

Gastroschisis and exomphalos

This leaflet gives more information about gastroschisis and exomphalos. If you have any questions or concerns, please speak to a doctor or nurse caring for your child.

What are gastroschisis and exomphalos?

They are 2 separate conditions that are frequently called abdominal wall defects. These happen when a baby's tummy (abdomen) does not develop properly in the womb. These conditions are usually found at your 20 week anomaly scan.

Gastroschisis is a defect of the abdominal wall, which results in your baby's bowel being outside of their tummy. This means the bowel is not protected by any membrane or sac.

Exomphalos is a defect at the base of the umbilical cord. It causes the bowel to push through (herniate) into the umbilical cord. In exomphalos, the bowel is protected by the membrane surrounding the umbilical cord.

Why do they happen?

Unfortunately, we do not know what causes these defects. There is no evidence that it is anything you have or have not done during the pregnancy. Gastroschisis is seen in about 5 in every 15,000 live births, and exomphalos in about 3 in every 15,000 live births.

Exomphalos is often associated with abnormalities affecting other body systems, but gastroschisis is generally an isolated problem. Your baby will be thoroughly checked and monitored for any other problems when they are born, by the medical team.

The delivery

We will plan for you to deliver your baby at St Thomas' Hospital. However, if you were to deliver at your local hospital, we would work with them on the plan for your baby. The neonatal transport service will be called to transport your baby to us.

Because there is a risk of reduced growth and wellbeing, particularly in months 6 to 9 (the third trimester), you will be closely monitored. Induction of labour is usually planned for around 38 week's gestation. You should be able to deliver naturally, unless the defect is very large or there are other medical reasons for needing a caesarean section.

Early management

In the delivery room

When you are ready to deliver your baby, the midwives will call for the neonatal team so that we are with you when your baby is born. A surgeon might also be in the delivery room, but this is not essential. When your baby is born the neonatal team will fully assess your baby and then wrap the exposed bowel in film to help protect it. This will also reduce heat and fluid loss. When your baby is stable, we aim to make sure you can see and cuddle your baby before they are moved to the neonatal unit. A parent or carer can go with the baby to the neonatal unit.

On the Neonatal Unit

Once your baby is on the Neonatal Unit, they will be weighed and placed in an incubator to help keep them warm and minimise fluid loss from the exposed bowel. Your baby will have a small tube (cannula) put into a vein so that we can give them intravenous (IV) fluids, as they will not be able to have milk feeds at first. This tube will also be used to give your baby any medicines that they need. Your baby will also have a small tube passed through their nose into their stomach. This is called a nasogastric tube (NGT), and will drain away the green fluid (bile) that collects in the stomach. This will reduce discomfort and lessen the risk of your baby being sick (vomiting).

Your baby will be examined by the neonatal doctors to check for any other problems. With gastroschisis and exomphalos, there can be complications such as infections and the bowel being blocked. During your baby's stay with us you will be kept fully informed of their progress.

Next steps

Soon after your baby is born, a surgeon will decide which treatment is best for your baby. They will talk to you about this, giving you the chance to ask questions, before getting your consent for any procedures that might be needed.

If the defect is small enough, it might be possible for the surgeons to put the bowel back into the abdomen soon after birth. This is called a primary closure. If this is not possible a staged approach (explained below) will be taken to repair the defect.

Gastroschisis

With gastroschisis a bag (silo) may be used, which covers and protects the bowel. The silo is usually put in place by the surgeon on the Neonatal Unit soon after the birth of your baby if a primary closure cannot be done. The bowel will then be gradually 'tucked' back into the abdomen by the surgeons until all of the bowel has been returned. This process can take several days. When this is done, the bag can be removed and the hole in the abdominal wall which the bowel came through can be closed. This can be done on the ward by applying a dressing which remains in place for 10 days, or the surgeons might take your baby to the operating theatre for this final stage.

What about feeding?

We will need to put in a central line. This is a drip which is put into a small vein in your baby's arm or leg and fed through into a larger vein. This line is needed so that we can give your baby a special IV fluid called total parenteral nutrition (TPN). TPN will be important for your baby as it provides key nutrients that they will not be getting while they are not allowed to eat. Feeding is unlikely to be started until the gastroschisis is closed.

Once milk feeds are started, it is usually a few weeks before your baby can tolerate any milk feeds. This is because their gut might not work properly to start with, because it has been exposed to amniotic fluid during the pregnancy, and has been handled. Your baby will start with small amounts of milk, which will gradually be increased through the NGT. As the milk feeds increase, the TPN will decrease until your baby is fully milk fed. Reaching full feeds can be something that takes some time and there may be many times when your baby's feeds are stopped or reduced as their gut slowly begins to work properly.

Exomphalos

The treatment for exomphalos is different from that for gastroschisis because the bowel is already covered and protected. If a primary closure cannot be done with exomphalos, we will probably use a special cream and dressing and wait for the sac to harden and the skin to grow. The time this takes will depend on the size of the exomphalos. Once there has been enough growth, the surgeons will repair the defect completely.

What about feeding?

As with gastroschisis, a long line will need to be inserted to give TPN to your baby. However, with exomphalos it is possible to start milk feeds before the defect is completely closed. This will depend on how your baby's bowel is working. Because of the time it takes for the skin to grow around the exomphalos, your baby might leave hospital before the defect is fully repaired. The neonatal and surgical teams will involve you in discussions and will give you information and support to help you prepare for this.

Long-term and follow-up

The Evelina London Children's Hospital might not be your local hospital. If this is the case, we might transfer your baby's care to your local hospital. This will not happen until the surgeons and neonatal team are happy with your baby's progress.

After your baby leaves hospital, they will have regular check-ups in the outpatient department to monitor their progress. This might be at your local hospital. Your baby should be able to feed and wean normally, but this might be slower than with other children. Some babies can also be slower in gaining weight and might have problems with constipation, but these are normally short-term problems.

More sources of information

Antenatal Results and Choices (ARC), **web:** www.arc-uk.org **tel:** 0845 0772290 or 020 77137486

Contact, **web:** www.contact.org.uk

NHS Fetal Anomaly Screening Programme, **web:** www.fetalanomaly.screening.nhs.uk

Contact us

If you have any questions or concerns about anything in this leaflet, or abdominal wall defects in general, please contact the Neonatal Unit, **tel:** 020 7188 4045.

For more information on conditions, procedures, treatments and services offered at our hospitals, please visit **web:** www.guysandstthomas.nhs.uk/leaflets

Evelina London Medicines Helpline

If you have any questions or concerns about your child's medicines, please speak to the staff caring for them or contact our helpline.

tel: 020 7188 3003, Monday to Friday, 10am-5pm, **email:** letstalkmedicines@gstt.nhs.uk

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