American Pediatric Surgical Association

Prenatal Counseling Series

Intestinal Atresia and Stenosis

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from the
Fetal Diagnosis and Treatment Committee

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FD&T Committee

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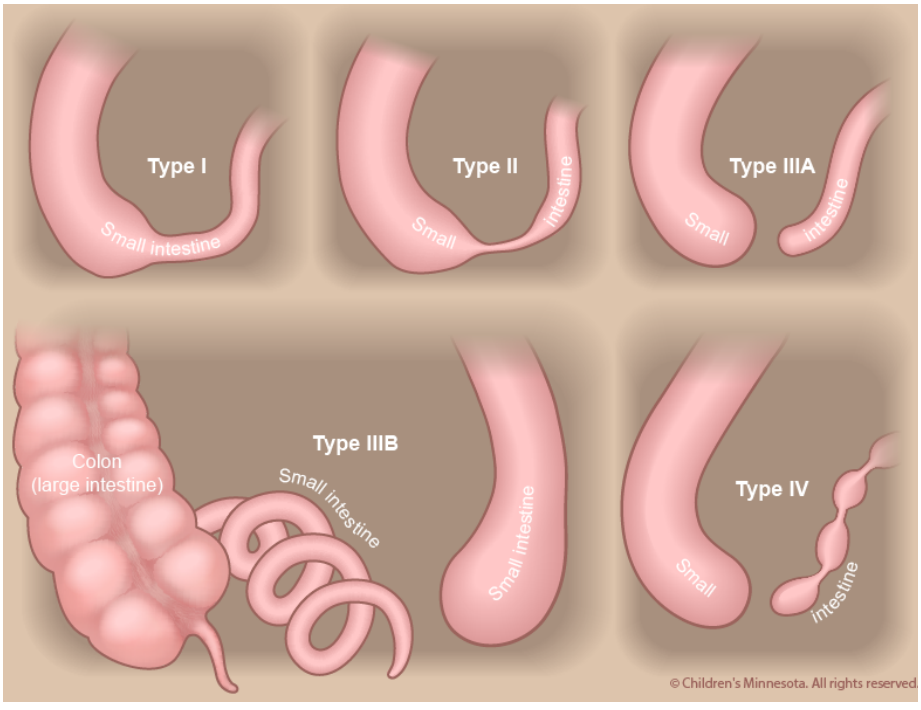
Intestinal Atresia and Stenosis

Intestinal atresia refers to congenital obstruction at any point in the intestinal tract, whereas intestinal stenosis typically refers to the narrowing of the intestinal lumen.

- Overall, intestinal atresias occur in nearly 1 in 5,000 live births.[1,2]
- Atresia of the proximal jejunum and distal ileum is the most common location and is more likely to be detected on prenatal imaging.
- Atresias are thought to result from in utero vascular compromise to the intestines.[3]
  - The etiology of duodenal atresia is unique in that it is thought to develop from failure of recanalization
- Cystic fibrosis is associated with up to 10% of cases.[4]
- All intestinal atresias and stenosis require surgical intervention to correct the obstruction.



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Differential Diagnosis

Differential diagnosis of prenatally diagnosed intestinal atresia and stenosis includes: malrotation with or without volvulus, meconium ileus, intestinal duplication, colonic atresia, total colonic aganglionosis (Hirschsprung disease), and anorectal malformations.

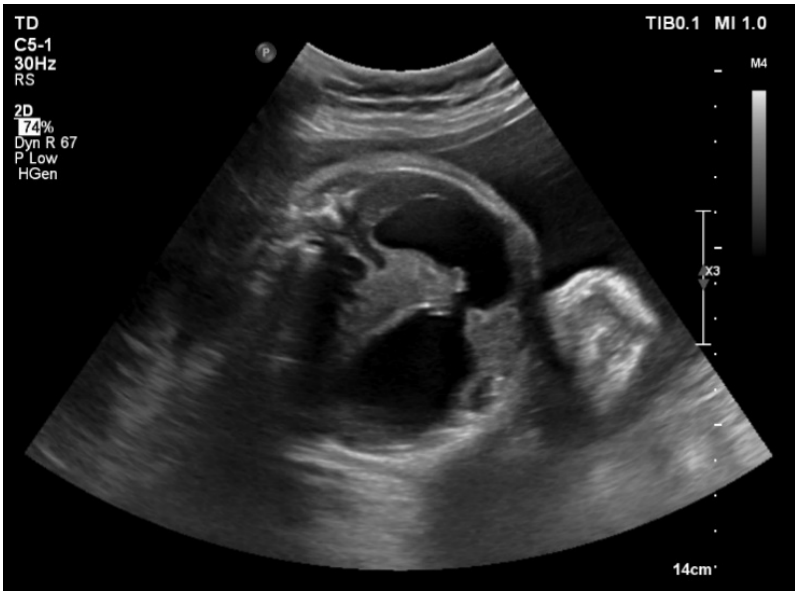
Initial Evaluation

Fetal anatomic ultrasound
Fetal magnetic resonance imaging (may not be offered at all centers)
Amniocentesis for karyotype
Cystic fibrosis DNA mutation analysis

Intestinal atresia is being increasingly diagnosed on prenatal ultrasound and MRI. The goal of prenatal imaging is to identify the level of the atresia, evaluate for additional congenital anomalies, exclude alternative diagnoses, and guide perinatal and postnatal management.

Fetal Anatomic Ultrasound[5]

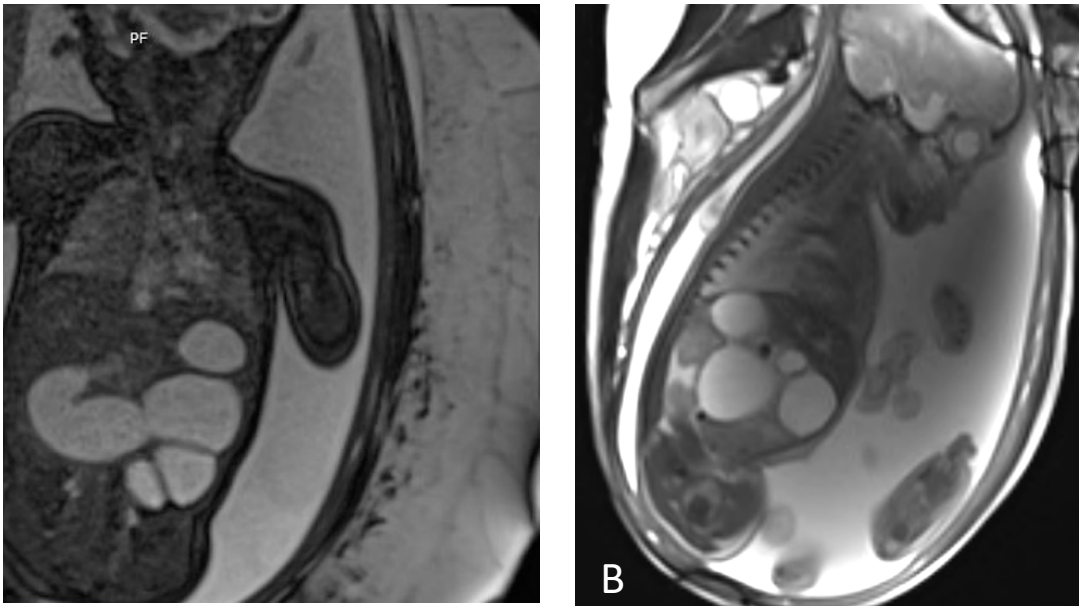
- Evaluate fetal growth.
- Evaluate the amount of amniotic fluid. Proximal atresias (pyloric and duodenal atresias) have a greater likelihood of developing polyhydramnios in the third trimester, which can predispose to preterm labor and delivery. On the other hand, the more distal the atresia, the less likely it is for polyhydramnios to develop.
- Evaluate for other congenital anomalies. Although rare to have other anomalies, if present, they are generally limited to the gastrointestinal tract and may include malrotation, intestinal duplication, and esophageal atresia.



Ultrasound imaging demonstrating double-bubble sign. Courtesy of Dr. Marjorie Treadwell, University of Michigan.

Fetal Magnetic Resonance Imaging (MRI)[6]

- MRI aids in characterizing the atresia and in demonstrating the anatomy of the remainder of the bowel.
- It is not routinely offered but can be used as an adjunct to ultrasound.



MRI of fetus demonstrating dilated loops of small bowel with associated polyhydramnios. Image A courtesy of Dr. Amir Alhajjat, and Image B courtesy of Dr. Brian Gray, University of Indiana,

Amniocentesis for Fetal Karyotype

- If echogenic bowel is present at time of ultrasound evaluation, performing amniocentesis should be considered along with cystic fibrosis DNA mutation analysis.[4]

Prenatal Counseling and Postnatal Considerations

After comprehensive review of the fetus with anatomic ultrasound, fetal MRI, and amniocentesis for karyotyping, comprehensive counseling can be performed and typically includes maternal-fetal medicine and pediatric surgery.

Prenatally diagnosed intestinal atresias are more likely to be severe compared to atresias diagnosed postnatally, even in the absence of other anomalies. Newborns with prenatally diagnosed intestinal atresia may have an increased risk of longer hospital course (including lengthier intensive care unit stay), need for intravenous nutrition, prolonged central venous access, and risk of short gut syndrome.

Fetal Intervention

At this time, there are no fetal interventions for for intestinal atresia and stenosis.

Management of Pregnancy and Delivery Planning

- Following the fetal diagnosis of intestinal atresia, further investigations will be required including a complete fetal anatomic ultrasound. If no other anomalies are identified, no further workup is required.
- Delivery should be anticipated at a center with high-risk obstetric, neonatal, and pediatric surgery support. A suspected diagnosis of intestinal atresia should not impact the method of delivery (vaginal versus Cesarean section).
- Management of intestinal atresia and stenosis immediately following birth include orogastric decompression and intravenous fluids for resuscitation and correction of any electrolyte abnormalities.
- If a prenatal evaluation for cystic fibrosis was not performed, a sweat chloride test (when the baby is of appropriate age and weight) or cystic fibrosis DNA mutation analysis should be ordered.

Postnatal Course

In general, outcomes in infants with intestinal atresia and stenosis are excellent. All intestinal atresias and stenoses require surgical management, though rarely emergently. Most newborns can be managed with a single elective operation during which the atresia is resected, and bowel anastomosis is performed.

Occasionally, there will be multiple atresias found, which may compromise intestinal length. In instances where significant bowel length has been lost, sequelae of short bowel syndrome will prolong hospitalization and complicate recovery. Long-term follow-up by a pediatric surgeon and pediatric gastroenterologist is recommended.

Postnatal Risks to discuss with families during prenatal consultation:

The following long-term outcomes should be explicitly discussed during the prenatal consultation for a fetus with intestinal atresia:

- Prolonged postoperative ileus with proximal atresia[7]
- Gastrostomy tube dependence[2]
- Failure to thrive
- Vitamin deficiencies (B12, if with loss of distal ileum)
- Short bowel syndrome (possible total parenteral nutrition dependence)[8]
- Pulmonary morbidity (if cystic fibrosis diagnosed)[4]
    - If the infant is tested cystic fibrosis positive, parental testing will be required as this could impact future pregnancy considerations
- Death[8,9]

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