Great Ormond Street Hospital for Children
NHS Foundation Trust

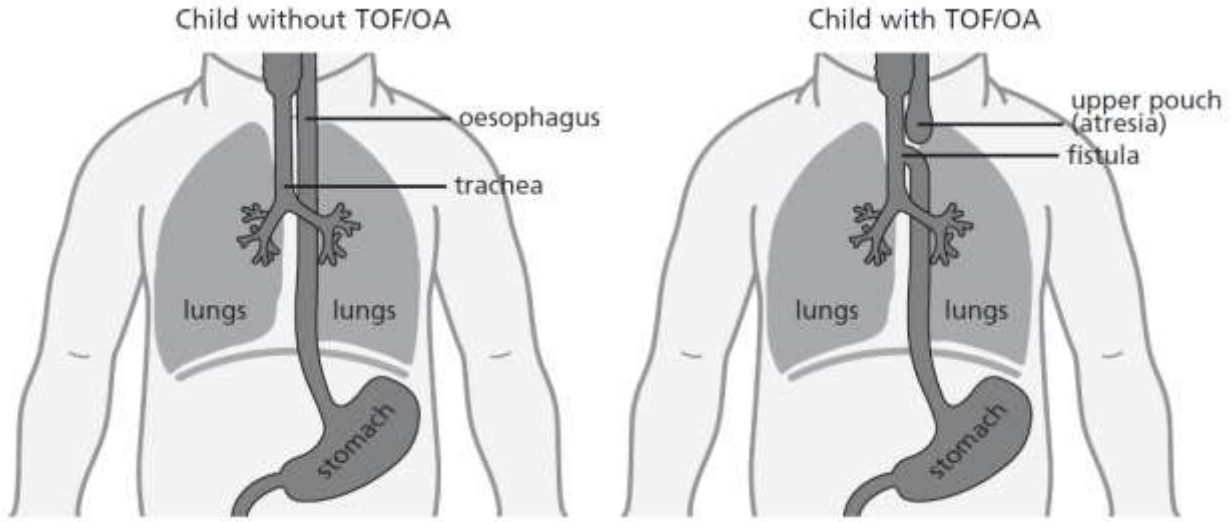
After your child has had oesophageal atresia and/or tracheo-oesophageal fistula repair: information for families

Oesophageal atresia (OA) and tracheo-oesophageal fistula (TOF) are both congenital (present at birth) problems. They can develop together or separately and are usually diagnosed soon after birth (or occasionally during a prenatal scan). Both conditions require repair in an operation under general anaesthetic lasting two to three hours.

Once the repair has taken place, it can take some time for your child to completely recover. This information sheet from Great Ormond Street Hospital (GOSH) explains what to expect during the recovery period as well as throughout childhood and adolescence.

Oesophageal atresia (OA) is a rare condition where a short section at the top of the oesophagus (gullet or foodpipe) 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) is another rare condition, which tends to occur alongside oesophageal atresia. This is where part of the oesophagus is joined to the trachea (windpipe).



Reflux

Normally, the oesophagus squeezes food down towards the stomach using an action called ‘peristalsis’. Gastro-oesophageal reflux (GOR) occurs when some of the food or stomach contents are squeezed back up the oesophagus. As the stomach contents are acidic, this can cause irritation.

GOR is more common in TOF babies, partly due to the abnormal development of the oesophagus but also due to the ring of muscle (sphincter) between the oesophagus and stomach being pulled upwards during the repair which weakens the anti-reflux mechanism.

Reflux can also lead to breathing difficulties and frequent chest infections as the food moving back up the oesophagus can enter the lungs. While this can cause lung problems in the long term, it often improves as the reflux is treated.

The severity of reflux varies greatly from one baby to another. Symptoms can include appearing uncomfortable after a feed, back arching or frequent vomiting after a feed. If left untreated, reflux can damage the lining of the oesophagus, which can lead to the development of strictures (see later section).

All babies at GOSH are given anti-reflux medication following OA/TOF repair. In addition, keeping babies upright after feeding can help. You can also raise the head of the cot so that your baby is sleeping at an angle – do this by raising the cot legs on blocks or putting a wedge under the head of the mattress. Never use a pillow directly under your child’s head.

Tracheomalacia

This is a condition where the trachea is floppy and collapses in on itself when breathing. This is common in children who have had OA/TOF repair as there are accompanying problems with how the trachea formed in the womb. For instance, the cartilage framework that holds the trachea together may not have formed properly leading to weak areas. Children who have had a TOF repair may also have areas of weakened cartilage where the oesophagus was disconnected from the trachea.

Usually the first signs are increased noise, effort and rate of breathing. A baby may also suffer from blue episodes when drinking. Children are at increased risk of developing chest infections and they may also find they cannot keep up with their friends as they have reduced exercise stamina. In severe cases, it can cause continuous serious breathing difficulties.

In extreme cases, a child may have ‘near death episodes’ where they seem to be choking and unable to breathe. This is why we teach you basic life support (BLS) skills and advise anyone else looking after your child to have training as well.

If an episode occurs, the child passes out which may open up the airway but mouth to mouth resuscitation should always be attempted.

Children with mild tracheomalacia may grow out of it without treatment. However, in more severe cases, an operation (aortopexy) may be needed to hold the trachea open.

‘TOF cough’

Children who have had OA/TOF repair can develop a distinctive cough, often referred to as a ‘TOF cough’. The distinctive sound is caused by coughing through the floppy section of the trachea. They also need to cough more often, as lining of the trachea is missing specialised cells that trap dust particles and clear them from the airway.

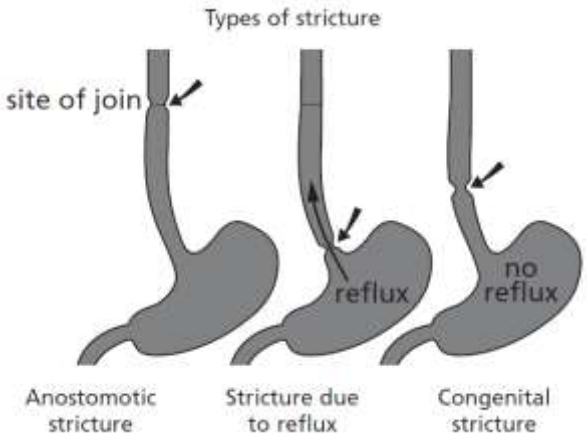
Chest infections

Children who have had an OA/TOF repair do not usually have more frequent infections than other children but are more likely to need treatment due to the problems coughing as described above.

As it is difficult for children to move mucus out of the airway, it may remain in the lungs and grow bacteria (germs), which will require treatment with antibiotics. They may also be more sensitive to viral infections that affect the respiratory system – these may cause tightening of the airway making breathing more difficult which can be scary but is similar to asthma so responds to similar treatment with inhalers.

Strictures

Narrowing can happen when scars or strictures develop following surgery to the oesophagus. This is common after all types of digestive system surgery, but especially following OA/TOF repair at the area where the atresia was opened and joined to the rest of the oesophagus.



The narrowed oesophagus can cause additional problems with food with bits or lumps, or hard foods that are not completed chewed before swallowing. The body will try to get rid of it by coughing – this is a built in self defence mechanism, but it can be scary for the child and anyone near them.

Narrowing of the oesophagus is usually treated with a procedure called oesophageal dilatation, which is always carried out while your child is under a general anaesthetic. Once your child is under general anaesthetic, the doctor passes a catheter (soft plastic tube) containing a balloon down the back of your child's mouth into their oesophagus. They watch where the catheter is by using X-rays and continue to pass it down the oesophagus until it reaches the narrowed section.

Once it is in place, the doctor inflates the balloon so that it stretches the narrowed section. Further X-rays are taken to check how much the balloon is inflated. At the end of the procedure, the balloon is deflated and with the catheter is brought back up the oesophagus and out of your child's mouth. The doctors may suggest a series of dilatations so that the oesophagus is gradually widened as this often gives the best long-term results.

Long-term follow-up

Children who have had OA/TOF repair will carry on coming back to GOSH regularly throughout childhood and adolescence. You will meet different members of the team depending on your child's needs – for instance, involvement from our dietitian or speech and language therapist may be helpful in addressing any feeding problems. When your child starts to approach adolescence we will discuss moving on to adolescent or adult services for their long term care. This is a gradual process to enable them to become more independent, taking charge of their own health and everyday life.

Further information and support

Talk to a member of the team if you have any questions.

You may also want to contact TOFS, the national support group for anyone affected by OA/TOF. Call them on 0115 961 3092 or visit their website at www.tofs.org.uk