



Exomphalos

Information for parents and carers



Your baby has been diagnosed with a condition called exomphalos. We understand that this may be a worrying and stressful time for you. This leaflet explains a bit more about this condition.

Key messages

- Exomphalos is a condition where the baby's bowel develops outside the tummy
- Your baby may need an operation
- Most babies survive to become healthy adults

What is exomphalos?

Very early in all pregnancies the baby's bowel develops outside the tummy in the umbilical cord. The bowel usually moves back inside the tummy a few weeks later. In exomphalos there is a weakness of the baby's abdominal wall where the umbilical cord joins it. This means that some of the bowel and other organs (such as the liver) continue to develop outside the tummy in a loose sac which is part of the umbilical cord. An exomphalos can be small (known as exomphalos minor – figure 1) or larger (known as exomphalos major – figure 2).



Figure 1: Exomphalos minor



Figure 2: Exomphalos major

How common is exomphalos?

Exomphalos occurs in about 1 in every 3000 pregnancies in Scotland.

Could my baby have any other problems?

At least a third of babies (1 out of 3) with exomphalos have other problems, such as heart defects and chromosomal disorders such as Edwards' syndrome. These will present other challenges to your baby and may affect the treatment they are offered and their likelihood of survival.

The cause of exomphalos is unknown but we do know that it is becoming more common.

In any future pregnancies you are much more likely to have a normal healthy baby than to have another baby with exomphalos. If your baby has other disorders associated with exomphalos you will be offered genetic counselling to discuss the chances of a problem in future pregnancies.

What will happen during my pregnancy?

Exomphalos is usually found at a routine ultrasound scan. This is usually at your booking scan but can be at a later scan. Exomphalos would not have been diagnosed if you had an ultrasound scan before 12 weeks as it is normal to see the bowel outside the tummy at this stage. If exomphalos is suspected, you will be offered a further, detailed scan by a fetal medicine specialist. They will discuss the results of your scan and offer you further tests to check for chromosomal abnormalities. If exomphalos is diagnosed before 15 weeks of pregnancy you will be offered chorionic villus sampling. If the diagnosis is after 15 weeks you will be offered an amniocentesis. These are both tests offered during pregnancy to detect chromosome abnormalities, the most common being Down's syndrome. They can also detect other genetic conditions. An initial result from these tests should be available within two to three working days, but it can take up to 14 days to get the full result.

You will continue to have regular ultrasound scans throughout your pregnancy to monitor the exomphalos and any problems associated with it. You will also have the opportunity to speak to a neonatologist (a specialist in newborn medicine) and a paediatric (children's) surgeon. They have experience of caring for babies with exomphalos and what your baby's diagnosis might mean for you and your family.

If your baby has another major abnormality, or their lungs are very small, you may choose not to continue with your pregnancy and have a termination. Your obstetrician and midwife will support you through this. If you choose to continue with your pregnancy, the fetal medicine team will plan with you where, when and how your baby will be delivered.

What happens when my baby is due?

Babies with exomphalos are cared for at neonatal surgical centres at:

- Royal Aberdeen Children's Hospital;
- Royal Hospital for Children and Young People, Edinburgh; and
- Royal Hospital for Children, Glasgow.

Exomphalos needs to be assessed and managed quickly after your baby is born. If the exomphalos is small there is no reason why you cannot have a vaginal delivery, but decisions about the type of delivery you have will depend on your individual case.

What will happen after the birth?

General care

The exomphalos will be covered in a clear bag or cling film after your baby is born to protect the organs outside the body. Where possible, you will be able to have some cuddles with your new baby before they are transferred to the neonatal intensive care unit. They will be kept warm and a drip will be put into a vein to give them medicines and fluids. A nasogastric tube will be put down your baby's nose into their stomach to drain any fluid that collects there. Your baby will then be thoroughly examined for any other problems associated with exomphalos. Investigations usually involve chromosomal and genetic tests for particular syndromes.

Surgical care

Depending on the size of the exomphalos, your baby may need to have it repaired in one operation or in several stages. The paediatric surgeon will see you shortly after your baby is born and explain the treatment your baby will have. If your baby needs surgery, you will be asked to sign a consent form to show that you understand and agree to the operation. Figures 3 to 6 on page 5 show some examples of healing exomphalos major.

Exomphalos minor	Exomphalos major
<p>If your baby is stable they may have an operation soon after they are delivered. If the hole is too large to close, a patch may be used to close the muscle layer. This is usually permanently in place and your baby's own tissue will grow around the patch.</p> <p>The wound will be covered by a protective dressing.</p>	<p>It may not be possible to repair exomphalos major after birth, so we would wait till your baby is slightly older. This allows time for the tummy cavity to get bigger and for the baby to grow some skin over the exomphalos to allow us to close the hole in the tummy wall. Your baby will have regular dressing changes to protect the sac and help them heal. You will be able to go home with your baby before their wound has healed, as healing can take several weeks or months.</p>



Figure 3



Figure 4



Figure 5



Figure 6

What happens next?

All babies are closely monitored after the operation. While your baby's bowel recovers and starts to work, they might be fed through a tube into their veins (this is known as parenteral nutrition or PN). Once your baby's bowel is working normally, this will be gradually replaced by milk given through a nasogastric tube.

What can I do to help my baby?

There are lots of things that you can do to help your baby including cuddles, positive touch and reading to them. The staff will also support you to become partners in your baby's care so that when you feel comfortable you can help with things like nappy changes and nasogastric tube feeding. We call this approach family integrated care.

Breast milk is the best milk for all babies, especially babies born with exomphalos, and many mothers find this a positive way to help their baby. However we recognise that not everyone chooses this option and other milks (including donated breast milk and specialist formula) are available.

What is the likely outcome for my baby?

The chance of your baby surviving depends on whether they have other major problems, such as a chromosomal disorder or serious heart defect. If your baby only has exomphalos there is a 90% (9 out of 10) chance of survival. Their chance of survival is lower if their exomphalos is very big or they have other major problems. In a very small number of babies the bowel may not work properly for months and they will need to be fed through a tube into a vein for a long period. This is called intestinal (bowel) failure.

How long will my baby be in hospital?

A baby with a small exomphalos and no other problems will be discharged from hospital once they are feeding well and gaining weight. This often takes a couple of weeks. A baby with a large exomphalos which is having regular dressing changes can go home once you have been trained to dress the exomphalos and your baby is feeding well and gaining weight. This may take several weeks or possibly even months. Sometimes other problems such as a heart defect can keep your baby in hospital for even longer periods.

Where can I get further information and support?

The fetal medicine, paediatric surgical and neonatology teams will be able to answer any questions that you have during your appointments.

In 2018, the Surgery for Conditions Affecting Newborns in Scotland (SCANS) was formed. This is a national network of doctors, midwives, nurses and parents. It was set up to produce standards of care so that you and your baby get the best treatment. You can find more information at www.scans.scot.nhs.uk. We have listed other support groups below.

Other useful sources of information

Gastroschisis Exomphalos Extrophies parents' support group (GEEPS):
www.geeps.co.uk

SCANS collects anonymised data (information that you and your baby cannot be identified from) about the different conditions we treat. We use this information to plan and deliver services to children with these conditions, understand how we can improve patient care and provide it in the best way possible for all of our patients. All information is handled securely and confidentially by NHS staff and we do not share it with outside organisations. If you have any questions or concerns about the information we collect or your data protection rights, please email nss.scans@nhs.scot.

You can find more information about how the NHS handles your personal information at [NHS Inform](https://www.nhs.uk/infomart).