

Gastroschisis in Children

Description/Etiology

Gastroschisis is a congenital full-thickness abdominal wall defect through which a portion of the bowel eviscerates. The defect in the abdominal wall occurs to the right of the umbilicus and the peritoneal membrane that normally covers the bowel is absent. Although the long-term prognosis for live-born infants is good, the condition is associated with significant morbidity and mortality during the perinatal period.

The defect is thought to occur between the fourth and eighth week of fetal development. The pathophysiology of gastroschisis is unclear; potential mechanisms include tearing at the base of the umbilical cord before the umbilical ring closes or failure of one or more folds in the abdominal wall to fuse properly and completely. In most cases, the small and large intestines are herniated and occasionally other abdominal organs are herniated. Prolonged exposure of the eviscerated bowel to amniotic fluid results in intestinal mucosal and muscular injury. Although associated congenital anomalies are relatively uncommon in cases of gastroschisis—occurring in 10–20% of cases—intestinal atresia, ischemic enteritis, and malrotation can occur as a result of the abdominal wall defect itself.

Gastroschisis is suspected based on blood test results that indicate elevated maternal serum alpha-fetoprotein (AFP) levels. Diagnosis can typically be confirmed by routine ultrasound, often before 20 weeks' gestation. (For more information on fetal gastroschisis, including the timing and mode of delivery, see *Quick Lesson About...Fetal Gastroschisis*) The abdominal wall defect is typically repaired surgically within the first few days of life (for details, see *Quick Lesson About...Gastroschisis in the Newborn*).

The long-term prognosis in children who have undergone repair of gastroschisis is generally excellent. Growth delay is common, but neurodevelopment is not delayed. Children with resolved gastroschisis can experience continued GI dysfunction, including diarrhea, constipation, poor gastric emptying, malabsorption, protein allergy, and necrotizing enterocolitis. They can experience complications associated with the abdominal surgical incision scar, intestinal adhesions, and intestinal failure. Gastroschisis is a common cause of short bowel syndrome (SBS; i.e., a disorder characterized by diarrhea, malabsorption of nutrients, steatorrhea, fluid and electrolyte imbalances, and malnutrition secondary to anatomic or functional loss of large segments of the small intestine; for more information, see *Quick Lesson About...Short Bowel Syndrome in Children*) and is the most common indication for intestinal transplantation in children.

Facts and Figures

Gastroschisis is the most common neonatal abdominal wall defect. The incidence of gastroschisis in the United States increased from 2.32:10,000 live births in 1995 to 4.42:10,000 live births in 2005; the increase primarily occurred in mothers under the age of 20 years for whom the risk of delivering an infant with gastroschisis is 11.45:10,000. The abdominal wall defect is equally common in males and females and appears to affect Whites at a higher rate than Blacks and Hispanics. The estimated survival rate for infants with gastroschisis is now 90–95%, compared with 60% in the 1960s. Researchers in a 2010 study reported a 49% 5-year survival rate in children with a history of gastroschisis who underwent intestinal transplant following intestinal failure (Lao et al., 2010).

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Risk Factors

No genetic risk factors have been identified and familial occurrence of gastroschisis is exceedingly rare. Likely risk factors include younger maternal age and smoking and/or use of alcohol, illicit drugs (e.g., ecstasy, methamphetamine), acetaminophen, aspirin, or pseudoephedrine during pregnancy. Reduced maternal intake of α -carotene and total glutathione and increased intake of nitrosamines may be risk factors.

Signs and Symptoms/Clinical Presentation

› For information, see *Description/Etiology*, above

Assessment

› Patient History

- Consult the patient's medical record and question the child's parents regarding the medical history and details of his/her case, including any complications

› Laboratory Tests That May Be Ordered

- CBC may indicate anemia
- Fecal tests may indicate the presence of occult blood and fat malabsorption
- A comprehensive metabolic panel may show malabsorption of specific nutrients (e.g., vitamins) in children with SBS

› Other Diagnostic Tests/Studies

- Abdominal X-rays, lower GI series, or CT scan may be performed to assess for abnormalities, including for intestinal adhesions

Treatment Goals

› Maintain Optimum Physiologic Status and Reduce Risk of Complications

- Monitor vital signs, assess all physiologic systems (especially gastrointestinal system), assess for pain and other discomfort, and review laboratory/other diagnostic test results; immediately report abnormalities and treat, as ordered
- Administer prescribed medications for pain and gastrointestinal dysfunction (e.g., diarrhea, constipation), as appropriate
- Promote adequate nutrition and request referral to a registered dietician for nutritional evaluation
- Follow facility pre- and postsurgical protocols if the patient becomes a candidate for surgery (e.g., intestinal transplantation); reinforce pre- and postsurgical education and verify completion of facility informed consent documents by a parent or legal guardian
 - Postsurgically, maintain adequate pain management and monitor for complications, including infection

› Provide Emotional Support and Educate

- Assess the anxiety level and coping ability of the child (as age-appropriate) and parents; provide emotional support, educate, and encourage discussion about the pathophysiology of gastroschisis, potential complications, treatment risks and benefits, and individualized prognosis
- Follow facility protocols for promoting parental involvement in patient care and rooming-in
- Request referral, if appropriate, to a
 - mental health clinician for patient counseling to reduce stress related to compromised gastrointestinal status and decreased self-image
 - social worker for identification of local resources for support groups

Red Flags

› GI and cosmetic issues can have a significantly detrimental effect on the patient's quality of life

What Do I Need to Tell the Patient/Patient's Family?

- › Reassure the patient/family that the long-term prognosis for children with gastroschisis is excellent
- › Advise parents of children with SBS secondary to gastroschisis of the need for lifelong medical surveillance
- › Encourage the patient to attend a support group for contact with others who face similar health challenges

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