

Omphalocele

Sometimes during pregnancy, the baby's abdominal wall does not close at the umbilicus. The intestine remains outside the abdomen in a sac. This is called an omphalocele, which is an abdominal wall defect. An omphalocele can be small, containing parts of the intestine, or larger, containing parts of the liver and other organs. (See image)



Cause

The cause of an omphalocele is not known. It is congenital, meaning it happens before your baby is born. Omphalocele develops in 1 in about every 5,000 live births. Approximately 25 to 40 percent of infants with an omphalocele have other birth defects such as genetic problems and heart defects. Many babies with omphalocele have an underlying genetic diagnosis

which may involve other birth defects and/or mental retardation. You may discuss prenatal diagnosis with the obstetric team.

How an omphalocele is found

You may have a prenatal ultrasound done while you are pregnant to learn about the health of your baby. An omphalocele is usually found during a routine prenatal ultrasound but sometimes it is diagnosed at birth. The survival rate for babies with omphalocele depends on the size of the defect, whether the baby is premature, if the sac ruptures, and if other problems are present.

Care during your pregnancy

A baby with omphalocele cannot be treated before birth. Your doctor will have a detailed plan to watch your pregnancy very closely. Your doctor will talk with you about how you will deliver your baby. If a cesarean section (C-section) is needed your doctor will discuss this with you. It is important to call your doctor if you have any questions or concerns during your pregnancy.

Ohio Fetal Medicine Collaborative

If this problem is found while you are pregnant, you will continue to receive care from your doctor and you may be referred to the Ohio Fetal Medicine Collaborative (OFMC).

Through the OFMC, you will see a doctor who specializes in the care of high-risk patients. You will also meet with a pediatric specialist at Nationwide Children's Hospital to learn more about your baby's care after he or she is born. Nurse coordinators will be

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available to help answer your questions and concerns; guide you through your pregnancy; and prepare you for what to expect.

The OFMC staff includes a Family Support Coordinator who provides emotional and spiritual support to parents and families before and after their baby's birth. This Coordinator also helps connect families with various hospital and community resources including chaplains, social workers and parent advisors.

You will deliver your baby at a hospital that is prepared to care for high-risk babies.

Care after your baby is born

At the delivery hospital, a neonatologist (a doctor who specializes in treating newborns) will supervise your baby.

- Your baby will receive care in the hospital's neonatal intensive care unit (NICU).
- A breathing tube will be placed if your baby is having difficulty breathing.
- The baby's sac of exposed organs is covered with warm, moist, sterile dressings or a sterile plastic bag.
- A tube will be put into your baby's nose or mouth. This tube goes into your baby's stomach and helps to keep the stomach empty.
- An intravenous line (IV) will be started in your baby's arm, hand, foot or scalp. The IV provides fluids, nutrients and antibiotics to prevent dehydration and infection.
- Your baby will be taken to Nationwide Children's Hospital, Main Campus, Neonatal Intensive Care Unit for further care.

Surgery

Your baby will need to have surgery. The goal of surgery is to place the abdominal organs back into the baby's body and repair the defect in the wall of the belly, if possible.

Primary Repair

Infants born with a small omphalocele have the surgery soon after birth. The omphalocele contents are returned to the abdomen. The muscle and skin are closed. If an infant has a large omphalocele and small abdomen, the abdominal muscle cannot be sewn together. Instead, the hole in the muscle is covered with Gore-Tex (an absorbable material) and the skin is closed over it.

Staged Repair

If the infant has a large omphalocele, the surgery may be done in stages.

The omphalocele sac is removed and a surgical pouch called a silo is placed around the intestines and liver. It is suspended over the baby. Slowly, over time, the intestines and liver are pushed back into the abdomen. This can take a few days or a few weeks. Surgery will be done right away if the sac breaks. When the intestines and liver can comfortably fit within the abdominal cavity, the silo is removed and the hole in the abdomen is closed.

Sometimes the omphalocele is so large that the contents cannot be placed back inside the infant's abdomen. The skin around the omphalocele grows and eventually covers the omphalocele. The omphalocele is wrapped, applying pressure over the sac so that the omphalocele is reduced in size. The abdominal muscles and skin can be repaired when the child is older.

Care after surgery

After surgery, your baby will receive care in the hospital's Neonatal Intensive Care Unit. Your baby will need to be on a breathing machine until he or she is fully recovered from surgery.

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Other treatments your baby will probably need after surgery are:

- Antibiotics to prevent infection
- Fluids and nutrients given through an IV
- Oxygen
- Pain medicines as needed
- A tube placed through the nose into the stomach to drain the stomach to keep it empty (nasogastric or NG tube)

Feedings are started once the intestines are fully healed after surgery. Feedings of breast milk or formula are started very slowly with either the bottle, nasogastric tube (NG) or a gastrostomy tube. The tube is removed when your baby's intestine works properly.

The total length of time your baby will stay in the hospital will vary. It will depend on whether your baby is born with other health issues. The usual length of stay is 4 to 8 weeks or longer depending on the health of your child after surgery. You may be able to take your baby home once he or she is feeding well and gaining weight.

Most of the time, surgery can correct omphalocele. How well your baby does depends on how much damage or loss of intestine there was, and whether your child has other birth defects.

Follow Up

Your baby will need regular follow-up appointments to measure growth, development and nutrition. You will want to find a local pediatrician to take care of 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.