

Let's Talk About...

Prenatal Counseling: Omphalocele

What is an omphalocele?

An **omphalocele** [ohm-FAL-oh-seel] occurs when a baby's abdominal (belly) organs push out of the body and into the umbilical cord during early fetal development. The omphalocele is almost always covered by a membrane (thin covering), which keeps the organs from floating freely in the amniotic fluid (liquid protecting the baby in the womb).

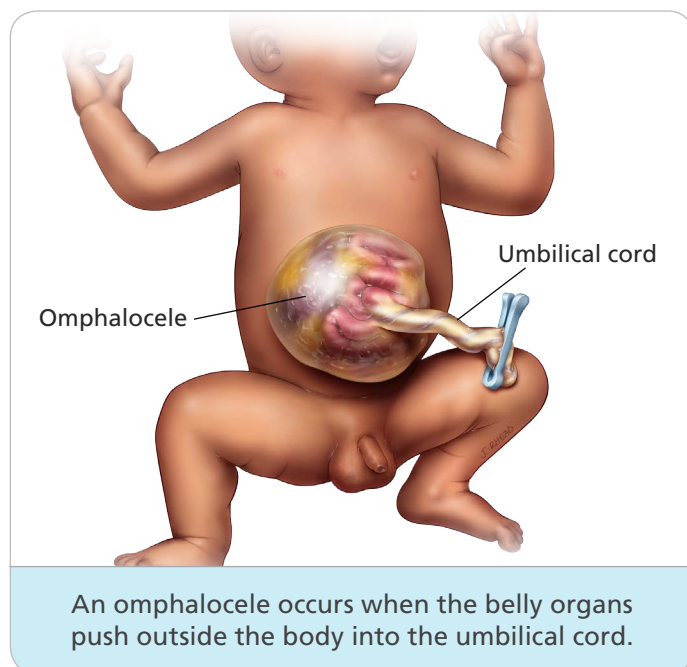
Omphaloceles can vary in size from small to large. A "giant" omphalocele contains most of the organs from the belly, including the liver. This may tell the doctor that there is a space between the belly muscles larger than 5 centimeters (about two inches). About 1 in 5,000 babies are born with an omphalocele each year, but only 1 in 10,000 babies have a giant omphalocele. We do not know what causes an omphalocele, but nothing the mother does or has done during pregnancy causes this problem.

The **Utah Fetal Center team** will help you make the best possible decisions about your baby's omphalocele. The team includes maternal-fetal medicine specialists for the pregnant mother, neonatologists who are specially trained to care for newborns, and pediatric specialists to help with your baby's needs before and after delivery.

How is an omphalocele diagnosed during pregnancy?

The fetal team usually finds an omphalocele during a pregnancy ultrasound around 20 weeks, but they can see it as early as 11 to 14 weeks. If the fetal team sees the defect, they may recommend follow-up tests, including:

- **A comprehensive fetal ultrasound.** This test provides more detail about your baby's other body structures that may be affected.



An omphalocele occurs when the belly organs push outside the body into the umbilical cord.

- **A fetal echocardiogram** [ek-oh-CAR-dee-oh-gram] or **ECG**. This ultrasound, performed by a pediatric cardiologist, looks at the structure and function of your baby's heart. Babies with omphaloceles have an increased risk of heart problems. In fact, the smaller the omphalocele, the higher the risk of a congenital heart defect. Even a baby with a small omphalocele needs an echocardiogram.
- **Formal genetic testing.** Three to 6 out of 10 babies with omphaloceles have another genetic problem, often congenital heart disease. A genetic counselor will tell you more about screening tests.
- **Fetal MRI.** Your baby may need an imaging test so the doctor can see how severe the omphalocele is and look at other structural problems. This is also important if the doctor thinks your baby has a giant omphalocele.

These tests give you and the fetal team the most accurate information so you can make decisions about treatment.

How does the omphalocele affect my baby?

The effect of an omphalocele on your baby's overall health can range from mild to significant depending on its size. Your baby's outcome often depends on other problems, such as genetic disorders or congenital heart disease. Many babies with omphaloceles have:

- **Pulmonary** [PUL-moh-nehr-ee] **hypoplasia** [HI-po-PLAY-zee-uh]. Small, underdeveloped lungs
- **Pulmonary** [pul-moh-nehr-ee] **hypertension** [HI-per-TEN-shun]. Increased blood pressure in the blood vessels of the lungs

When your baby has small lungs and increased pulmonary blood pressure, it's hard for them to get enough oxygen while breathing. This puts a strain on your baby's heart.

These findings can only be diagnosed after your baby is born.

How is an omphalocele managed during pregnancy?

The fetal center will review your tests and tell you more about omphaloceles, recommend ways to manage your pregnancy, and help you meet the specialists who'll care for your baby after delivery.

There is no fetal treatment for omphaloceles, which means your baby will need surgical treatment after they're born. You may deliver your baby early, so your doctor should closely monitor your pregnancy. If your baby has a giant omphalocele, you may need a cesarean [sess-SAY-ree-an] delivery (C-section) to protect their liver from injury during birth.

Where should I deliver my baby?

You should deliver your baby at a hospital with a neonatal intensive care unit (NICU) that specializes in treating omphaloceles so a pediatric surgeon can immediately evaluate your baby after birth.

If your baby has a giant omphalocele or has an omphalocele and a severe congenital heart defect, you should deliver at the University of Utah Hospital. Your baby will then receive care at Primary Children's Hospital.

How is the omphalocele managed after delivery?

A specially trained pediatrician called a neonatologist [NEE-oh-nay-TAHL-oh-jist] will manage your baby's medicine, feeding, and daily needs in the NICU. They will evaluate your baby right after their birth and make sure their heart and lungs are working right.

Your baby may need oxygen to help them breathe or may even need a ventilator (breathing machine). They may also need:

- **An NG or OG tube.** A healthcare provider inserts a nasogastric [NAY-zo-gas-trick], or NG, tube in your baby's nose or an orogastric [or-oh-GAS-trick], or OG, tube in your baby's mouth. The tube goes all the way to the stomach and sucks out any fluid. This prevents your baby from choking or breathing stomach contents into the lungs.
- **A bowel bag.** This plastic bag covers your baby's body below the armpits. It protects the omphalocele and keeps it warm and moist until the surgeon arrives. The surgeon then evaluates the omphalocele and places a special dressing around it to protect it and keep it clean.
- **A PICC line.** A peripherally [per-IF-er-uh-lee] inserted central catheter (PICC line) is a tiny tube inserted through the umbilical cord or an arm or leg. Your baby will receive fluids, antibiotics, other medicines, and nutrition through the PICC line.
- **TPN.** Your baby won't be allowed to eat at first, so they'll receive total parenteral [pah-REN-ter-ull] nutrition, or TPN, through the PICC line. TPN contains protein, fat, sugar, vitamins, and minerals and will meet all your baby's nutritional needs.

Your baby may need another ECG after their birth depending on their fetal ECG results. The fetal center will consult a pediatric cardiologist to help care for your baby if needed.

Because the omphalocele is closed by a protective covering, there is no urgency to repair it. The fetal team can take time to stabilize your baby in the NICU and do more tests.

How is an omphalocele treated?

There are usually 3 treatment options, depending on the omphalocele's size and your baby's health:

- **Primary closure.** If your baby is stable and the omphalocele is small enough, a surgeon can close it with one surgery. They will remove the omphalocele sac and put the organs back into the belly. They will then close the muscle with stitches and create a belly button.
- **Staged closure (Schuster [shoo-ster]).** If your child has a giant omphalocele but is stable, the surgeon can slowly push the omphalocele in and close it over several days to weeks. Your baby usually needs multiple surgeries.
- **Sclerotherapy ("paint and wait").** If your baby is stable, sclerotherapy [sk-LER-oh-THEY-ruh-pee] is an option. The doctor treats the omphalocele with medicine, usually betadine [bay-tuh-dyne] or silver sulfadiazine [sul-fuh-DY-uh-zeen], to keep it clean and allow new skin to grow over the omphalocele. This usually takes several months, but you can do treatment at home. When your baby is big enough, the surgeon closes the omphalocele. Sclerotherapy is often the only option for children who are unstable, have severe congenital heart disease, or who need a lot of breathing support.

You'll discuss these options with the pediatric surgeon during your fetal center evaluation and again after your baby is born. They will help you decide which approach is best for your baby.

What outcome can I expect for my baby after surgery?

Your baby's outcome can vary depending on the omphalocele's size, other birth defects, and how well your baby's lungs work. Overall, 9 in 10 babies born with omphaloceles survive.

Your baby may stay in the NICU for several weeks to months, depending on their condition. They may need heart surgery to fix a congenital heart defect.

Rarely, your baby may need a tracheostomy [TRAY-kee-AH-stoh-mee], or tube in their windpipe, for long-term breathing help with a ventilator if they have severe lung disease.

After you go home, your baby will need close follow-up with a pediatrician to monitor their growth and development. You may also need to follow up with other heart, lung, stomach, esophagus, and brain specialists.

Some babies also need home health nursing, occupational therapy, physical therapy, speech therapy, and nutritional therapy. Many babies will need to see a pediatric surgeon often to care for the omphalocele or make sure it is repaired well.

For additional information

Utah Fetal Center

[intermountainhealthcare.org/locations/primary-childrens-hospital/medical-services/utah-fetal-center](https://www.intermountainhealthcare.org/locations/primary-childrens-hospital/medical-services/utah-fetal-center)

American Pediatric Surgical Association (APSA)

[eapsa.org/parents/conditions/f-o/omphalocele](https://www.eapsa.org/parents/conditions/f-o/omphalocele)

Centers for Disease Control and Prevention (CDC)

[cdc.gov/ncbddd/birthdefects/diaphragmaticernia.html](https://www.cdc.gov/ncbddd/birthdefects/diaphragmaticernia.html)

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MOO: Mothers of Omphaloceles

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