

# EXOMPHALOS

## What is exomphalos?

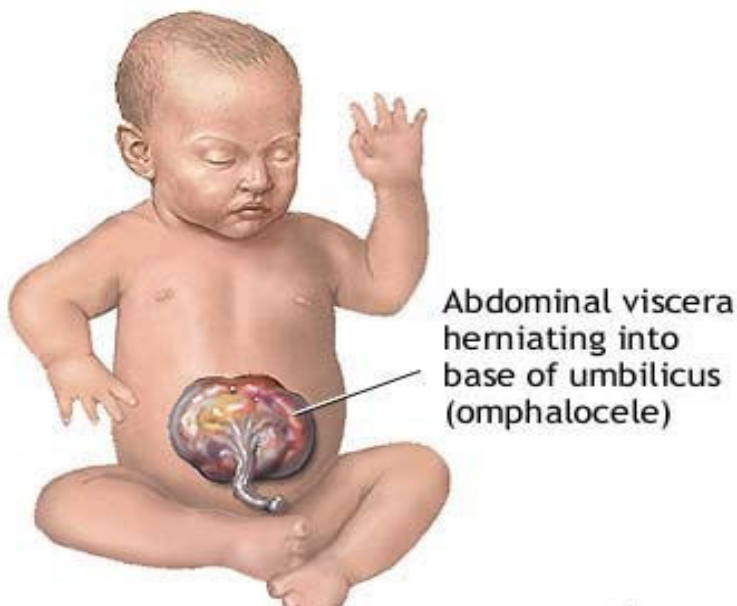
Exomphalos (also known as omphalocele) is a condition that occurs very early in pregnancy, usually between 3 to 8 weeks of gestation. During early pregnancy the intestine develops outside of the abdominal cavity in the umbilical cord and is supposed to return to the abdomen by 10 weeks. Exomphalos is a defect in the development of the abdominal wall that allows the intestine, and frequently the liver, to stay outside of the abdominal cavity in the sac of the umbilical cord.

## Why does exomphalos occur?

There is no known reason why in some babies a defect occurs in the abdominal wall allowing the intestines to develop outside of the abdomen. This condition occurs in 1 in every 6-10,000 pregnancies. Babies with an exomphalos frequently have other problems including abnormalities in the number of chromosomes (the structures in cells that contain our genetic material) and abnormalities of the heart.

## How is exomphalos diagnosed?

The majority of infants with an exomphalos are diagnosed in the antenatal period during routine ultrasounds. However, occasionally it may be diagnosed soon after delivery. Before birth, your obstetrician will look for other abnormalities that are known to be associated with exomphalos. Investigations may include an amniocentesis (procedure where the fluid surrounding the baby is withdrawn by a needle inserted through the mother's abdomen) which looks at the number of chromosomes and may diagnose some syndromes. Following these investigations further management of the pregnancy will be discussed with you.



[http://www.dhss.mo.gov/Genetics/TalkCornerArchives/11\\_07AbdominalWallDefects.html](http://www.dhss.mo.gov/Genetics/TalkCornerArchives/11_07AbdominalWallDefects.html)

**How is exomphalos treated?**

After diagnosis, your obstetrician will monitor your baby's health and the development of the liver and intestine carefully. The intestine can twist on itself and if this occurs is called a volvulus. If the blood supply to the intestine becomes blocked because of a volvulus it may be necessary to deliver your baby early to help save these areas of intestine.

After your baby has been delivered he/she will need to be looked after in the nursery. The baby can lose a lot of body heat and fluid through the intestines being outside of the abdomen. Your baby will need to be cared for in a warm environment and the intestines covered to prevent infection and fluid losses. As your baby will not be able to feed, intravenous fluids will be given to provide nutrition and antibiotics. A special intravenous line called a PICC line will be inserted to provide your baby with nutrition. As with all intravenous lines they have the potential to become blocked, tissue or migrate out of the vessel, but these possible complications are outweighed by the need for intravenous access for nutritional requirements. Most babies with an exomphalos will have an ultrasound of the heart (echocardiogram) to determine whether there are any abnormalities.

When your baby is medically stable he/she will be taken to the operating theatre and the paediatric surgeons will place the intestine and liver back into the abdominal cavity. This is usually done within 24 hours of birth, and often within the first few hours. The surgeons will need to look carefully at the intestines before putting them back into the abdomen to determine whether there has been a volvulus. If there has been a volvulus these areas of intestine may need to be removed and the two ends joined together.

In cases where a lot of the intestine and liver has developed outside of the abdomen the abdominal cavity may not be big enough and not all of the intestine and liver can be replaced at the initial surgery. This may mean that your baby will need to have more than one operation before all of the intestine and liver can be replaced into the abdomen. The appendix is normally in the lower right side of the abdomen, but with an exomphalos it may not be in this position. As this may cause problems with diagnosing appendicitis in later life, the appendix is usually removed at the time of the surgery.

**What happens after surgery?**

Once the intestine and liver have been returned to the abdomen, this may put pressure on the diaphragm and lungs and your baby may need help with their breathing. This may mean that your baby needs to be attached to a ventilator for some time after the surgery. Following surgery your baby's intestine needs time to recover and he/she will be fed with intravenous fluids. Your baby will then commence oral feeds, initially in small amounts. It may take several weeks for your baby's intestines to absorb the feeds properly, and during this time they will continue to receive nutrition from intravenous fluids. When your baby is ready to go home depends upon when they can tolerate all oral feeds. If you intended to breast feed your baby, this is still possible. The nursing staff will support you with expressing your breast milk and then breast feeding when your baby is able to tolerate oral feeds.

Infants who have a small exomphalos and no other medical conditions usually do very well and have no further problems. Very occasionally scar tissue may develop following the surgery, which may cause an obstruction and require further surgery. Infants who have a large defect of the abdominal wall and other medical complications may not survive for a number of reasons. The medical staff will discuss with you on a regular basis the outcomes expected for your baby.

If you have any further questions please ask the medical and nursing staff.

Approved by Canberra Hospital Neonatal Intensive Care Unit, 2012  
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