Exomphalos

Neonatal surgery

Patient information

What is exomphalos?

Exomphalos is a weakness of the baby’s abdominal wall, where the abdominal wall fails to close around the base of the umbilicus. This weakness allows the abdominal contents, mainly the bowel and the liver to protrude outside the abdominal cavity.

These organs outside the abdomen are contained within a sac which is a protective membrane. As the contents are lying outside the baby’s abdomen, the abdominal cavity often does not develop properly and remains small in size.

Why does exomphalos occur?

The cause of this condition is unknown. It is a rare problem and occurs in about four in every 10,000 births. Exomphalos is usually noticed on the ultrasound scan carried out between 18 and 21 weeks of pregnancy.

Up to 80% (eight in ten) babies with exomphalos will have other serious problems such as heart, lung, kidney defects and chromosomal problems. Detailed scans will be performed and some of the disorders such as heart problems can be diagnosed. Other disorders can only be diagnosed by tests such as chorionic villus sampling (CVS) or amniocentesis.

What happens at the delivery?

It should be possible for you to deliver your baby vaginally unless the Exomphalos is very large, or there are other reasons for requiring a caesarean section. We would recommend that your baby be delivered here at St George’s Hospital. Your baby will need to be transferred to the neonatal unit soon after delivery.
What happens next?

After your baby is born, the sac will be wrapped in a cling film to reduce heat and fluid loss. Your baby will not be able to feed for the first few days. If you wish to breastfeed, the nurses or midwives will show you how to hand express and store your milk, ready for when your baby is able to feed.

A drip will be placed into a small vein so that intravenous fluids can be given. A tube will be passed through your baby’s nose into the stomach to drain away the green fluid (bile) that collects in the stomach. This lessens the risk of your baby vomiting and reduces discomfort.

Your baby will be examined in order to identify any problems with other body systems and may need further tests. If there are associated lung problems assistance may be required with breathing.

When will my baby have surgery?

The size of the exomphalos can vary. It is therefore impossible to say exactly what type of operation is required until your baby is born and the actual size of the exomphalos can be seen. There are a number of different treatments, depending upon the size of the exomphalos:

**Primary repair**

If the exomphalos is small, it may be possible to return the organs back into the abdominal cavity and close the abdomen in one operation.

**Staged repair**

If the exomphalos is large, it may not be possible to close it in a single operation. Therefore, your baby would require an initial operation to construct a temporary covering of plastic sheeting (silo) outside the abdomen allowing the abdominal contents to gradually return to the abdomen over seven to ten days (approx). A final operation is necessary to close the abdominal wall. If the closure is very tight your baby may need to be on a ventilator for some days to help with breathing.

**Conservative treatment**

If the exomphalos is very large it may not be possible to repair it soon after the birth. It will be repaired when your baby is older. This is because the abdominal cavity is small and your baby will need to grow skin over the exomphalos to allow closure. This can take several weeks or months. Your baby will have daily dressings to protect the exomphalos and help the healing process.

Your baby may be able to go home before the exomphalos is fully covered by skin. The time that your baby spends in hospital with this condition will depend on the type of treatment needed and any other problems affecting other body systems.
Feeding

Some babies can feed within a few days of their operation but for other babies the bowels can take several weeks to work normally.

During this time your baby will need drip feeding (parenteral nutrition) through a long line. The long line is placed in a small vein in an arm or leg and fed through into a large vein. If you wish to breastfeed, the nurses or midwives will show you how to hand express and store your milk, ready for when your baby is able to feed. Milk feeds will be slowly introduced and increased as your baby is able to tolerate them. Once recovered your baby should be able to feed normally either by breast or bottle.

Long term and follow-up

Following discharge from the ward there will be regular check-ups in order to monitor your baby’s progress. Your baby will be seen in the outpatient’s department. It may be possible for this follow up to take place in your local hospital.

Following an operation there is always a small risk of a future obstruction occurring. If your baby has a bilious (green) vomit or a distended (swollen) tummy, medical advice should be sought.

If St George’s Hospital is not your local hospital

Once your baby has had surgery and made a good recovery, i.e. when their specialist medical and nursing requirements are less, the baby will be transferred back to the care of your local hospital. This transfer is a sign of progress and will not occur until the baby is ready. It will allow you to be closer to home and become familiar with your local healthcare professionals.