HSC Belfast Health and Social Care Trust

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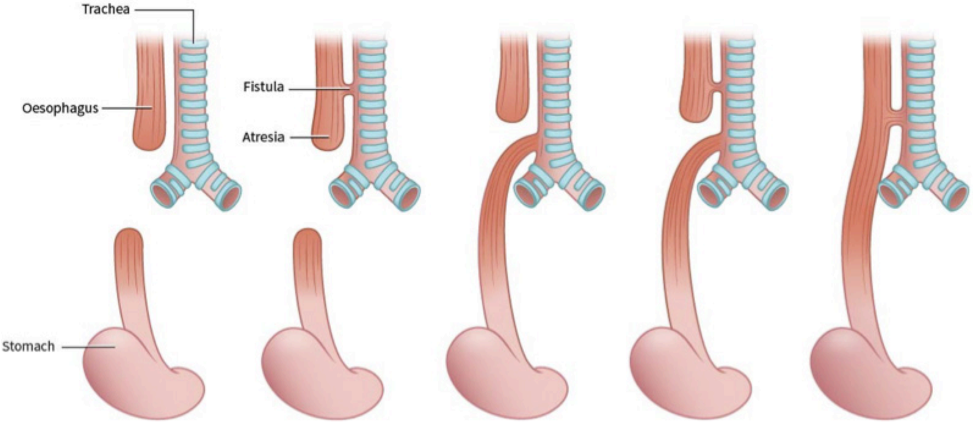
Oesophageal Atresia with Tracheo-Oesophageal Fistula

Oesophageal atresia (OA) is a rare condition where a short section at the top of the gullet (oesophagus) has not formed properly so is not connected to the stomach. This means food cannot pass from the throat to the stomach. Tracheo-oesophageal fistula (TOF) tends to occur alongside oesophageal atresia. This is where part of the oesophagus is joined to the windpipe (trachea). This page explains about oesophageal atresia and tracheooesophageal fistula, how they are treated and what to expect when your child comes to Royal Belfast Hospital for Sick Children for treatment.

How are OA and TOF diagnosed?

Oesophageal atresia and tracheo-oesophageal fistula: different types



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Both OA and TOF tend to be diagnosed soon after birth. The midwife may try to pass a tube through your baby’s nose into their stomach (naso-gastric tube or NG tube) and find that it is not possible. Occasionally, OA and TOF might be discovered in an ultrasound scan during pregnancy, particularly if the stomach is not very clear on the scan.

What causes them?

We do not know what causes OA and TOF. It is not due to anything that happened during pregnancy. It is very rare, occurring in between one in 3500 and one in 5000 births. OA and TOF can occur in different combinations (figure 1) - the most common forms are shown below. OA and TOF can be associated with other problems, so doctors will examine your child closely to check if this is the case.

How are OA and TOF repaired?

They are both repaired in an operation, which lasts between two and three hours.

Are there any alternatives?

No. OA and TOF always require treatment, to enable your baby to feed.

What happens before the operation?

Your baby will be transferred to RBHSC from the maternity hospital where he or she was born. To begin with, your child will have a Replogle tube passed through his or her nose into the oesophagus. This will drain off any saliva (spit) which is in the oesophagus. He or she will also have an intravenous infusion (drip) of fluids and medicines. You child may be nursed in the intensive care unit, depending on his or her general condition.

The surgeon will explain about the operation in more detail, discuss any worries you may have and ask you to give permission for your child to have the operation by signing a consent form. An anaesthetist will also visit you to explain about the anaesthetic.

What does the operation involve?

The operation is carried out while your child is under general anaesthetic. There are two ways of carrying out the operation: using thoracoscopic (keyhole) surgery or open surgery. The surgeon will discuss the most appropriate method with you. To repair the TOF, the surgeon will separate the oesophagus from the trachea and repair the part of the trachea where the oesophagus was originally joined.

The method used to repair the OA depends on the distance between the ends of the oesophagus. In most cases, the surgeons will cut the blind end of the oesophagus and then join the two ends together to form a continuous passage from the throat to the stomach.

In rare cases called ‘long gap OA’ where the distance between the ends of the oesophagus are too large for the surgeon to be able to join them straightaway, different treatment is needed. If this is likely to be the case, the doctors will explain this to you.

Are there any risks?

All surgery carries a small risk of bleeding during or after the operation. Even if the gap between the ends of the oesophagus is quite small, it can still be difficult to join together. After the operation, the join may leak or it may narrow over time, but these can both be treated in another operation. Your child will be reviewed regularly for a long time after the operation.

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.

What happens afterwards?

Your baby will go to the intensive care unit to recover. You will be able to visit as soon as he or she is settled back in the incubator. For a while after the operation, your baby will need help with breathing so will be connected to a ventilator. Some babies may be kept still with a muscle relaxant for several days in intensive care, all babies are closely monitored after the operation and be given pain relief through the 'drip'. Your baby will be transferred to a surgical ward at RBHSC when he or she no longer needs intensive care.

While your baby's intestines recover and start to work, he or she will have an intravenous drip of fluids. Feeding usually starts a few days after the operation, with breast or bottled milk given through a naso-gastric tube (tube passed through a nostril and into the stomach). Some babies' intestines take a little longer to recover, so need to be fed through a tube into the veins (total parenteral nutrition or TPN). Naso-gastric feeding is tried when his or her intestines start to show signs of recovery. As your baby recovers, you will be able to feed him or her from the breast or bottle.

Once your baby is feeding properly and gaining weight, you will be discharge home or transferred to your local hospital. Soon after your baby leaves hospital we will send you a letter with details of your outpatient appointment.

You should seek urgent medical help from your family doctor (GP), local hospital or team at RBHSC, if your baby:

- Is coughing or choking when feeding
- Has difficulty in swallowing saliva or feeds
- Is failing to gain weight

What is the outlook for children with OA and TOF?

If the OA and TOF occur on their own, with no other associated problems, the outlook for children who have them is good, with the majority growing up to live normal lives.

However, there are a couple of problems which may occur and which you should know about. They tend to happen most often in the first few years after the operation and improve as the child grows older.

- Chest problems can occur, which are sometimes serious enough to need a stay in hospital. These tend to improve with age and have little effect on the child's breathing when older.

- The area of the trachea which was repaired may become floppy (tracheomalacia) which can cause breathing problems. If your child's breathing is very noisy or he or she is having 'blue spells', please take your child to your local hospital and contact the team at RBHSC. Some children however, continue to have a distinctive 'TOF cough' but this is not at all serious. A further operation called an aortopexy is rarely needed - we will give you further information if required.

- Feeding problems may also occur due to the oesophagus narrowing where it was originally repaired (strictures). Sometimes it needs widening (dilatation) and this will be done in an operation under general anaesthetic.

- Some children also complain of problems with swallowing and need to have a drink with all food. This is often caused by poor oesophagus and swallowing coordination (working together). This can be investigated by a speech and language therapist.

- Some children who have had OA and TOF develop a problem with **gastro-oesophageal reflux** when they are older. This is where the contents of the stomach flow back up the oesophagus causing pain and irritation. In many cases, we start babies on an anti-reflux medicine after the operation to reduce the risk of this developing.

- **Weaning** onto solid foods can also be more problematic for children who have had OA and TOF. It can take longer for children to manage food with chunks and certain foods, such as white bread, can cause problems. Children should remain on Stage 2 foods (puree with soft lumps) for longer than usual to reduce the risk of choking. Feeding should also be done under close. Our Dieticians and speech therapists will advise you on weaning.

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Unofficial guide to Surgery: Core Operations, Chapter 12

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