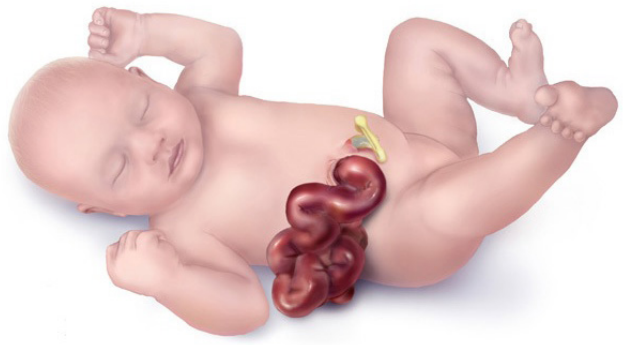
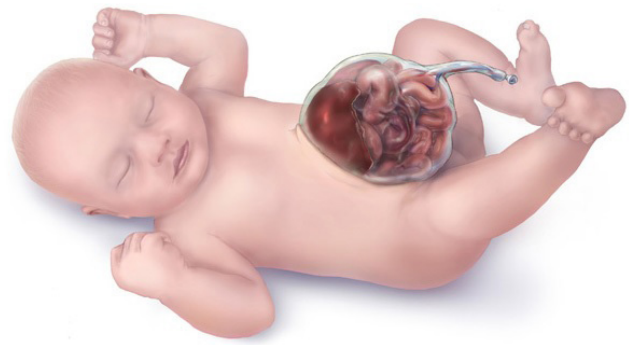
Gastroschisis and Omphalocele

The two most common congenital abdominal wall defects are gastroschisis and omphalocele. Both involve incomplete closure of the abdominal wall during fetal development, and for both, their cause is unknown. A gastroschisis is usually an isolated congenital defect, whereas a baby with an omphalocele often has chromosome anomalies, cardiac conditions, and other major birth defects.

A *gastroschisis* is a herniation of abdominal contents through a defect in the abdominal wall, usually just to the right of the umbilicus. An *omphalocele* is a herniation of abdominal contents into the umbilical cord itself. The contents of a gastroschisis are directly exposed to amniotic fluid, whereas the contents of an omphalocele are usually covered with a protective membranous sac.



Gastroschisis. Courtesy of the Centers for Disease Control and Prevention.

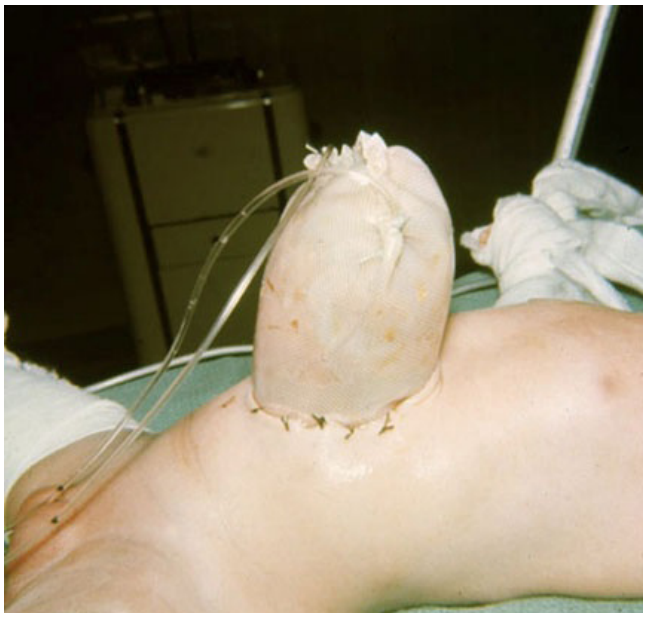


Omphalocele. Courtesy of the Centers for Disease Control and Prevention.

At delivery, the ABC (airway, breathing, circulation) rule should be followed for babies with gastroschisis or omphalocele. Immediately afterward, protection of the herniated contents and management of evaporative loss should be accomplished. Abdominal contents should be wrapped in warm, saline-soaked gauze and covered with plastic wrap. Alternatively, the baby should be placed in a sterile bowel bag up to the nipple line. Preventing evaporative fluid loss is particularly important for the baby with gastroschisis because of the lack of the protective membranous covering of the abdominal contents. Diligent observation of the color and perfusion of the abdominal contents of a baby with gastroschisis is imperative. The baby should be placed on his or her right side with abdominal contents supported with additional gauze or blankets to prevent kinking of the mesentery blood vessels. An echocardiogram also should be considered to rule out potential cardiac anomalies (Escobar & Caty, 2016).

Babies will need IV fluids started; the baby with a gastroschisis will need a higher than normal IV fluid rate to prevent dehydration. Both will need placement of a Replogle tube to low intermittent wall suction (30–40 mmHg) to prevent gastrointestinal distention. Both will need to have broad-spectrum antibiotics started.

These babies will require surgical intervention after birth, but the timing of surgery in each case may differ. If the defect is small, surgery may be done shortly after birth with a primary closure. Closure for a gastroschisis must be done on a more urgent basis than that of an omphalocele to prevent continued damage to the exposed abdominal contents. If the defect is large, the size of the abdominal cavity may not be spacious enough to safely replace all of the herniated contents. If the contents are replaced under pressure, this could compromise respiratory function and vascular perfusion and result in the loss of bowel tissue. Therefore, with a large gastroschisis, a silo (sterile bag suspended above the abdomen) will be placed surrounding the abdominal contents so they can



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be gradually reduced into the abdomen over a period of days to weeks. The contents of an omphalocele also may be reduced in this manner.

Multiple alternative staged methods of closing omphaloceles also have been described. Some use the baby's own tissue; others use grafts from other sources. A nonsurgical way of initially closing a particularly large omphalocele, a so-called giant omphalocele in which multiple organs have herniated, has been to harden the membranous sac covering the contents with various topical chemicals. This method provides long-term protection, allowing skin to eventually grow over the sac. Once skin has covered the sac, closure is then surgically performed. Another recent method for closing giant omphaloceles is negative pressure wound therapy, which has a low complication rate (Aldridge et al., 2016).

Outcome for the baby with a gastroschisis depends on time of closure (Gonzalez, Cooper, St Peter, Minneci, & Deans, 2017), how much viable intestine is available, whether or not there were associated atresias, and how long it takes for gastrointestinal function to take place. Outcome for the baby with an omphalocele largely depends on the extent of complications with the other associated congenital defects. For babies with either defect, it can be a long road to achieve full feeds; gastroschisis babies usually take longer to accomplish this. It is common for babies with either defect to have many episodes of feeding intolerance. These babies also are subject to short- and long-term dependence on parenteral nutrition with all their associated potential problems of infection, poor growth, and liver failure. Parents of these babies need substantial encouragement and support.

It may take a long time for babies with gastroschisis and omphaloceles to be able to eat normally and have normal intestinal function, especially those with gastroschisis. However, using human milk for feedings has been shown to reduce length of hospital stay for infants with gastroschisis (Gulack et al., 2016). In many cases, it is "two steps forward and one step back" for feedings. There also can be complications with the IV and special IV fluids these babies need to grow until they are able to digest their food well. However, almost all of these babies are eventually able to eat on their own.

Bibliography

Aldridge, B., Ladd, A. P., Kepple, J., Wingle, T., Ring, C., & Kokoska, E. R. (2016). Negative pressure wound therapy for initial management of giant omphalocele. *The American Journal of Surgery, 211*(3), 605–609.

Christison-Lagay, E. R., Kelleher, C. M., & Langer, J. C. (2011). Neonatal abdominal wall defects. *Seminars in Fetal & Neonatal Medicine, 16, 164–172.*

Escobar Jr, M. A., & Caty, M. G. (2016). Complications in neonatal surgery. *Seminars in Pediatric Surgery, 25*(6), 347–370.

Gonzalez, D. O., Cooper, J. N., St Peter, S. D., Minneci, P. C., & Deans, K. J. (2017). Variability in outcomes after gastroschisis closure across U.S. children's hospitals. *Journal of Pediatric Surgery,* pii: S0022-3468(17)30253-1.

Gulack, B. C, Laughon, M. M., Clark, R. H., Burgess, T., Robinson, S., Zhang, A., . . . Smith, P. B. (2016). Enteral feeding with human milk decreases time to discharge in infants following gastroschisis repair. *The Journal of Pediatrics, 170*, 85–89.

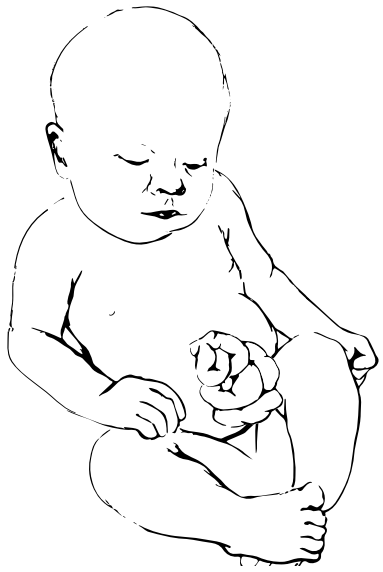
Islam, S. (2012). Advances in surgery for abdominal wall defects, gastroschisis and omphalocele. *Clinics in Perinatology, 39*, 375–386.

Ledbetter, D. J. (2012). Congenital abdominal wall defects and reconstruction in pediatric surgery, gastroschisis and omphalocele. *Surgical Clinics in North America, 92*, 713–727.

Gastroschisis and Omphalocele: Information for Parents

Gastroschisis and omphalocele are the two most common abdominal wall birth defects. Their causes are unknown, and they are not due to anything a mother did or didn't do during pregnancy.

Babies with gastroschisis and omphaloceles are the same in some ways and different in other ways. Both are types of hernias where the parts of the body that should be inside the abdomen are on the outside instead. With a *gastroschisis*, the hernia is through a hole in the muscle wall of the abdomen. With an *omphalocele*, the hernia is through the area of the belly button. Babies with omphaloceles also are more likely to have other birth defects



Infant with gastroschisis

When babies are born with these hernias, they will have several extra things done in the delivery room. The parts that are on the outside will be carefully protected. The abdomen may be covered with warm, wet sterile gauze and plastic wrap, or the baby's whole body may be put in a big clear plastic bag up to the chest. This is done to keep the babies from losing fluid from the parts that shouldn't be exposed to the air. These babies also will have a tube hooked up to a suction machine and put through their mouth or nose to their stomachs. This will keep air from getting in and swelling their intestines. An intravenous (IV) line and IV fluids will be started. Antibiotics will be started, too.

Surgery will be needed to put the herniated parts back into the abdomen. If there is enough space in the abdomen, it may be possible to do this in one step. But sometimes the space is not big enough; in that case, it may take a few days to weeks to put everything back in comfortably. Your baby also may have a sonogram of their heart (to rule out any problems) and their abdomen (to make sure the organs on the inside are normal).

It can take a long time for babies with gastroschisis or omphaloceles to be able to be held and eat normally and have normal intestinal function, especially babies with gastroschisis. Once the baby recovers from surgery and the abdomen is closed, they can be held. Starting feedings is a slow process. Your baby may have an IV for a long time until he or she can digest food well. However, almost all of these babies are eventually able to eat on their own.