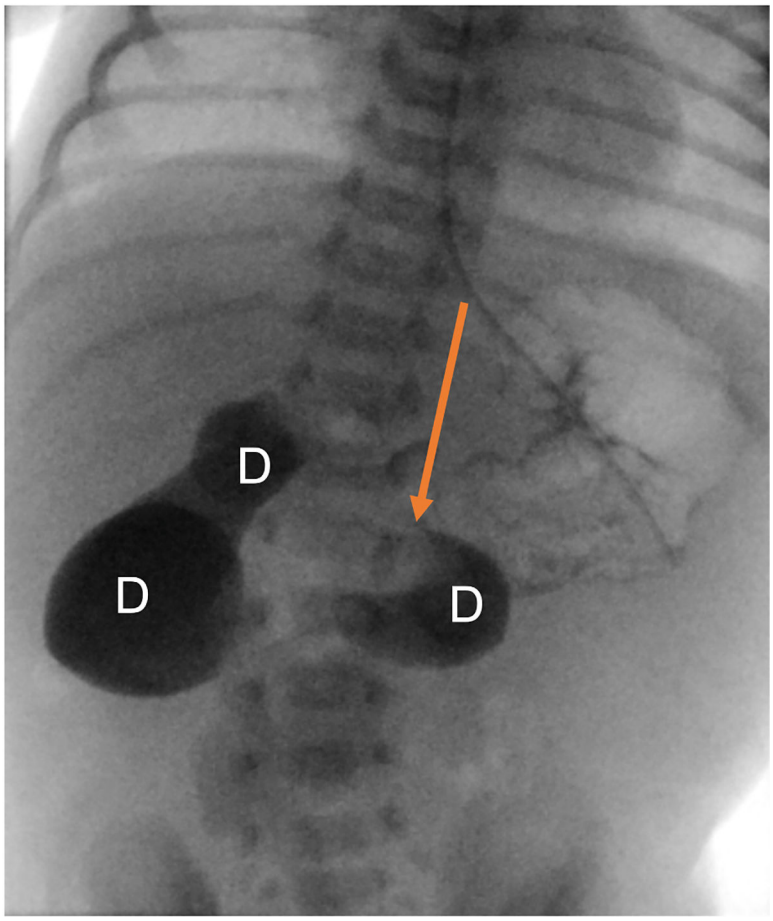

Fig. 4.

Frontal fluoroscopic image from an immediate postnatal upper GI exam redemonstrates duodenal dilation (D) and abrupt obstruction of D3 (arrow) without passage of contrast beyond that, consistent with the prenatal diagnosis of malrotation with midgut volvulus.

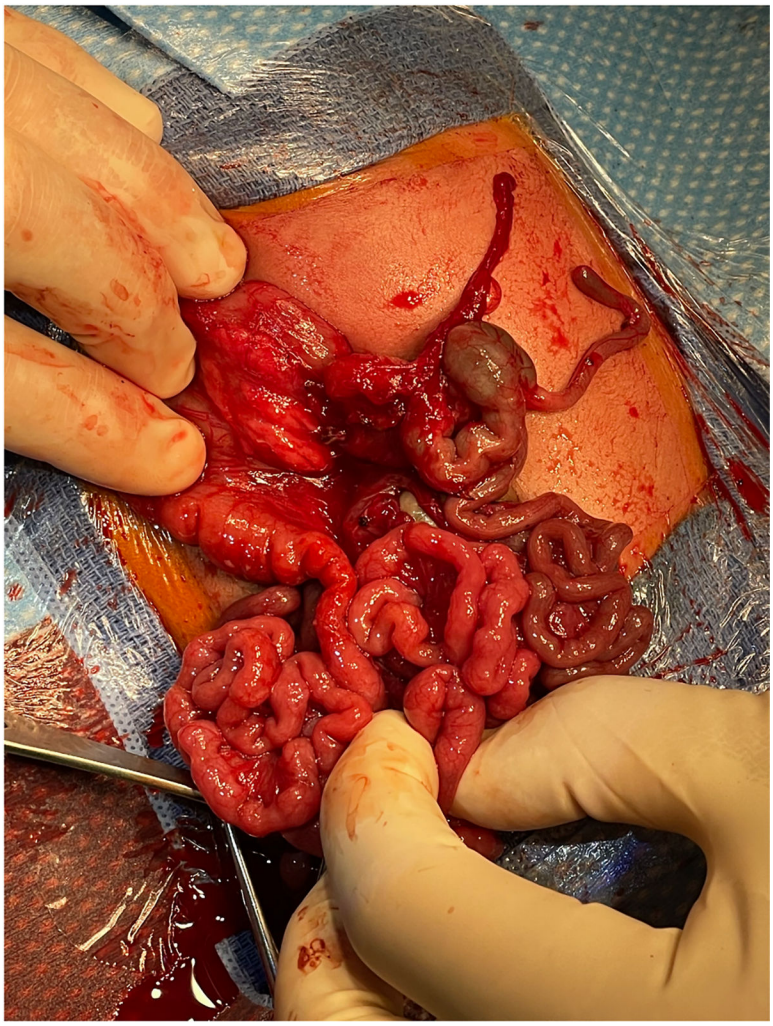

Fig. 5.
Non-ischemic bowel following reduction of midgut volvulus.