



# Exomphalos

**This information sheet explains about exomphalos or omphalocele, what causes it and what to expect when your child comes to Great Ormond Street Hospital (GOSH) for treatment.**

Exomphalos is a type of abdominal wall defect. It occurs when a child's abdomen does not develop fully while in the womb. Early in all pregnancies, the intestine develops inside the umbilical cord and then usually moves inside the abdomen a few weeks later. In exomphalos, the intestines and sometimes other organs such as the liver remain inside the umbilical cord but outside the abdomen.

## What causes exomphalos?

We do not know what causes exomphalos although we do know that they are becoming more common, affecting around two in every 5000 children born each year. Exomphalos can be associated with other problems but the doctors will examine your child closely to check if this is the case.

## What are the signs and symptoms of exomphalos?

Exomphalos is immediately recognisable because the child's intestines are outside the body covered in a membrane. The size of the bulging membrane containing the intestines and other organs varies from a small protrusion to quite a large lump.

There are two types of exomphalos: **exomphalos minor** where the opening is less than 4cm and only contains the intestines, and **exomphalos major** where the opening is greater than 4cm and/or with the liver inside the cord.

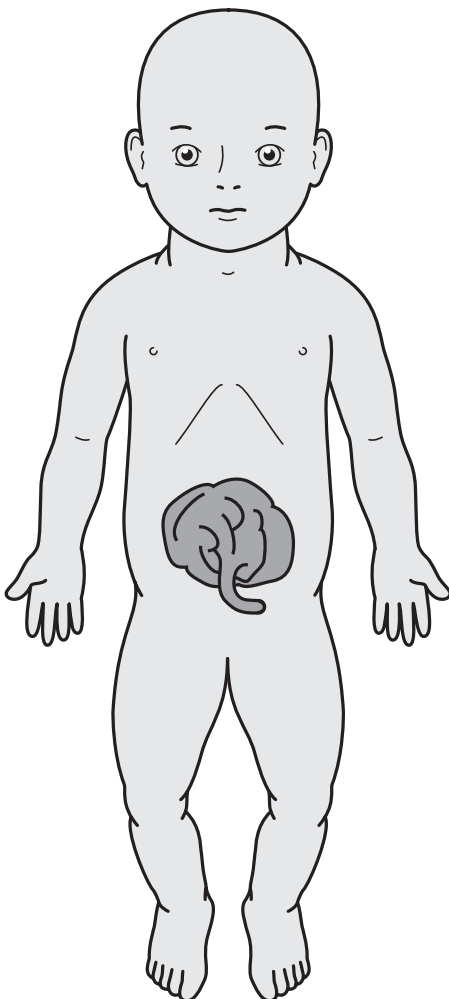
## How is exomphalos diagnosed?

In many cases, exomphalos is visible on prenatal ultrasound scanning, which is useful because it gives time for discussions and planning for when and where to give birth. Generally children are born naturally (vaginal childbirth) but some, especially if they have a very large exomphalos, may need a caesarean section.

## How is exomphalos treated?

Exomphalos is a serious condition so needs prompt treatment soon after birth. Children born with exomphalos are usually transferred to Great Ormond Street Hospital (GOSH) within a few hours of birth.

Immediately after birth, if the membrane covering the intestines is intact, your child will be kept warm and hydrated until they are





transferred to GOSH, either to our intensive care unit or another of our specialist wards. Depending on the size of the exomphalos, your child may need to have it repaired in one operation or in several stages. If the exomphalos is small and your child is stable, they may have an operation soon after transfer, where the surgeon replaces the contents back inside the abdomen and closes up the base of the umbilical cord. If the exomphalos is larger, contains the liver, and/or your child needs to be stabilised, doctors may place a silo or pouch over the intestines, which is closed over a period of days to weeks, to allow your child to grow so that there is room inside the abdomen.

### **What happens before the operation?**

The doctors will explain the operation in more detail, discuss any worries you may have and ask you to give permission for the surgery by signing a consent form. Another doctor will visit you to explain about the anaesthetic.

### **What does the operation involve?**

If your child is having a one-stage repair under general anaesthetic, the surgeons will replace your child's entire intestine into the abdominal space and close up the hole at the base of the umbilical cord. Sometimes, they may need to use a 'patch' of material if the hole is quite large. They will cover the area with a dressing to protect the wound while it heals.

In a staged repair, there is too much intestine outside of the abdomen to put back without causing further damage or the space inside the abdomen is too tight. While your child is under general anaesthetic, the surgeon will make a mesh sac and put it over the intestine which keeps it contained and protected. This sac is then suspended above your child so that gravity gradually moves the intestines back inside the abdomen. It is tightened regularly until all the intestine is inside the abdomen,

which usually takes a few days to a few weeks. Your child will then have an operation under general anaesthetic to close up the skin and muscles.

Occasionally, it is not possible to safely enclose all of the intestine inside the abdomen. If this is the case, the patch will be left in situ and the skin allowed to heal over the affected area forming a 'hernia', which will need treatment at a later date.

### **Are there any risks with the operation?**

All surgery carries a small risk of bleeding during or after the operation. During the operation, the surgeon will minimise any bleeding by sealing off the blood vessels affected. There is a very small chance that nearby structures in the abdomen could be damaged during surgery but this is a very rare occurrence.

Every anaesthetic carries a risk of complications, but this is very small. Your child's anaesthetist is a very experienced doctor who is trained to deal with any complications.

It can take a while after the operation for the intestine to start working properly so your child may need to be fed intravenously using total parenteral nutrition (TPN) for a while. This affects many children and is explained further in the next section.

Some babies with exomphalos have breathing problems which may require more support for a longer period.

### **Are there any alternatives to the operation?**

No – abdominal wall defects need to be repaired to prevent fluid and body heat loss from the exposed intestines and reduce the risk of damage. Ultimately, the condition needs to be treated to allow your child to grow and develop.



## What happens after the operation?

Your baby will come back to recover either on the intensive care unit or our surgical ward. 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 fed through a tube into their veins (total parenteral nutrition or TPN). This will gradually be replaced by breast or bottled milk, given through the naso-gastric tube when your child is able to tolerate this. As your baby recovers, you will be able to feed them from the breast or bottle. Over time, the drips and monitors will be removed one by one.

The nurses on the ward will encourage you to look after your baby as much as you feel able 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. If you are worried about caring for your baby, please talk to the nurses. You will be able to go home or be transferred back to your local hospital once your baby is feeding properly and gaining weight.

Before you go home or at your follow up clinic appointment, we might arrange for your child to have a contrast scan to check the position of the intestines inside the abdomen. Your local health visitor or community paediatric nurse will visit you regularly. We will send you details of your outpatient appointment in the post, soon after you leave hospital.

## When you get home

If your child is unwell, please take your child to your local hospital. The doctors there will discuss any concerns with the team at GOSH.

**If you have any questions, please telephone 020 7405 9200 and ask for the ward from which your child was discharged.**

## What happens next?

The outlook for children born with exomphalos varies depending on the size of the defect and any other problems. Many children have grown up to lead normal lives. Some children need to continue with TPN for a longer period so that they can gain weight to reach the right size and weight for their age. They may seem smaller than other children of the same age for the first few years but the majority catch up in time.

There is a very small chance that despite treatment the intestines may not work properly to absorb nutrients. This is called intestinal failure and requires long term TPN which can occasionally cause liver problems. Children who have had a exomphalos repair may develop hernias in the years after the operation. This is because the abdomen has fewer muscles than usual. If you notice a bulge in your child's abdomen, please talk to your doctor.

## Is there a support group?

The GEEPS (Gastroschisis, Exomphalos and Exstrophy Parents Support) group can offer help and support. You can visit their website at [www.geeps.co.uk](http://www.geeps.co.uk).