Belfast Health and Social Care Trust

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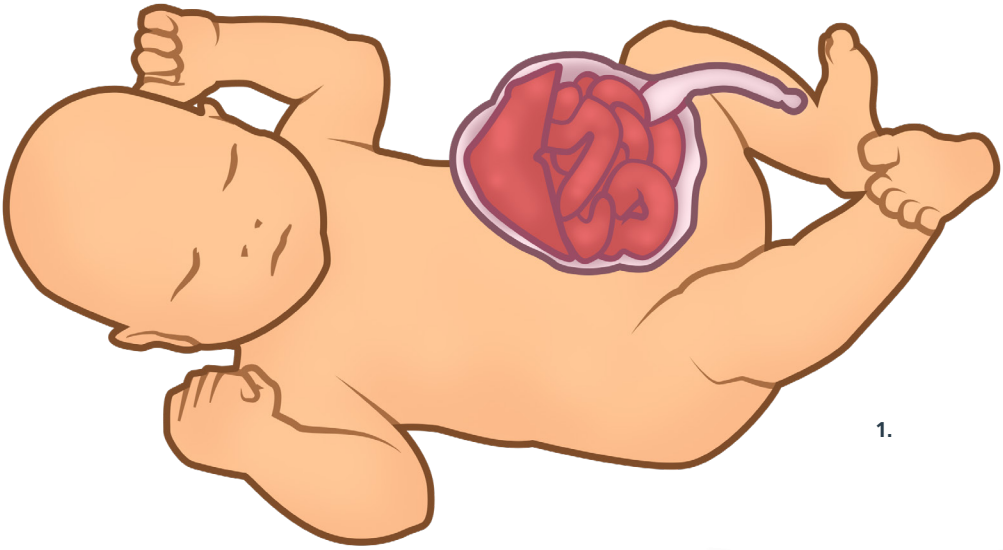
Exomphalos

Exomphalos is a type of abdominal wall defect. It occurs when a child's abdomen does not develop fully while in the womb. This page explains about exomphalos or omphalocele, what causes it and what to expect when a child comes to the Royal Belfast Hospital for Sick Children (RBHSC) for treatment.

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



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What causes exomphalos?

We do not know what causes exomphalos, although we do know that it is becoming more common, affecting around two in every 5,000 children born each year. Exomphalos can be associated with other problems, but doctors will examine children 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 and 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 containing the intestine, 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. Generally children are born naturally (vaginal childbirth) but some, especially if they have a very large exomphalos, may need a caesarean section.

Initial Management?

Exomphalos is a serious condition so needs prompt treatment soon after birth. Children born with exomphalos are usually transferred to Royal Belfast Hospital for Sick Children (RBHSC) within a few hours of birth.

Immediately after birth, if the membrane covering the intestines is intact, the child will be kept warm and hydrated until they are transferred to RBHSC, either to our intensive care unit or another of our specialist wards.

Depending on the size of the exomphalos, the infant may need to have it repaired in one operation or in several stages.

Primary repair: If the exomphalos is small, it may be possible to return the organs back into the abdominal cavity and close the abdomen in one operation.

Staged repair: If the exomphalos is large, it may not be possible to close it in a single operation. Therefore, your baby would require an initial operation to construct a temporary covering of plastic sheeting (silo) outside the abdomen allowing the abdominal contents to gradually return to the abdomen over seven to ten days (approx.). A final operation is necessary to close the abdominal wall. If the closure is very tight your baby may need to be on a ventilator for some days to help with breathing.

Conservative treatment: If the exomphalos is very large it may not be possible to repair it soon after the birth. It will be repaired when your baby is older. This is because the abdominal cavity is small and your baby will need to grow skin over the exomphalos to allow closure. This can take several weeks or months. Your baby will have daily dressings to protect the exomphalos and help the healing process. Your baby may be able to go home before the exomphalos is fully covered by skin.

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

What happens after the operation?

The child will come back to recover either on the intensive care unit or our surgical ward. All children are closely monitored after the operation, and so the child will be connected to monitors to check their breathing, heart rate and oxygen levels. If the 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 the 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 the child recovers, they will be able to feed from the breast or bottle. Over time, the drips and monitors will be removed one by one.

The nurses on the ward encourage families to look after their child as much as they feel able while they are recovering. This can be daunting, especially while their child is connected to drips and monitors, but it will become easier with time. If worried about caring for their child, families can talk to the nurses. They will be able to go home or be transferred back to their local hospital once their baby is feeding properly and gaining weight.

Before they go home, or at a follow up clinic appointment, we might arrange for the child to have a contrast scan to check the position of the intestines inside the abdomen.

A local health visitor or community paediatric nurse will visit regularly. We will send details of any outpatient appointment in the post, soon after leaving hospital.

When they get home
If the child is unwell, we suggest taking the child to a local hospital. The doctors there will discuss any concerns with the team at RBHSC.

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 live 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 a bulge in the child's abdomen is noticed, please talk to your doctor.

Contact Details

Barbour Ward (out of hours): 028 961 50337

Paediatric Surgical Secretaries

Miss McCullagh/ Mr Dick: 028 961 55679
Mr Philip: 028 961 6039
Miss Milliken/ Miss Lawther: 028 9504 7666

1. Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities. https://www.cdc.gov/ncbddd/birthdefects/omphalocele.html December 2019

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