Perinatal Survival of a Fetus with Intestinal Volvulus and Intussusception: A Case Report and Review of the Literature

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Fetal intestinal volvulus is a rare life-threatening condition. Late diagnosis of volvulus contributes to high rate of morbidity and mortality. It has variable degrees of presentation and survival. Intrauterine volvulus may be complicated by intestinal atresia due to ischemic necrosis. To our knowledge, there are three reported cases of term fetal demise. We report a case of fetal intestinal volvulus with perinatal survival of the largest term infant described with this complication to date. The volvulus was associated with type 3A jejunal atresia and intestinal pathology was noted on prenatal ultrasound. The infant was born via urgent cesarean delivery at 37\(\frac{6}{7}\) weeks of gestation and underwent emergent exploratory laparotomy with resection of small bowel and primary end-to-end anastomosis. Intrauterine intestinal volvulus may be suspected on prenatal ultrasound but only definitively diagnosed postnatally. Signs of fetal distress and volvulus are rarely associated with reports of survival in the term fetus. We review reported cases of prenatally suspected volvulus in infants documented to survive past the neonatal period. As fetal volvulus and most intestinal atresias/stenoses manifest during the third trimester, we recommend that the limited fetal anatomical survey during growth ultrasounds at 32 to 36 weeks routinely include an assessment of the fetal bowel.
Follow-up ultrasound to assess interval growth at 36\(^{2/7}\) weeks revealed fetal mega-cisterna magna at 14 to 16 mm and striking tubular dilatation of the lower digestive tract concerning for complete intestinal obstruction. At that time, the differential diagnosis entertained the following pathological entities: jejunal atresia, ileal atresia, colonic atresia, meconium ileus, intestinal volvulus, imperforate anus, Hirschsprung disease (aganglionic megacolon), and meconium peritonitis. Umbilical artery velocimetry was normal, and neither Doppler interrogation of the middle cerebral artery nor brain-sparing reflex activation found any evidence of fetal anemia (peak systolic velocity, 0.97 multiples of the median). Appropriate interval fetal growth was noted (60th percentile to 51st percentile between the two scans); the placenta and uterus were unremarkable and there was normal amniotic fluid volume (amniotic fluid index = 20.5 cm). Interestingly, the standard abdominal circumference views were normal (\(\text{Fig. 2}\)) and the intestinal dilatation was noted by the examining sonographer on a more caudal plane. Prolonged external fetal monitoring and testing on that day was reassuring (biophysical profile was 10/10) and cautious outpatient management was continued with twice weekly fetal surveillance until planned prelabor repeat cesarean delivery by 39\(^{0/7}\) weeks. Prenatal neonatology and pediatric surgical consultations were initiated.

The gravida presented to the obstetrical triage at 37\(^{6/7}\) weeks with decreased fetal movement for the past 2 days. Cardiotocography revealed minimal short-term variability and repetitive late decelerations that failed to respond to intrauterine resuscitation. A male infant of 3,353 g was born via urgent cesarean delivery due to non-reassuring fetal heart tones. At delivery, there was meconium-stained amniotic fluid and a single nuchal cord that was reduced without difficulty. Apgar scores were 1, 5, and 7 at 1, 5, and 10 minutes, respectively, and abdominal distention was immediately noted on neonatal examination.

The infant was found to have increasing abdominal tenderness and distension in the ensuing hours. An orogastric tube was placed and yielded dark and then bloody secretions. Initially, the neonate had spontaneous breathing but subsequently developed periods of apnea and desaturations requiring intubation. Umbilical venous blood showed pH of 7.22, PCO\(_2\) of 44, PO\(_2\) of 49, bicarbonate of 17.2, and base deficit of 9 (umbilical artery specimen clotted) and initial glucose was < 20 mg/dL. Initial complete blood count revealed anemia with hemoglobin of 10.8 g/dL, platelet clumping with normal count (183,000/μL), and leukocytosis at 25,600 WBC/μL predominantly lymphocytic (54%). Because of pediatric concern for hypoxic ischemic encephalopathy (HIE), the infant was placed on a head-cooling protocol with induced hypothermia for brain protection.

Upon transfer to a quaternary neonatal intensive care unit for further specialized management, the neonate was found not meeting the HIE criteria and was gradually rewarmed in anticipation of exploratory laparotomy. Chest X-ray revealed ascites, KUB (kidney, ureter, and bladder X-ray) revealed small stomach bubble with no gas in bowel, and bedside abdominal ultrasound revealed multiple distended loops of small bowel filled with fluid. An urgent upper gastrointestinal series revealed the duodenojejunal junction to the right of the midline consistent with malrotation without radiographic evidence of obstruction. Emergent laparotomy was performed for suspicion of intestinal malrotation on the first day of life. Surgical exploration revealed markedly distended ischemic jejunum with about a 240-degree volvulus adjacent to an area of a “type 3A” intestinal atresia (complete separation of the two blind bowel ends by a mesenteric defect). On the distal end of the atresia, there was a 57-cm segment of small bowel-to-small bowel intussusception. Approximately 50 cm of bowel distal from the intussusception to the ileocecal valve was found to be patent. The entire colon appeared normal. The proximal bowel was distended and there was 17 cm of healthy proximal bowel from the orthotopic ligament of Treitz down to the area of bowel ischemia. Surgical resection of the small bowel volvulus and intussusception, with primary end-to-end small bowel anastomosis, were performed and the infant recovered well and was discharged from the hospital on 29 days of life. Pathological
Intestinal volvulus is suspected. In association with ascites on prenatal ultrasound and fetal assessment for fetal anemia when dilated loops of bowel are noted in the case report recommended the use of Doppler velocimetry to detect the presence of ischemia.\footnote{Ojuoba et al \textit{et al} propose sonographic examination of the umbilical cord in fetuses with intestinal atresia to identify those at risk of hemorrhage.\textsuperscript{10}} When the elongating intestine returns to the abdominal cavity, the midgut loop rotates 90 degrees counter-clockwise around the axis of the superior mesenteric artery (SMA). This brings the duodenojejunal loop to the right and the ileocolic loop to the left side. During the 10th to 11th weeks, the intestines return to the fetal abdominal cavity. Both the proximal and distal loops undergo a total of 270 degrees of rotation. The duodenojejunal loop ends up posterior to the SMA and the ileocolic loop anterior to the SMA. The duodenojejunal junction lies in the left upper quadrant of the abdomen and the ileocecal junction in the right lower quadrant.\footnote{Nijagal et al propose sonographic examination of the umbilical cord in fetuses with intestinal atresia to identify those at risk of hemorrhage.\textsuperscript{10}}

Volvulus with malrotation may result in variable degrees of ischemic necrosis.\footnote{American Journal of Perinatology Reports Vol. 3 No. 2/2013} The bowel loops twist around the fetal mesenteric artery or its branches resulting in intestinal obstruction and vascular compromise, which can cause necrotic bowel perforation or even hemorrhagic ascites, anemia, and fetal demise.\footnote{Perinatal Survival of Fetus with Midgut Volvulus Ojuoba et al} Midgut volvulus involves the entire bowel from the ligament of Treitz to the mid-transverse colon and the prognosis is very poor.

Sonographic findings of intestinal volvulus were first reported in 1983.\footnote{Perinatal Survival of Fetus with Midgut Volvulus Ojuoba et al} Prenatal diagnosis and sonographic findings suggestive of in utero volvulus are important for multi-disciplinary planning at the time of delivery. In addition to dilated loops of bowel, the “whirlpool sign” (bowel loops with accompanying mesentery and vessels wrapping around the main SMA) and the “coffee bean” or kinked loop sign (which is a distention of a very short segment of bowel, which resembles a coffee bean) are signs of pathognomonic for intestinal volvulus (\textsuperscript{7} Fig. 3). Polyhydramnios has been a sign consistent with complete obstruction from intestinal volvulus.\footnote{Ojuoba et al} One case report recommended the use of Doppler velocimetry to assess for fetal anemia when dilated loops of bowel are noted in association with ascites on prenatal ultrasound and fetal intestinal volvulus is suspected.\footnote{Perinatal Survival of Fetus with Midgut Volvulus Ojuoba et al} Fetal magnetic resonance imaging may be helpful in establishing the exact site of obstruction prenatally; however, its cost and sparse availability of required expertise make this imaging modality impractical for most pregnancies—especially as the diagnosis is typically acutely suspected close to delivery. Postnatally, plain radiography is sufficient, and upper barium studies are rarely needed.\footnote{American Journal of Perinatology Reports Vol. 3 No. 2/2013}

The sonographic findings of fetal intestinal intussusception at term may include a “target-like” lesion; Shimotake reported that this is a round hyperechoic area of fetal bowel mucosa surrounded by relatively hypoechoic ring of bowel wall.\footnote{Perinatal Survival of Fetus with Midgut Volvulus Ojuoba et al} Intussusception can lead to vascular impairment and bowel necrosis resulting in intestinal atresia; similarly, fetal intestinal volvulus may be complicated by ischemia and can lead to intestinal atresia.\footnote{Perinatal Survival of Fetus with Midgut Volvulus Ojuoba et al} Clinically, sluggish fetal movements or the observation of a nonreactive tracing on cardiotocography helps in reaching the diagnosis of volvulus.\footnote{Perinatal Survival of Fetus with Midgut Volvulus Ojuoba et al} Fetal distress such as decreased fetal movement and minimal/absent short-term heart rate variability can be presenting signs, as in our

![Image 305x567 to 533x738](image1)

![Image 306x75 to 533x249](image2)

![Image 306x75 to 533x249](image2)

Fig. 3 “Coffee bean” sign at 36 weeks of gestation sonogram.

![Image 306x75 to 533x249](image2)

Fig. 4 Volvulus at 36 weeks of gestation showing striking tubular dilatation, suggested the development of fetal intestinal pathology.
Table 1  Perinatal and neonatal findings of fetal volvulus that survived past the neonatal period

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Study</th>
<th>Age (wk)</th>
<th>Delivery</th>
<th>Sex</th>
<th>Weight (g)</th>
<th>Fetal ultrasound findings</th>
<th>Procedure</th>
<th>Alive at age</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Molvarec et al</td>
<td>32</td>
<td>Cesarean</td>
<td>F</td>
<td>2,150</td>
<td>Dilated small bowels, mild polyhydramnios</td>
<td>Jejunoileostomy</td>
<td>28 mo</td>
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<tr>
<td>2</td>
<td>Molvarec et al</td>
<td>36</td>
<td>Cesarean</td>
<td>M</td>
<td>2,350</td>
<td>Dilated small bowels</td>
<td>Ileoileostomy</td>
<td>16 mo</td>
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<td>3</td>
<td>Durand et al</td>
<td>34</td>
<td>Cesarean</td>
<td>F</td>
<td>2,180</td>
<td>Dilated loops of bowel</td>
<td>Unknown</td>
<td>10 y</td>
</tr>
<tr>
<td>4</td>
<td>Durand et al</td>
<td>39</td>
<td>Cesarean</td>
<td>F</td>
<td>2,660</td>
<td>Dilated loops of bowel</td>
<td>Unknown</td>
<td>15 mo</td>
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<td>5</td>
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<td>34</td>
<td>Cesarean</td>
<td>F</td>
<td>2,190</td>
<td>Dilated loops of bowel</td>
<td>Unknown</td>
<td>5 mo</td>
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<td>6</td>
<td>Leung et al</td>
<td>34</td>
<td>Vaginal</td>
<td>F</td>
<td>3,000</td>
<td>Ascites meconium peritonitis</td>
<td>Unknown</td>
<td>1 mo</td>
</tr>
<tr>
<td>7</td>
<td>Crisera et al</td>
<td>37</td>
<td>Vaginal</td>
<td>U</td>
<td>N/A</td>
<td>No polyhydramnios</td>
<td>Jejunostomy</td>
<td>9 wk</td>
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<tr>
<td>8</td>
<td>Usmani and Kenigsberg</td>
<td>29–30</td>
<td>Vaginal</td>
<td>F</td>
<td>1,200</td>
<td>Polyhydramnios</td>
<td>Bowel resection</td>
<td>7 y</td>
</tr>
<tr>
<td>9</td>
<td>Samuel et al</td>
<td>34</td>
<td>Cesarean</td>
<td>U</td>
<td>N/A</td>
<td>Unknown</td>
<td>Ileostomy, colostomy</td>
<td>2 mo</td>
</tr>
<tr>
<td>10</td>
<td>Black et al</td>
<td>36</td>
<td>Cesarean</td>
<td>M</td>
<td>2,860</td>
<td>Unknown</td>
<td>Anastomosis</td>
<td>2 y</td>
</tr>
<tr>
<td>11</td>
<td>Black et al</td>
<td>34</td>
<td>Undocumented</td>
<td>M</td>
<td>2,170</td>
<td>Dilated loops, aperistaltic</td>
<td>Anastomosis</td>
<td>18 mo</td>
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<td>12</td>
<td>Has and Gunay</td>
<td>34</td>
<td>Vaginal</td>
<td>M</td>
<td>2,000</td>
<td>Polyhydramnios, ascites, dilated loops of bowel</td>
<td>Bowel resection</td>
<td>36 d</td>
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<tr>
<td>13</td>
<td>Park et al</td>
<td>33</td>
<td>Vaginal</td>
<td>M</td>
<td>2,790</td>
<td>Dilated loops of bowel, coffee bean sign</td>
<td>Anastomosis</td>
<td>4 mo</td>
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<tr>
<td>14</td>
<td>Noreldeen et al</td>
<td>31</td>
<td>Cesarean</td>
<td>M</td>
<td>1,860</td>
<td>Dilated loops of bowel ascites</td>
<td>Anastomosis</td>
<td>1 y</td>
</tr>
<tr>
<td>15</td>
<td>Yu et al</td>
<td>37</td>
<td>Undocumented</td>
<td>F</td>
<td>2,900</td>
<td>Polyhydramnios, whirlpool sign, dilated loops of bowel</td>
<td>End-to-end anastomosis</td>
<td>28 mo</td>
</tr>
<tr>
<td>16</td>
<td>Current case</td>
<td>37</td>
<td>Cesarean</td>
<td>M</td>
<td>3,353</td>
<td>Dilated loops of bowel</td>
<td>Bowel resection anastomosis</td>
<td>24 mo</td>
</tr>
</tbody>
</table>

Abbreviations: F, female; M, male; N/A, not available; U, unknown.
Heterozygosity for mutations at perinatal survival at 39 weeks of a fetus with compound arthrogryposis. Midgut volvulus; the fetus also had intestinal atresia and gestation, due to cardiovascular failure and shock from fensen et al reported a case of fetal demise at 38 weeks of a fetus with compound arthrogryposis. Midgut volvulus; the fetus also had intestinal atresia and gestation, due to cardiovascular failure and shock from fensen et al reported a case of fetal demise at 38 weeks of gestation; autopsy in that case revealed intestinal volvulus with extended hemorrhagic infarction of the small bowel but no perforation. The fetus was notably affected by cystic fibrosis—characterized by viscous mucous secretions that can obstruct the bowel lumen, thus predisposing to volvulus. In contrast, Durand et al reported a case of perinatal survival at 39 weeks of a fetus with compound heterozygosity for mutations at AF508 and Y1092X. Steffensen et al reported a case of fetal demise at 38 weeks of gestation, due to cardiovascular failure and shock from midgest volvulus; the fetus also had intestinal atresia and arthrogryposis. Lee et al reported a case at 38 weeks of fetal ileal atresia and volvulus; however, it is not clear if that infant survived the end-ileostomy repair. Various etiologies for intestinal volvulus have been described in infants alive in the neonatal period, including meconium peritonitis and intestinal mesentery defects. Pathognomonic sonographic findings in the third trimester ultrasound examination, such as the whirlpool sign, aids in rapid antenatal diagnosis of intestinal volvulus. Characteristics of these cases are summarized in Table 1.

Currently, the American College of Obstetricians and Gynecologists and the American College of Radiology recommend routine sonograms in the first and/or second trimester, but state that no evidence exists to recommend routine sonograms in the third trimester. Our case involves fetal volvulus, intussusception, and intestinal atresia with perinatal and long-term survival of a term infant. Cases reviewed of survival of fetal intestinal volvulus demonstrate that prenatal diagnosis and neonatal anticipation affect outcomes decisively. Molvarec et al reported two cases of fetal intestinal volvulus, which were delivered promptly on the basis of ultrasound findings and biophysical monitoring of the fetus. Bawa and Kannan demonstrate different outcomes of fetal intestinal volvulus based on the performance of prenatal scanning; one fetus identified with dilated bowel underwent prompt surgical intervention and survived, whereas another succumbed with the postnatal diagnosis of volvulus when no prenatal sonogram was performed. They suggested that a single third trimester scan for fetal anomalies may be an effective strategy to reduce perinatal mortality.

Sonographic detection of dilated fetal bowel usually represents a normal variant without clinical significance. Cystic fibrosis (meconium ileus), intestinal obstruction/ataresia, and cloacal malformations make up the majority of pathological conditions associated with dilated fetal bowel; these conditions should also prompt prenatal pediatric surgery and genetics consultations and management in a tertiary care center. For the general obstetrician gynecologist, it is important to note that severe and abrupt fetal bowel dilatation in a previously normal fetus with hyperperistalsis and poor fetal activity are strongly associated with fetal volvulus. This can result in a poor prognosis for perinatal outcome without prompt and appropriate intervention.

The main barrier to routine third trimester sonography is cost-effectiveness—there is no doubt that some urological and intestinal anomalies would be identified with likely improvement of perinatal outcomes, but the cost of screening the entire population in the third trimester renders this approach unattractive from a public health perspective. Our case and the related literature suggest that particular attention should be paid to the fetal digestive tract at the time of fetal growth sonograms after 32 weeks. In a society of delayed childbearing, evolving obesity epidemic and associated comorbidities (chronic hypertension, diabetes mellitus), as many as two-thirds of all pregnant women will have an indication for a third trimester scan—we suggest that this is an opportunity to screen for fetal intestinal anomalies that could substantially affect perinatal care and avert late fetal loss or serious neonatal morbidity. The new onset of striking tubular intestinal dilation in the third trimester in combination with clinical suspicion for fetal intestinal volvulus should prompt multidisciplinary consultation and planning with neonatology and pediatric surgery to help optimize perinatal outcomes in a tertiary care hospital.

References
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Perinatal Survival of Fetus with Midgut Volvulus  Ohuoba et al.