

## GASTROSCHISIS\*

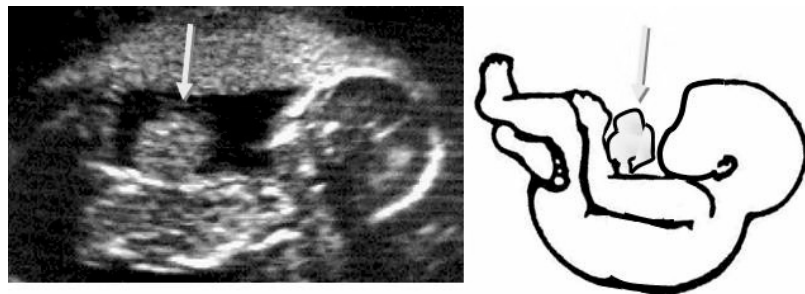
\*This information is intended to supplement your consultation with a pediatric surgeon, regarding your unborn child with gastroschisis. The same information is also available on our web site: [www.fetal-treatment.org](http://www.fetal-treatment.org). Underlined words are explained in a glossary at the end.

*Hasbro Children's Hospital,  
the pediatric division of Rhode Island  
Hospital, a Lifespan partner  
Brown Medical School  
Women & Infants' Hospital*

Maternal-Fetal Medicine  
Pediatric Surgery  
Genetics  
Fetal Cardiology and Echocardiography  
Newborn Medicine  
Pediatric Urology  
Pediatric Neurology and Neurosurgery  
Perinatal Pathology  
Obstetrical Anesthesia  
Perinatal Infectious Diseases  
Prenatal and Special Testing  
Molecular and Cytogenetics Laboratory  
Fetal and Pediatric Imaging  
Pediatric Hematology  
Medical Ethics

### WHAT IS GASTROSCHISIS?

Gastroschisis (sometimes called "laparoschisis") means the presence of a hole in the abdominal wall of the fetus, through which loops of intestines (and sometimes stomach, liver and other organs) protrude. The term only applies to those conditions where the hole is located *to the side* of the umbilicus (umbilical cord); practically speaking, this hole is almost always to the left of the umbilical cord. Gastroschisis is not the same as Omphalocele, which refers to a hole in the abdominal wall *in* the belly button. Although both conditions appear the same (intestines protruding outside the abdomen), each condition has its own features. Abdominal wall defects can be detected by ultrasound from the third month of pregnancy on (14 to 15 weeks). As the pregnancy progresses, diagnosis becomes more accurate: loops of intestine can then be seen outside the abdomen, "floating" into the amniotic cavity (arrow).



Ultrasound image of a fetus with an abdominal wall defect, showing intestines protruding from the hole in the abdominal wall (arrow)

### HOW COMMON IS IT?

Gastroschisis occurs in approximately 1 of every 2,000 live births, making it a relatively "common" congenital anomaly. In fact, its incidence seems to be increasing in recent years, for reasons unknown. There seems to be a relationship with young maternal age, although it can occur at any age. At our institution, we treat 6-10 infants with gastroschisis every year.

*The Fetal Treatment Program  
593 Eddy Street  
Providence, RI 02903*

*(401) 228-0559  
Fax (401) 444-6603*

[www.fetal-treatment.org](http://www.fetal-treatment.org)

## WHAT CAN BE DONE BEFORE BIRTH?

Gastroschisis can be diagnosed with fairly good accuracy from the 14th week of gestation (3 months). It is now possible to intervene during pregnancy for a number of anomalies. It would be tempting, therefore, to try and treat the fetus with gastroschisis before birth. However, extensive research has shown that patients with gastroschisis (and omphalocele) are best treated *after* they are born, and that most in utero interventions would be too risky for mother and child.

We can intervene in other ways, though: with advance knowledge of an abdominal wall defect, it is possible to change the plans for delivery of the baby. One can change the mode, place and time of delivery.

### 1. Mode of delivery

If intestines and other organs are outside the abdomen, it would seem logical that they would be at an increased risk of being damaged during normal delivery. Some have therefore advocated Cesarean section ("C-section") for all cases of gastroschisis and omphalocele. In fact, the risk of injury is only theoretical, and vaginal delivery does not put the baby at an increased risk of complications. For that reason, most (although not all) physicians now recommend normal delivery, even for gastroschisis, unless there are obstetrical reasons to proceed with a C-section.

### 2. Place of delivery

As long as he or she is inside the womb, the fetus with a gastroschisis is relatively well shielded from trauma and complications. After birth, however, the exposed intestines have to be protected from direct trauma, dehydration and infection. The baby can be safely transported to a treatment center, as long as certain precautions are taken. However, if the diagnosis of gastroschisis has been made beforehand, it would seem logical to have the baby be born directly in such a treatment center (i.e., a center with a neonatal intensive care unit and immediate access to a pediatric surgery service). Therefore, we generally recommend that, if you are pregnant with a fetus with gastroschisis or omphalocele, you plan to deliver in such a tertiary institution. Your care will likely be transferred to a Maternal-Fetal Medicine specialist at our institution, to facilitate the transition to peri- and postnatal care.

### 3. Time of delivery

One of the concerns with gastroschisis is that the exposed bowel becomes so damaged, that function is impaired and the baby may end up staying in the intensive care for a long time. It is known that many infants with gastroschisis have what appears to be damaged bowel, with very thick, rigid loops of intestines containing a "peel." One of the theories for this peel (and for the fact that some babies have little or no peel at all) is that prolonged exposure of the bowel to the amniotic fluid causes progressive damage. In other words, limiting the amount of time that the bowel is floating in this fluid (or even diluting that fluid by infusing sterile saline water inside the womb) could theoretically decrease the amount of peel and intestinal damage.

Many centers have therefore recommended early delivery (between 35 and 37 weeks of gestation, instead of the normal 40 weeks). Unfortunately, there are no good scientific studies proving the benefits of this. In fact, at our institution, we have reviewed all babies born with gastroschisis in the last 10 years, and have found no benefit at all of early delivery. For that reason, we recommend that your baby be born as close to term as possible.

## WHAT WILL HAPPEN AT BIRTH?

If everything goes as planned, you will deliver at a tertiary care center with direct access to a neonatal intensive care unit. The neonatologists will be present at delivery, so that they can immediately assess your baby and start treatment, if necessary. At the same time, the pediatric surgeons will be alerted, so that surgical correction can be performed as soon as possible. In most cases, however, you will be able to see (and hold) your baby after delivery.

Your baby will be "stabilized" in the intensive care unit. An intravenous line will be placed in an arm or a leg, so that fluids can be given. Because of the exposed intestines, your baby is likely to lose a lot of fluid by evaporation, and is likely to cool off more rapidly as well. Your baby will therefore be placed under a warmer, and the loops of bowel will be carefully wrapped to protect them from the outside. If your baby shows signs of distress, it is possible that he will be intubated, so that we can help him breathe better.

Once it is clear that there are no other major problems, your baby will be ready to undergo surgical repair of the defect. How this is done will depend on how much intestines and other organs are exposed, and how big your baby is. In many cases, all the intestines can safely be placed back in the abdomen (so-called "primary repair"), and the abdominal wall can be closed. Of course, this is done in the operating room with your baby under anesthesia. Often, however, there is so much out that this cannot be safely replaced all at once. In that case, we try at least to protect the intestines until they are ready to be put back in the abdomen. For this, we place a "silo" (a clear plastic or silicone pouch) over the intestines, so that they are now shielded from trauma, infection and dehydration. This can be done at the bedside, in the intensive care unit, or in the operating room.

Once the swelling has gone down and the abdomen has become used to the presence of more bowel, the silo can be removed and the abdomen closed over the intestines. This typically takes a few days to a week.

### WHAT HAPPENS NEXT?

As mentioned before, the intestines have suffered somewhat during pregnancy, and they will need some time to recover. On average, it may take 2 to 3 weeks before the intestinal tract functions properly again. During that time, your baby will be fed through the veins only, by "total parenteral nutrition," or TPN. He will get all the calories necessary to grow, until he can be fed by mouth again. Once gut function returns, it will likely take a while before your baby can tolerate full feeds, and that nutrition through the veins can be stopped. Your baby is likely to stay in the hospital for at least 1 month. Sometimes, this can be much longer, depending on the degree of prematurity and the condition of the bowel.

### COMPLICATIONS AND LONG-TERM OUTCOME

The overall outcome of gastroschisis is excellent: some infants may have minor intestinal problems in the first few months, but will recover from that and lead a completely normal life. Although the belly button may not look perfectly normal, there should be minimal scarring.



Typical scar after repair of a gastroschisis

In some rare cases, however, there may be some complications. While gastroschisis is usually *not* associated with other anomalies, there may be intestinal defects in 5 to 10%. These represent in utero "accidents," where a piece of intestine becomes necrotic and disappears. As a result, there may be a missing portion of intestine (intestinal atresia), which will have to be fixed. Often, this is not discovered until a few weeks after birth. An additional operation will then be necessary. Very rarely, a large portion of intestine suffers and dies off. In those rare instances, bowel function may suffer.

## GLOSSARY

**Amniotic cavity:** The space within the uterus in which the fetus resides, and bound by the amniotic membrane

**Atresia:** An absent portion of an organ. Intestinal atresia (ileal or jejunal atresia) refers to a missing portion of small bowel, a known complication of gastroschisis. If present, this will require an operation, to reconnect the two ends of bowel

**Beckwith-Wiedemann syndrome:** An anomaly that includes omphalocele, enlargement of some of the organs (often the pancreas), a large tongue and various degrees of gigantism (large baby). These children have a risk of developing some childhood tumors, such as Wilms tumor and hepatoblastoma.

**Cloacal exstrophy:** A complex anomaly of the abdominal wall, the intestinal tract and the pelvic organs. Infants with cloacal exstrophy have a short intestine, an absent anus, a large defect of the abdominal wall (omphalocele) and the bladder (which is exposed and lacks a front wall), and anomalies of the pelvic bone and genitalia. This is not the same as a bladder exstrophy (defect of the bladder, with or without anomaly of the penis in boys), which is a less severe condition.

**Diaphragm:** A large muscular sheath that separates the chest from the abdomen. Anomalies of the diaphragm include diaphragmatic hernia, a congenital defect in that muscle. The most common form is a posterolateral hernia, or Bochdalek type (whereby the hole is mostly on the side and in the back of the diaphragm, causing intestines to move into the chest). The anterior form, or Morgagni, is a defect behind the sternum (breast bone); this is the one most commonly associated with omphaloceles, as part of the Pentalogy of Cantrell.

**Gastroschisis:** refers to a defect (hole) in the abdominal wall of the fetus or newborn, through which intestines or other abdominal organs can protrude. In gastroschisis, the hole is to the (patient's) left of the belly button.

**Omphalocele:** Abdominal wall defect in the fetus, located in the umbilicus. Often, a membrane covers the exteriorized intestines. "Giant" omphaloceles contain not only intestines, but liver as well. If only a very small defect is present, this is often referred to as "hernia of the umbilical cord." Omphaloceles are to be differentiated from gastroschisis, where the defect is to the side (usually left) of the belly button. Omphaloceles are often associated with other anomalies, including congenital heart defects. Omphalocele can also be part of a syndrome, such as Beckwith-Wiedemann syndrome, pentalogy of Cantrell and cloacal exstrophy.

**Peel:** The thick layer of scar often seen in exposed bowel loops in gastroschisis.

**Pentalogy of Cantrell:** A rare syndrome that includes five anomalies: an omphalocele, a diaphragmatic hernia (hole in the diaphragm), a heart defect, a defect of the pericardium (the membrane that envelops the heart), and a defect the sternum (breast bone), which in very severe cases can cause the heart to protrude out of the chest.

**Silo:** A silicone plastic ("silastic") sterile bag or membrane that can be placed over the exposed bowel loops in gastroschisis, to prevent dehydration and further damage to them. It is used for temporary cover if the intestines can not immediately be replaced in the amniotic cavity.

**Total Parenteral Nutrition (TPN):** Basic nutrients (carbohydrates, essential fats, proteins, vitamins and trace elements) given intravenously (through a vein), usually via a central venous catheter, that provide enough calories for an patient to survive *and* grow without any feeding by mouth. This life-saving technique allows a baby with gastroschisis to receive enough nutrition until he can feed normally (often 3-4 weeks later).

**Trisomy:** The presence of an extra chromosome (normally, humans have 2 sets of 23 chromosomes, or 46 chromosomes, in each cell; individuals with a trisomy have an extra copy of one chromosome per cell). The most common trisomy syndromes are trisomy 21 (Down syndrome), trisomy 13 and trisomy 18. The latter two are sometimes associated with an omphalocele.