

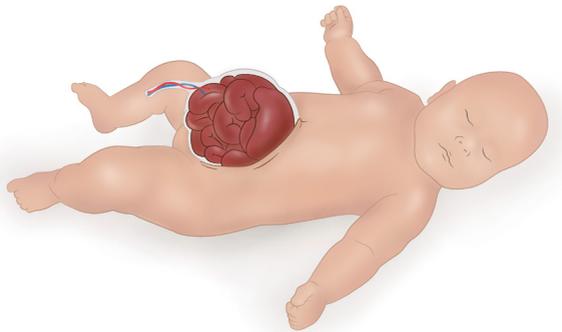
Information leaflet for parents and carers

Exomphalos



What is an Exomphalos?

Exomphalos is a weakness of the baby's abdominal wall where it joins the umbilical cord. This weakness allows the abdominal contents, (usually the bowel, and often with the liver) to protrude into a sac that surrounds the umbilical cord. As the contents are lying outside the abdominal cavity, then the abdominal cavity often does not develop properly and remains small in size.



What causes Exomphalos?

The cause of this condition is unknown and is a rare problem. About half of all babies with Exomphalos will have problems affecting other body systems. Most commonly affected are the heart, lungs, and kidneys. Detailed scans will be performed but it is not always possible to detect problems before your baby is born. There is also an increased risk of a chromosomal abnormality and you may be offered an amniocentesis or to have your baby's bloods taken to check the baby's chromosomes.

How is Exomphalos diagnosed?

Exomphalos is usually diagnosed on the ultrasound scan carried out between 18 and 21 weeks of pregnancy. After diagnosis ultrasounds will be carried out regularly to monitor your baby's growth.

How will my baby be born?

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. Your baby will need to be transferred to the neonatal unit soon after delivery.

Immediate care after delivery

After your baby is born, a drip will be placed into a small vein so that intravenous fluids and antibiotics 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 reduces the risk of your baby vomiting and reduces discomfort. If your baby has difficulty breathing they will be given breathing support.

Your baby will be examined in order to identify any problems with other body systems and may need further tests.

Feeding:

Breast milk is very important for every baby but especially sick babies and even more so for those with bowel/ stomach problems. Breast milk is far more than just food, it contains essential nutrients your baby needs to grow, develop, and recover from surgery or illness.

Although your baby cannot feed straight away, in order to establish and/or maintain your breast milk supply for when they are well enough to feed, we encourage you to express your breast milk. It is important that you start expressing milk within 6 hours of birth and continue to do this at least 8-10 times a day, including at least once every night.

There is a quiet, private area on the ward that you can use to breast feed your baby or express breast milk. Expressing milk can be done by hand, which is useful in the first few days after birth when you can expect to express small amounts of sticky clear or yellow fluid called 'colostrum'.

Electric breast pumps are available for use on the ward. Your expressed breast milk will be stored safely until your baby is well enough to feed. The nursing staff will be able to show you how to express and store your milk and arrange for you to have access to a breast pump.

Surgery:

The size of the sac and its contents can vary greatly from being very small to very large. It is therefore impossible to say exactly what type of operation is required for Exomphalos until your baby is born and the actual size can be seen.

Small Exomphalos

If the exomphalos is small and with no liver in the exomphalos sac then it is possible to do a primary repair. This is an operation under general anaesthetic within the first few days of life to return the intestines back inside the abdomen, and to close the muscles and skin.

Large Exomphalos

A large Exomphalos containing liver and bowel in the sac can sometimes be closed as a primary repair but more often surgery is delayed until 18 months to two years of age. This is due to the abdominal cavity being too small to put the intestines and liver back inside.

If surgery is delayed special honey dressings will be used to protect the sac and reduce infection while it heals. As it heals the skin will grow up and cover the exomphalos sac. This can take weeks up to a couple of months. Your baby will be able to go home before surgery and you will receive help and support to care for and dress the exomphalos at home until the skin has healed. Your child will be seen regularly by the surgeon in outpatients clinic. Surgery for a large exomphalos may involve one operation or for more complex cases it may require a number of operations known as a staged repair.

After Surgery:

Your baby will come back to recover either on the intensive care unit or our surgical ward at Birmingham Children's Hospital. You will be able to visit as soon as they are settled. All babies are closely monitored after the operation, and so your baby will be connected to monitors to check their breathing, heart rate and oxygen levels. If your child needs help with breathing, they will be nursed on the intensive care unit and connected to a ventilator. They will also be given pain relief through the intravenous infusion (drip).

While your child's intestines recover and start to work, they might be given fluid through a drip. This will gradually be replaced by breast or bottled milk when your child is able to tolerate this.

The nurses on the ward will encourage you to look after your baby as much as you feel able to while they are recovering. This can be daunting, especially while your baby is connected to drips and monitors, but it will become easier with time.

You will be able to go home or be transferred back to your local hospital once your baby is feeding properly and gaining weight.

Is there a risk to my babies life?

The outlook for babies with this condition depends mainly on the presence and severity of any other associated problems. These problems are not always apparent until after the baby is born. The majority of babies with an Exomphalos and no other problems survive and grow up to lead normal lives.

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. Your baby should be able to continue with feeding and weaning and progress as normal.

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

Is there a support group?

The support group for families of children with abdominal wall defects is:

GEEPS

104 Riverside Road
Romford RM5 2NS
Tel: 01708 738 134
Contact@geeps.co.uk
www.geeps.co.uk

BLISS

BLISS is a support group which is able to offer support to families with babies with a range of conditions

68 South Lambeth Road
London SW8 1RL
Helpline: 0870 7700 337
information@bliss.org.uk
www.bliss.org.uk

Looking after and sharing information about your child

We have a duty of care to help patients and families understand how information about them is kept and shared and we include the following information in all our patient leaflets:

Information is collected about your child relevant to their diagnosis, treatment and care. We store it in written records and electronically on computer. As a necessary part of that care and treatment we may have to share some of your information with other people and organisations who are either responsible or directly involved in your child's care. This may involve taking your child's information off site. We may also have to share some of your information for other purposes; such as research etc. Any information that is shared in this way will not identify your child unless we have your consent. If you have any questions and/or do not want us to share that information with others, please talk to the people looking after your child or contact PALS (Patient Advice and Liaison Service) on 0121 333 8403.

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