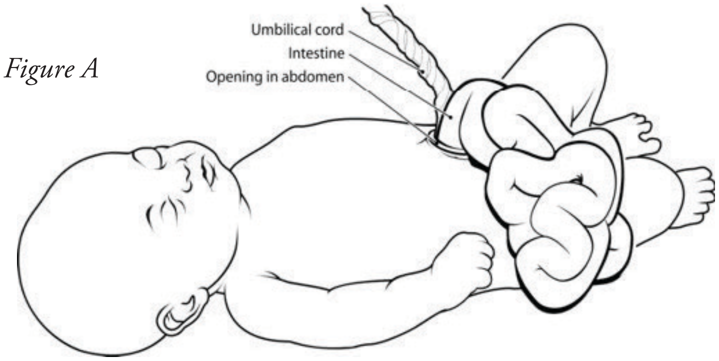
Gastroschisis

Figure A



Overview

Normally, during prenatal developmental, the muscles and skin that form the abdominal wall completely close by the 12th week of pregnancy. When the muscles and skin fail to form normally, this results in an abdominal wall defect. Gastroschisis is the most common of the abdominal wall defects. Babies born with gastroschisis have a hole in the abdominal wall. The intestines, and sometimes other organs, come through a small opening to the right of the umbilical cord, and float freely in the amniotic fluid. (See Figure A). Because the intestines have been exposed to the amniotic fluid, they may be swollen and covered with a thick film, which can make the intestines function poorly. About 10% of babies with gastroschisis will have another problem within the bowel, such as an interruption in the intestine (atresia, absence of a portion of the intestine), and, less commonly, a perforation, or rupture causing a hole in the intestine. Problems with organs other than the intestines are very uncommon in babies with gastroschisis. The survival rate for babies born with gastroschisis is 95%.

Cause

The exact cause of gastroschisis is not known. Gastroschisis affects both males and females in equal numbers. Most babies with gastroschisis are born to young mothers in their first pregnancy. It is congenital, meaning it happens before your baby is born. Usually, the condition is not inherited, and future pregnancies are not affected. Usually there is not a family history of gastroschisis.

How Gastroschisis is Diagnosed

Gastroschisis may be found during routine prenatal ultrasound. If your doctor suspects a problem, additional tests may be ordered. These may include:

- **Fetal echocardiogram**: A fetal echocardiogram is a special ultrasound used by a pediatric heart doctor (cardiologist) to look closely at your baby's heart and surrounding blood vessels.
- **Fetal MRI** (magnetic resonance imaging): This type of imaging gives more detailed pictures of your baby's organs.

These tests are not typically needed unless your doctor suspects there may be other problems with your baby.

Other Tests

Some congenital conditions are a result of changes in DNA, or a baby's genetic make-up. The doctors may suggest genetic testing to find out more about your baby's DNA.

- **Cell-Free Fetal DNA testing**: Your doctor can take a sample of your blood to look for copies of fetal (baby) DNA. This is only a screening test. A positive result means that there could be problems with the DNA. This should be confirmed with another test to be sure of the result. This additional testing can be done before or after birth depending on your needs and the needs of the baby.
- **Amniocentesis**: A small sample of the amniotic fluid that surrounds the baby in the womb is taken and tested for DNA problems. This test is usually done in the middle of the second trimester. An amniocentesis can be used to confirm findings on the cell-free fetal DNA test.
- **Testing the baby**: After birth, a test is done with a sample of your baby's blood to look for chromosome problems. This is called either a "karyotype" (basic chromosome study), or microarray (a more detailed look at smaller pieces of DNA).

Genetic changes are very uncommon in babies with gastroschisis.

Care During Your Pregnancy

If a test finds you are carrying a baby with gastroschisis, you will continue to receive care from your obstetrician. Your obstetrician may refer you to the Ohio Fetal Medicine Collaborative (OFMC), which will help you connect with a team of fetal medicine experts.

There, you will meet with a multidisciplinary team including neonatologists (doctors who specialize in treating newborns), pediatric surgeons (doctors who perform surgery), and nurses to learn more about your baby's care after he or she is born. Nurse coordinators will help answer your questions and concerns, guide you through your pregnancy, and prepare you for what to expect.

Your doctor will watch your pregnancy closely. There is a risk of preterm delivery when a baby has gastroschisis. You should deliver your baby at a hospital that is ready to care for high-risk babies like yours. You should discuss your plans for labor and delivery with your obstetrician. Contact your doctor right away if you have any concerns.

Care After Your Baby is Born

At the delivery hospital, a neonatologist (a doctor who specializes in treating newborns) will manage the care of your baby. The neonatologist will assess your baby's breathing and heart rate, and examine the baby for other problems.

- Your baby will briefly receive care in your delivery hospital's neonatal intensive care unit (NICU).
- As soon as your baby is born, a sterile plastic bag called a bowel bag will be placed over the baby's feet and pulled up over the bowel to the chest, to help protect the intestines. This bag helps to prevent infection by keeping germs from getting to the intestines, and helps keep your baby warm and hydrated. This bag stays on until he or she is evaluated by the surgeons at the hospital.
- A tube will be put through your baby's nose (nasogastric, NG) or mouth (orogastric, OG) down into the stomach. This tube helps to keep the stomach and intestines empty and decompressed.
- An intravenous line (IV) will be started in your baby's arm, hand, foot or scalp. The IV provides fluids and antibiotics to prevent dehydration and infection.
- Your baby may need extra oxygen to help ensure there is good blood flow to the intestines.
- Your baby will be immediately transferred to the Children's Hospital neonatal intensive care unit (NICU) for further care.

Treatment

- Your baby will not be able to drink breast milk or formula until after the intestines have healed from surgery. A special IV called a PICC (Peripherally inserted central catheter) will be used to give your baby IV nutrition called TPN (total parenteral nutrition). TPN has all of the calories and nourishment needed for your baby to grow.
- We strongly encourage you to pump breastmilk to provide to your baby when he or she is ready to start feeds. Breastmilk is best for all babies, and is particularly important for babies with gastroschisis, as breastmilk is very gentle on the intestines.
- The NG or OG will stay in place through the time of surgery, and after surgery until the intestines recover and start to function well.

Surgery

Once your baby has been admitted to the NICU, the pediatric surgery team will evaluate the intestines and decide on which surgical approach would be best. See Figure B.

Figure B

Primary Repair

Less than 10% of the time, the surgeon is able to place all the intestines back into the abdomen and close the opening on the first day of life.

Staged Repair

A staged procedure is done when there is a lot of intestine on the outside, or if the bowel is too swollen and inflamed right after birth. Placing the swollen intestines into the abdomen puts too much pressure on the lungs, making it hard for your baby to breathe, and puts too much pressure on the bowel, putting them at risk for poor blood flow. In this situation it is safer to do the procedure in steps or stages.

For the first stage, a plastic surgical pouch called a silo is placed around the intestines. See Figure C. This helps protect the intestines from infection and dehydration. Every day the surgeon will gently push part of the intestines into the abdomen until everything is inside. It can take up to a week or more to get all the intestines back into the belly. Then your baby will go back to surgery to close the hole in the abdomen.

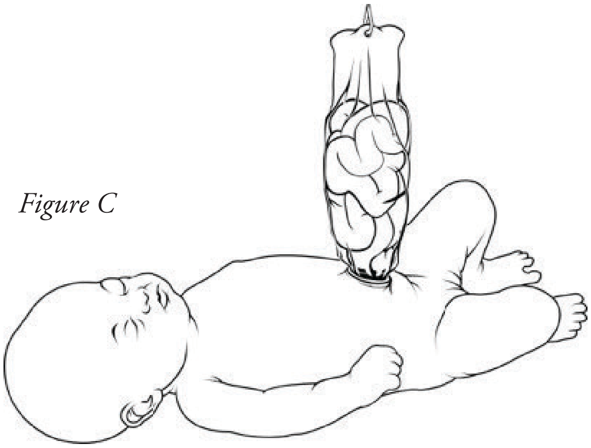


Figure C

In both cases, the surgeons will try to make sure your baby will have a belly button, but it may be slightly to the left.

If your baby has an atresia or other complication, additional surgeries may be needed.

Babies cannot breastfeed or take bottles until they are off the ventilator or CPAP and only requiring low amounts of oxygen flow by nasal cannula. Sometimes it can take babies some time to learn how to breastfeed or bottle feed. Your baby can continue to receive feeds though the NG tube while learning this skill.

Care After Surgery

After surgery, your baby will receive care in the hospital's NICU. Your baby will need to be on a breathing machine until he or she is fully recovered from surgery. Other treatments your baby will probably need after surgery are:

- Antibiotics to prevent infection
- Fluids and nutrients given through the PICC
- Oxygen
- Pain medicines as needed
- The NG or OG tube will stay in place until your baby's intestines begin to work properly. This may take several weeks. Your baby will not be able to eat until the intestines have healed.

Feeding

Feedings are started once the intestines are fully healed after surgery. It may take several weeks for the intestines to heal and start to work. Your baby will be started off with small amounts of breast milk or formula, and the amount will be increased gradually over time. The feeds can be given by bottle, if your baby is ready and interested in a bottle, but sometimes needs to be given via an NG feeding tube. When your baby is taking enough breast milk or formula, by bottle or tube, to grow and gain weight, the central line will be removed. Once your baby is tolerating full volumes of milk, we will encourage you to start working on direct breastfeeding. A lactation consultant is available to assist you.

Complications:

- Infection: Your baby will be on antibiotics right after birth and for several days after surgery. The surgeon and staff will monitor your baby very closely for any signs of infection.
- Sometimes babies need a blood transfusion.
- Feeding problems: It takes the bowel several weeks to heal properly. Your baby will not be able to eat large amounts of formula or breast milk right away. This is one of the major reasons why your baby has to stay in the hospital for a long period of time.
- Twisting of the bowel during development may cause bowel atresia (absence of a portion of the intestine) in 10% of cases. The bowel may have perforated (ruptured) in 5% of cases. If either of these complications occur, your baby will need additional surgeries.

Discharge Planning

You will be able to take your baby home once he or she is taking enough breast milk or formula to grow and gain weight. The average length of hospital stay is about 4-6 weeks. A few babies will do especially well and go home earlier. Some babies, particularly those with any complications, may need to stay longer until their intestines are functioning properly.

Looking Ahead

You will want to find a local pediatrician to take care of all the routine checkups, immunizations and doctor's visits. The program coordinator can help if needed. The pediatric surgeon will follow up with your baby's surgical needs for as long as necessary.

Most babies with gastroschisis will go on to feed and develop normally throughout childhood.

About the Ohio Fetal Medicine Collaborative

The Ohio Fetal Medicine Collaborative (OFMC) is a group of care providers and hospitals committed to achieving optimal outcomes for high-risk expectant mothers and their babies with suspected fetal diagnoses.

Members of the OFMC include:

- Dayton Children's Hospital
- Miami Valley Hospital
- Mount Carmel Health Systems
- Nationwide Children's Hospital
- OhioHealth
- The Ohio State University Wexner Medical Center

For more information about OFMC or to learn more about gastroschisis, visit OhioFetalMedicine.org