American Pediatric Surgical Association

Prenatal Counseling Series

Gastroschisis

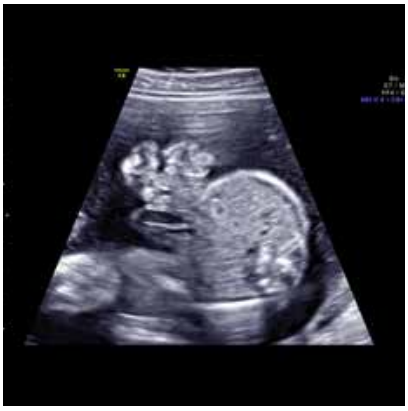
APSA
American Pediatric
Surgical Association
Saving Lifetimes™

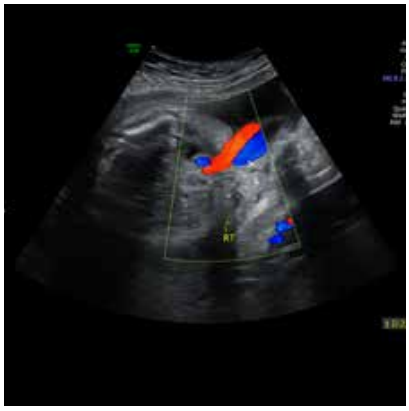
from the
Fetal Diagnosis and Treatment Committee
of the American Pediatric Surgical Association
Editor-in-Chief: Ahmed I. Marwan, MD

Special thanks to: Oliver Muensterer, MD,
and Jill Stein, MD

Definition and Differential Diagnosis

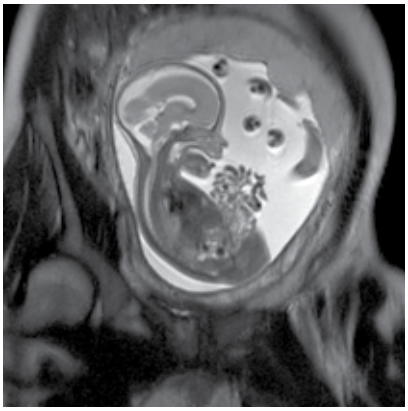
- Gastroschisis is a condition in which the abdominal wall does not form correctly during development.
- It results in a defect of varying size mostly to the right side of where the umbilical cord attaches to the baby.
- Protruding organs may include: small bowel, together with large bowel, stomach, liver, bladder, as well as the ovaries and fallopian tubes in females.





Axial ultrasound images show an abdominal wall defect with multiple loops of non-dilated bowel located external to the fetal abdomen. The defect is most commonly located to the right of the umbilicus. No overlying membrane is seen. *Courtesy of Jill Stein, MD – Colorado Fetal Care Center – Children's Hospital Colorado*

- In contrast to omphalocele, a similar condition that occurs in the midline of the baby rather than the right side, the organs in gastroschisis are not covered by a sack or membrane.



Sagittal MRI image of a 22-week-gestation fetus with an abdominal wall defect adjacent to the umbilicus. Multiple loops of non-dilated bowel are protruding external to the abdomen without overlying membrane. *Courtesy of Jill Stein, MD – Colorado Fetal Care Center – Children's Hospital Colorado*

- Outer surface of the bowel is exposed to the amniotic fluid, which usually leads to inflammation and thickening of the wall, causing the bowel not to work correctly for several weeks after birth, even if the protruding content is brought back to the belly and the defect is closed. During this time, the newborn baby needs to be fed intravenously rather than through the gut.
- Gastroschisis happens in as many as 1 out of 2,000 births.
- The risk factors are maternal young age and smoking. Overall, the incidence seems to have increased over the last decades.

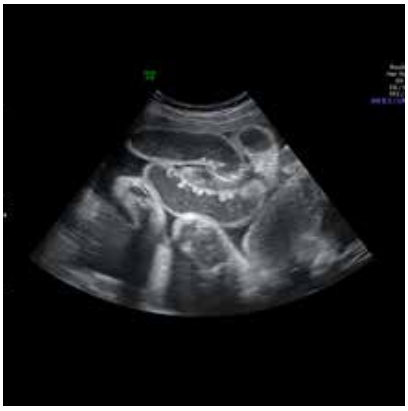
Prenatal Diagnosis

- Gastroschisis can be detected by prenatal ultrasound in as early as the 12th week of pregnancy. Typically, the defect is detected to the right side of the cord insertion.
- It is mostly an isolated defect, however associated intestinal atresia may be seen.
- Alfa-Fetoprotein (AFP) levels in the blood are usually evelated in mothers carrying a fetus with gastroschisis. Most often, there are no specific genetic anomalies detected in affected babies.

Prenatal Considerations

Once a prenatal diagnosis of gastroschisis is established, birth should be planned in a specialized center that incorporates high-risk obstetrics, neonatology and pediatric surgery. Some babies with gastroschisis are growth restricted, and some are born prematurely. The goal, however, should be to allow the pregnancy to progress to 38 weeks before birth. Current studies suggest that babies with gastroschisis can be born safely via a normal vaginal delivery if there are no other factors that warrant a cesarean section.

- Close antenatal surveillance is recommended due to the possibility of late gestational fetal demise.
- Dilated thickened bowel loops may be related to complicated gastroschisis, however this is neither sensitive nor specific.



Ultrasound images of multiple loops of bowel located external to the fetal abdomen that are mildly dilated with diffuse wall thickening.
Courtesy of Jill Stein, MD – Colorado Fetal Care Center – Children's Hospital Colorado

- When counselling families, it is important to introduce the concept of simple versus complicated gastroschisis.
- Complicated gastroschisis may be due to associated intestinal atresia, in utero volvulus, or chronic intesintal pseudo-obstruction.

Delivery and Post-natal Considerations

Neonatal Transition

- Gently wrap the protruding organs in moist sterile dressings and then place the entire body from the nipples downward in a sterile clear plastic bag to protect them.
- Transfer to the neonatal intensive care unit.
- Judicious intravenous fluid resuscitation balancing insensible fluid losses, neonatal fluid status and kidney function and iatrogenic bowel wall edema.
- Full examination by the pediatric surgeon to determine if a primary repair is feasible, or if a silo bag should be placed (depending on the degree of viscero-abdominal disproportion).

Management of Simple Gastroschisis

- When primary repair is possible, the organs will be returned to the abdomen and the defect is closed either surgically or via a sutureless umbilical cord patch.
- Otherwise, the organs will be placed in the silo, and gently brought into the belly over the following few days, until closure is possible.
- Parenteral nutrition via a central venous line until the bowel starts working. This may take several weeks and requires a lot of patience.

Management of Compicated Gastroschisis

- Initial mangement is similar to simple gastroschisis.
- Sometimes the diagnosis of associated intestinal atresia is not apparent at first.
- Various approaches are available for management of these babies including delayed surgical correction after initial closure, immediate definitive surgical correction or temporary decompression via a stoma.

Other Post-natal Considerations

- Some boys with gastroschisis have undescended testicles, which need to be corrected at a later time.