

Patient information factsheet

Exomphalos

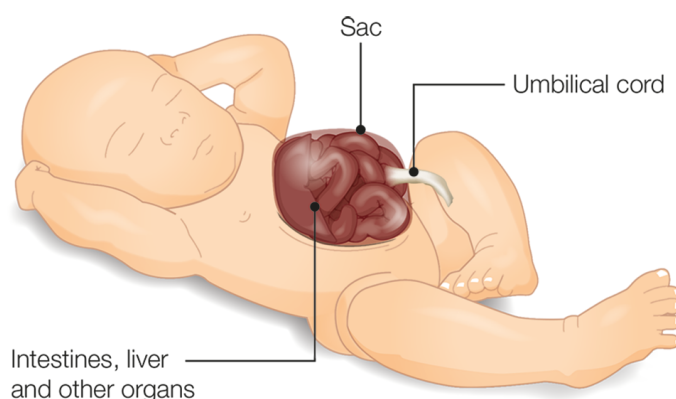
A scan has shown that your baby has a condition called exomphalos (also known as an 'omphalocele'). This factsheet has been designed to accompany the individualised discussions you will have about your care and the care of your baby both during pregnancy and after your baby's birth.

We hope it will help to answer some of the questions you may have. If you have any further questions or concerns, please speak to a member of the fetal medicine or neonatal surgical team.

What is exomphalos?

Exomphalos occurs when a baby's abdominal (tummy) wall does not fully close around the base of their umbilical cord (where the tummy button will form). This allows the organs that usually grow inside a baby's abdomen to grow outside of their abdomen in a protective membrane (a loose sac) that surrounds the umbilical cord.

Exomphalos usually affects a baby's intestines (bowel), but it may also affect their liver and other organs. The size of a baby's exomphalos will depend on the number of organs growing outside of their abdomen.



What causes exomphalos?

The cause of exomphalos is not always known. Exomphalos occurs in approximately one in every 2,500 births.

What does this mean for your baby during pregnancy?

Exomphalos can be associated with other physical conditions. This may be a condition affecting the development of your baby's heart, or a genetic or chromosomal condition.

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You will be offered a detailed scan of your baby to examine their development and growth. However, it is important to be aware that antenatal scans have limitations and are not always 100% accurate. You may also be offered a diagnostic test to determine whether or not your baby has a genetic or chromosomal condition. Diagnostic tests include chorionic villus sampling and amniocentesis. The fetal medicine team will discuss diagnostic tests with you in more detail. It is important that you take time to consider your options and ask any questions you may have before you decide whether or not having a diagnostic test is the right choice for you.

Your baby's wellbeing during pregnancy and after birth will depend on:

- the size of the exomphalos
- the presence of any additional chromosomal or genetic conditions

We will offer you regular ultrasound scans throughout your pregnancy to assess your baby's wellbeing. However, it is important that you become familiar with your baby's usual daily pattern of movements and contact your local maternity day assessment unit immediately if you feel that the movements have changed.

The birth of your baby

Place of birth

We recommend that you give birth to your baby on the main labour ward at Princess Anne Hospital in Southampton. Your baby will require extra care and support from our neonatal (baby) surgical team immediately after birth. If your baby's exomphalos is very large or they are born prematurely (before 37 weeks), they may need additional support with breathing. We will arrange for you to meet with the neonatal surgical team during your pregnancy. This will provide you with the opportunity to discuss the care your baby will receive once they are born, answer any questions you may have and have a tour of the neonatal unit.

Giving birth to your baby

Your baby's wellbeing will be closely observed throughout your pregnancy, and if concerns are identified, we may recommend your baby is born a little earlier than normal. If this is the case, your obstetrician may discuss induction of labour with you.

Although it is possible to give birth to your baby vaginally, if your baby's exomphalos is large, we may recommend having a caesarean section.

Your baby's care immediately after birth

When your baby is born, they will be cared for by the neonatal team who will assess their wellbeing and wrap the exomphalos in a plastic film. This will protect your baby's bowel and any other exposed organs. It will also allow the neonatal team to monitor it.

Once the neonatal team are happy that your baby is in a stable condition your baby will be transferred to the neonatal unit and nursed in an incubator or heated cot. If your baby's lungs have been affected by the exomphalos or through premature birth, they may also need help with their breathing.

Your baby will have a cannula (small tube) placed into a vein, to allow us to give them intravenous fluids via a drip, as they will not be able to feed at first. This tube will also be used to give your baby any medicines that they need.

If you are planning to breastfeed your baby, you will be given support to express and store your milk until your baby is ready for milk feeds.

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A small tube (nasogastric tube) will also be passed through your baby's nose or mouth, down into their stomach to drain away the fluid that collects there due to their bowel not working properly. This will help to stop your baby being sick.

The neonatal team will involve you as much as possible in your baby's care and will explain the reason for any treatment they are receiving. They will also be happy to answer any questions you may have.

Treatment

The size of your baby's exomphalos (and the organs it contains) can vary greatly. It is therefore necessary to wait until your baby is born before deciding on the most appropriate form of treatment for your baby. Treatment options include:

Primary repair

If your baby's exomphalos is small, it may be possible to place the contents of the exomphalos inside their abdomen, and repair their abdominal muscles and skin during one operation.

The timing of the operation will depend on your baby's wellbeing, but it will usually be performed in the first few days after their birth. Your baby will require a general anaesthetic (medicines used to send your baby to sleep).

Staged repair

If your baby's exomphalos is large and contains their liver, placing the contents of the exomphalos inside their abdomen, and repairing their abdominal muscles and skin during one single operation may not be possible. Instead your baby may be offered an operation to construct a temporary envelope of plastic sheeting (known as a silo). This silo will cover the organs exposed outside of your baby's abdomen. During your baby's stay on the neonatal unit, the silo will gradually be made smaller. This will push your baby's organs back inside their abdomen as they grow over a period of about 10 to 14 days.

A second operation is necessary to completely replace the contents of the exomphalos inside your baby's abdomen, and repair the abdominal muscles and skin. If closing the abdomen is difficult, your baby may need help with their breathing for a number of days after their operation. Your baby's surgeon may decide to only close the skin of the abdomen over the exomphalos and not repair the muscles. If this is felt to be the most appropriate course of treatment for your baby, they may have a bulge in the tummy wall (called a ventral hernia). A further operation to repair this hernia will be arranged when your child is a bit older, usually about three years old. In the meantime, your baby will be able to go home with you and adopt a normal lifestyle.

For all of these procedures, your baby will be given a general anaesthetic (medicines used to send your baby to sleep). We will also give your baby pain relief (analgesia) after their surgery to keep them comfortable.

Conservative management

If your baby's exomphalos is extremely large, it may not be possible to place the contents inside your baby's abdomen until your baby is older and the size of their abdomen has grown. Skin will gradually grow over the exomphalos over a period of several months. Your baby may be able to return home before the exomphalos is fully covered by skin.

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In this situation, your baby will have a ventral hernia (bulge of the tummy wall). An operation to repair this hernia will be arranged when they are about three years old.

The overall length of time your baby spends in hospital will vary depending on the type of treatment they need and whether or not they have any other conditions.

Feeding your baby

Introducing milk feeds will depend on how well your baby's bowel is working. Most babies are able to have milk feeds within a few days of their operation. You will be given help and support with feeding your baby.

Some babies cannot tolerate milk feeds straight away. If this is the case, your baby will be fed intravenously (straight into their veins) through a central line with a special drip called total parental nutrition (TPN). The central line will be placed in a small vein in your baby's arm or leg and fed through into a larger vein. You will be given a separate factsheet about TPN.

We will slowly introduce milk feeds through your baby's nasogastric tube, increasing the amount and frequency as your baby is able to tolerate them. As the milk feeds increase, the TPN will decrease until your baby is fully milk-fed. Once your baby's bowel has recovered, your baby should be able to feed normally by breast or by bottle.

Long-term care and follow up

After your baby leaves the neonatal unit, you will be offered regular appointments to monitor your baby's progress. We will try to arrange these appointments to suit your family's needs. In some situations, Southampton may not be your local hospital. If this is the case, we may transfer your baby's care to your local hospital. This will not happen until the neonatal team are happy with your baby's progress.

Most babies born with exomphalos (without additional genetic or chromosomal conditions) will make a full recovery after receiving treatment.

Contact us

If you have any questions or concerns, please contact us.

Fetal medicine team

Telephone: **023 8120 6025** (Monday to Friday, 9am to 5pm)

Surgical specialist nurses

Telephone: **023 8120 8564** (Monday to Friday, 9am to 5pm)

For urgent queries outside of these hours, please call the maternity triage line on:

0300 123 9001 (if your maternity care is provided by University Hospital Southampton NHS Foundation Trust) or call your local maternity day assessment unit.

Your GP, midwife and obstetrician may also be able to give you more information.

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Useful links

Antenatal Results and Choices (ARC)

ARC is a national charity that supports people making decisions about screening and diagnosis and whether or not to continue a pregnancy.

Website: www.arc-uk.org



GEEPS

A support group for parents of babies with exomphalos and gastroschisis.

Website: www.geeps.org

Wessex Healthier Together

Website: www.what0-18.nhs.uk/pregnant-women/concerns-during-pregnancy/over-20-weeks/reduced-baby-movements-after-24th-week-pregnancy

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For help preparing for your visit, arranging an interpreter or accessing the hospital, please visit **www.uhs.nhs.uk/additionalsupport**