

Abdominal wall defects: exomphalos (omphalocele)

Information for parents



Information to help you understand more about exomphalos.

1. What is exomphalos?

Exomphalos (sometimes known as omphalocele) is what doctors call an abdominal wall defect.

An abdominal wall defect is where a baby's abdomen (tummy) does not develop fully while in the womb (uterus).

With exomphalos, a baby's abdomen fails to close around the base of the umbilical cord. This means that some organs develop outside the abdomen.

It is usually just the intestine that is outside the abdomen, but sometimes the liver and other organs can also develop outside the baby's abdomen.

The organs outside the abdomen are contained within a sac which is usually covered in a protective membrane, so it is like a balloon at the base of the umbilical cord.

Up to 80% (8 in 10) of babies with exomphalos have other serious problems such as heart defects and chromosomal disorders.

Some of the disorders, such as heart problems, can be diagnosed by ultrasound scan. Others, which are suspected after an ultrasound scan, can only be diagnosed by tests such as chorionic villus sampling (CVS) or amniocentesis. There is more information on CVS and amniocentesis in leaflets called *Chorionic villus sampling (CVS) – information for parents* and *Amniocentesis test – information for parents*. These are available on our website at www.fetalanomaly.screening.nhs.uk/publicationsandleaflets.

2. How common is it?

The condition occurs in about 4 in every 10,000 births.

3. How is it diagnosed and confirmed?

Exomphalos is usually noticed at the Fetal Anomaly ultrasound scan carried out between 18 weeks and 21 weeks of pregnancy.

Sometimes it is noticed during a scan earlier in the pregnancy, but usually after 12 weeks. This is because a baby's abdominal wall does not normally close before this time.

You will need a second scan to confirm the diagnosis.

4. Is there any treatment?

All babies with exomphalos need an operation after they are born.

Your specialist doctor will talk to you about the type of treatment your baby will need, although this may not be clear until after your baby is born. Your baby's treatment will depend on the size of the exomphalos as well as whether your baby has other disorders.

5. What is the outlook for the baby?

The outlook for your baby depends on whether your baby just has an exomphalos or also has other disorders.

- If your baby just has an exomphalos there is a 90% (9 in 10) chance of them surviving. The chances of survival are lower if the exomphalos is very large.
- If your baby has other disorders, they may only have a 10% (1 in 10) chance of surviving. The chances of survival depend on how serious the other disorders are.

6. What happens next?

You will be given the chance to talk to specialists about what having a baby with exomphalos might mean for you and your family.

They will offer you another scan to check whether the baby has other disorders, and will discuss tests such as CVS (chorionic villus sampling) or amniocentesis to check for genetic disorders. There is more information on CVS and amniocentesis in leaflets called *Chorionic villus sampling (CVS) – information for parents* and *Amniocentesis test – information for parents*. These are available on our website at www.fetalanomaly.screening.nhs.uk/publicationsandleaflets.

Your doctor may talk to you about the option of having a termination to end your pregnancy. You will have the opportunity to discuss the possible implications of continuing or ending your pregnancy.

If you choose to continue your pregnancy, your healthcare team will help you plan how your care, including delivery, is managed.

You will be offered regular ultrasound scans to monitor your baby. Arrangements will be made for you meet some of the paediatric team, including one of the paediatric surgeons, who will care for you and your baby during pregnancy and after the birth.

There are a range of options for the delivery of your baby. Your health professional will discuss these options with you.

If you choose to have a termination, your health professional will talk to you about the procedure and support you through the process.

Whatever you decide, your decision will be respected and you will be supported by your midwife and doctor.

7. How likely is it to happen in a future pregnancy?

You are much more likely to have a normal, healthy baby in your next pregnancy than to have another baby with an exomphalos.

There is no way to prevent this condition. It is not due to anything you have or have not done.

Even when there are other disorders, the chance of your next baby having an exomphalos is also likely to be low, but this depends on the specific disorders.

If your baby had other disorders, you will be offered genetic counselling to discuss the chances and your options in future pregnancies.

8. Where can I get more information and support?

You may feel you only want to talk to your family and friends, or a particular doctor or midwife from the hospital. However, there are other people and organisations that can provide information, help you make your decisions and support you in your pregnancy and afterwards. You may also want to talk things through with the hospital chaplain or your own minister or faith leader.

9. Further information, charities and support organisations

The following organisations can offer you support. There are details of other support organisations on our website at www.fetalanomaly.screening.nhs.uk. If you have any questions about the information in this leaflet or where the information came from, email us at enquiries@ansnsc.co.uk.

Antenatal Results and Choices (ARC)

Email: info@arc-uk.org

Helpline: 0845 077 2290

Website: www.arc-uk.org

Antenatal Results and Choices (ARC) provides information and support to parents before, during and after antenatal screening and diagnostic tests, especially those parents making difficult decisions about testing, or about continuing or ending a pregnancy after a diagnosis. ARC offers ongoing support whatever decisions are made.

GEEPS

Email: geeps@btinternet.com

Website: www.geeps.co.uk

GEEPS is an international network of families and friends of children born with abdominal wall defects. GEEPS is run by the families and friends of affected children and is a non-profit-making network. The aim of GEEPS is to support families through the shock of diagnosis and beyond in the hope that some of the stress can be relieved by sharing thoughts and fears with other parents who have been in a similar situation.

This information has been produced on behalf of the NHS Fetal Anomaly Screening Programme for the NHS in England. In other countries, check with a health professional to find out whether there are any differences in approaches to screening.

This leaflet has been developed through consultation with the NHS Fetal Anomaly Screening Programme expert groups.

All of our publications can be found online at www.fetalanomaly.screening.nhs.uk.

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