

# GASTROSCHISIS

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## GASTROSCHISIS

## Common Associated Conditions

### Condition and Description

Gastroschisis is a congenital condition where there is incomplete formation of the abdominal wall so that the contents protrude outside the body. There is no membrane (peritoneum) covering the abdominal contents. The opening will be located to the right of the umbilicus, which will be normal. Typically the opening is very small, but it can sometimes be quite large in size. The amount and type of abdominal contents that is protruding varies as well, but it is typically intestinal tissue. The liver is hardly ever protruding through the opening, but sometimes the stomach will be involved. The bowel tissue will float in the amniotic fluid before birth, and this contact with amniotic fluid may damage the bowel tissue, causing it to be abnormal in appearance and function.

Sometimes gastroschisis can be detected very early in pregnancy on a fetal ultrasound or by an elevated maternal or amniotic serum alpha-fetoprotein level. Once detected, the baby may sometimes be delivered early (perhaps at 35 weeks instead of at 40 weeks, which is full-term) to limit the damage to the bowel tissue.

### Prevalence

Gastroschisis occurs in about 2 of 10,000 live births, and 75% of the time it occurs in first-born children. Slightly more boys than girls are born with this condition. It is very rare for this condition to recur in subsequent pregnancies.

Most babies (55%) with gastroschisis are born prematurely, and most (77%) will have intrauterine growth retardation (below-average growth before birth) because the unborn baby will lose nutrients through the exposed bowel. Cardiac problems and shortened bowel length are common problems associated with gastroschisis. Maternal illness and infection, prenatal drug use and smoking, and genetic abnormalities are sometimes associated with gastroschisis. Babies with gastroschisis may occasionally have other intestinal abnormalities such as intestinal atresia (about 10% of the time).

### Short-term Treatment and Outcomes

Babies with gastroschisis are best cared for in a specialized pediatric hospital that has experience in treating children with complex conditions, so transfer to such a facility will be needed if they are born elsewhere. Surgery is performed as soon as the baby is stable after delivery to prevent infection or damage to delicate tissues. A tube will be inserted through the baby's nose or mouth prior to surgery to remove all fluids from the gastrointestinal tract using suction. They will be given antibiotics to combat infection.

If the opening is small (which it is in 80% of the cases), the bowel contents will be carefully placed into the abdomen if the tissue appears healthy and the hole will be surgically closed. However, sometimes this is not possible and a silicon patch will be used to cover the exposed bowel to close the abdomen. Babies will have



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intravenous feedings of hyperalimentation (also called total parenteral nutrition or TPN) solution until the bowel is healed (which takes several weeks). Once the bowel is functioning, oral feedings of breast milk or formula can be safely tolerated.

### Long-term Treatment and Outcomes

Improved surgical techniques and neonatal care have resulted in an 85-90% survival rate of babies with gastroschisis in recent years. However, babies with gastroschisis will require long hospitalizations, and complications are quite frequent, especially those involving intestinal function.

### Common Complications

Because of the exposed bowel, babies born with gastroschisis are prone to hypothermia (low body temperature), dehydration (lack of body fluids), sepsis (infection throughout the body), and hypoglycemia (low sugar level in the blood), all of which cause the baby's condition to be very serious.

After surgery, parents will need to watch the babies very closely for any signs of bowel obstruction, whether from infection, scarring or twisting on itself (called volvulus). Babies with a bowel obstruction will typically vomit bile-colored fluid, be irritable and have a swollen abdomen that is firm to touch (abdominal distention).

A major complication of gastroschisis is short bowel syndrome (also called short gut syndrome) that can occur if a large section of the baby's bowel is damaged prior to birth or becomes infected after surgery. A condition where the bowel becomes infected after birth is called necrotizing enterocolitis (NEC), and it needs to be promptly treated when it is suspected. The bowel is needed to absorb nutrients and if it is not functional and has to be removed, the child's nutrition can be severely

affected. Some children do have to be on life-time TPN because of short-bowel conditions. Intestinal transplantation would be the option for children with short bowel syndrome (though it is not always successful).

A hernia of the abdominal wall can occur in later childhood or adulthood, and that would require further surgery to repair. Children may require periodic assessment from gastrointestinal specialists, nutritionists and pediatric surgeons.

### Implications for Children's Development

The effects on a child's development will vary according to the extent of the condition and the number of complications that occur. Typically the baby is treated in infancy and if there are no short-bowel syndrome complications, the treatment is very successful (85% success rate). The prolonged hospitalizations in infancy can affect the baby's early experiences with the family, and parents must be supported to be at the hospital with the baby doing as much of the care as possible in order to form a strong bond between baby and parent. Illness, immobility, lack of good nutrition, and multiple surgeries may delay the baby's development, but once the initial crises pass, babies can often catch up with their development with the assistance of early infant stimulation programs. They may be prone to infection until the healing has occurred and good nutrition is established.

Surgical scars might be extensive and may cause self-consciousness, especially in adolescence. Children will need to be supported to understand their condition and be self-confident, independent individuals.

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