

Omphalocele

What is an Omphalocele?

The muscles of the abdomen normally close by 14 weeks in pregnancy and an omphalocele occurs when this does not form correctly. Omphalocele refers to an opening in your baby's abdomen, which allows the abdominal contents (intestines, liver, stomach etc) to slip outside the baby's body into the umbilical cord. A see through membrane usually covers these organs. Omphaloceles are usually detected during routine ultrasound, and are small (contain intestine only) or large (containing more than intestine). Approximately 1 in every 3000-4000 pregnancies results in a baby with an omphalocele.

What Causes Omphalocele?

Nothing you did or didn't do during your pregnancy caused this to happen. The cause of omphalocele is unknown. It appears to be a random event that can happen to anyone.

What Does This Mean?

Approximately 30% of babies with omphalocele will have a chromosomal abnormality or syndrome. It has also been shown that 40-50% will have another health problem such as a heart defect or digestive difficulties. There are many other conditions that can be associated with omphalocele. It is important to check the baby for these other problems because it will help us tell you what it means for the development or survival of your baby.

What Other Tests Should We Consider?

Omphalocele is associated with chromosome abnormalities or problems such as Trisomy 18 or Trisomy 13. Some genetic problems make it hard for babies to live longer than a few hours, days or months after birth and some genetic problems are so severe that babies are unable to survive after birth. You will be offered an amniocentesis to look for genetic or chromosome problems. During an amniocentesis, a small amount of amniotic fluid is taken from the area around the baby and tested for these genetic problems. An amniocentesis may not find all genetic causes for omphalocele. Your doctor may also recommend a fetal echocardiogram, which is a detailed ultrasound of your baby's heart.

What Will Happen Around the Time of the Baby's Birth?

Your health care provider will discuss with you the best way for your baby to be born. Babies with omphalocele need to be born in a hospital with a neonatologist (specialist in newborn babies) and a paediatric surgeon (specialist in surgery for children). Babies with omphaloceles require care in a neonatal intensive care unit after they are born.

The team will stabilize the omphalocele and wrap it with bandages and a clear film or “saran wrap” to keep it clean and protected. Efforts will be made for you to see your baby after s/he is born.

Babies with large omphaloceles often have underdeveloped lungs despite being born close to their due date. Because of this they may need help from a breathing machine or ventilator.

What Are the Treatment Options?

There are three approaches to treating omphaloceles: primary repair, staged repair and the non-operative approach.

A primary repair operation usually occurs within 1-2 days of your baby’s birth. This will be done if your baby has a small omphalocele where the abdominal contents can easily be placed back in the baby’s abdomen. A staged repair involves the covering of the omphalocele with a special plastic pouch. Over 5-7 days the surgeons will gently push a little more of the omphalocele back into your baby’s abdomen until it can be closed with an operation.

Until your baby’s intestines have recovered from surgery, your baby will be unable to eat, and will be nourished with IV fluids. You can still breastfeed your baby, just not right away. If you wish to breastfeed, it is important to begin expressing or “pumping” milk as soon as possible after the birth. Once the surgeon is satisfied your baby’s intestines are working properly, your baby will be fed your milk. Sometimes it can take weeks before a baby with omphalocele can take all their nutrition in the form of milk.

More uncommonly, a non-operative approach is needed. This is usually the only option available for large omphaloceles. This is because there are more abdominal organs outside the body (usually liver, stomach and intestines) and often these babies do not have a big enough abdomen to fit everything back inside right away. In this case, your baby’s omphalocele will be covered with special cream to help grow new skin over the membrane, which covers the organs. Eventually, when your baby is older s/he will have the operation to place the organs back into the abdomen.

Your surgeon and neonatal team will discuss with you the best option for your baby.

What Does This Mean For My Baby’s Future?

If the chromosomes are normal and there are no additional birth defects other than the omphalocele, the severity of the defect depends entirely on size of the omphalocele. Small or medium sized omphaloceles are more easily repaired with a simple operation and a shorter stay in the neonatal intensive care unit. Large omphaloceles may require complex reconstruction over weeks, months, or even years.

Babies born with omphalocele can have long-term problems and often need regular follow up appointments after going home from the hospital. Your surgeon will monitor your baby over the next few years and you may have regular assessments to monitor your baby's development as well. Children with omphaloceles may have feeding difficulties throughout their childhood years but in general the outcomes are quite good.

What Do We Do Now?

You will meet with a doctor that specializes in high-risk obstetrics, and potentially a genetics specialist. You will also meet the neonatologist and the pediatric surgeon. These doctors will discuss with you in detail your options for further testing, discuss with you test results and provide you with treatment options. They will be able to answer any questions you may have. There is no right or wrong answer on what to do next. Your doctor will assist you in making an informed decision. We will support you in whatever decision you choose to make.

Where Can We Find More Information?

University of California
The Fetal Treatment Center – Omphalocele
<http://fetus.ucsfmedicalcenter.org/omphalocele/>

The Children's Hospital of Philadelphia
Center for Fetal Diagnosis and Treatment – Omphalocele Surgery and Repair
<http://www.chop.edu/service/fetal-diagnosis-and-treatment/fetal-diagnoses/omphalocele.html>