About OA, TOF, and VACTERL

A leaflet provided courtesy of TOFS. Particularly useful for new parents of babies born unable to swallow.

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lifelong support
for those born
unable to swallow

Contents

Summary of Content

This leaflet is particularly useful for new parents of babies born with oesophageal atresia (OA) and tracheo-oesophageal fistula (TOF).

It introduces OA and TOF, explains why surgery is necessary for these conditions and outlines the likely course of events after surgery.

Basic information about VACTERL association, of which OA and TOF may be features, is also provided.

Cover photo: Sonny McGarry, born with OA and TOF

What is OA? What is TOF?

Oesophageal atresia (OA) and Tracheo-oesophageal fistula (TOF) are conditions which affect around 1 in 3,500 births.

Most people do not realise how early in pregnancy the body's organs start to develop.

The oesophagus (food-pipe or gullet) is the tube that allows us to swallow food and drink. It passes from the back of the mouth, down through the chest, to the stomach. The oesophagus starts to form as early as four to six weeks after the egg is fertilised. Whether the child will have OA is determined before many women even know they're pregnant.

The trachea (windpipe) is a separate tube through which air passes into the lungs.

The oesophagus and the trachea initially form as a single structure. During normal development this structure divides into two, with the part at the front forming the trachea and the part at the back forming the oesophagus.

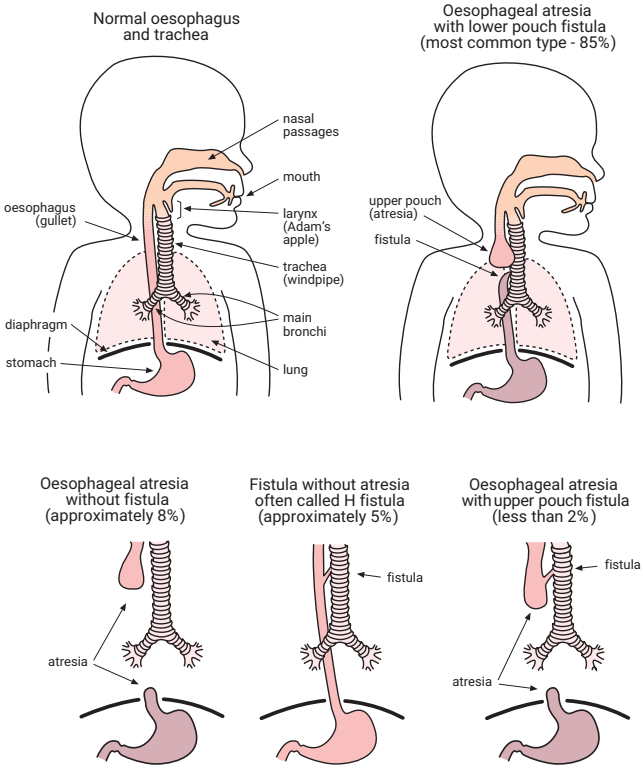
In babies born with oesophageal atresia/tracheo-oesophageal fistula, (OA/TOF), this separation has not happened as it should.

A baby with oesophageal atresia has a gap in his or her oesophagus. This means they are unable to swallow saliva, or milk. This inability to swallow saliva is often the first sign after birth that a baby has OA. The baby is often frothy or bubbly.

A baby with a tracheo-oesophageal fistula (TOF) has an abnormal connection between the food pipe and the airway.

Hearing that your baby has OA/TOF can be very alarming for parents. The first thing to understand is that you have done nothing wrong. We don't fully understand the cause of OA/TOF but we do know it is nothing to do with anything the mother or father have or have not done during the pregnancy.

We hope that this leaflet will give you some basic information, so that you know what to expect. For most parents their first concern is whether their baby will survive. It will be reassuring to hear that almost all babies with OA or TOF survive – prematurity or the presence of serious heart anomalies are the main factors which will affect their chances.



OA and TOF can occur on their own or together. Most 'TOF babies' actually have both OA and TOF with the fistula (connection) occurring from the oesophagus below the atresia, though other variations can occur.

Nurses and doctors at the hospital will explain how your baby is affected. You might get them to draw a circle around the picture relevant to your baby.

Transfer to another hospital

Your OA/TOF baby needs treatment at a specialist centre where the staff are experienced with OA and TOF. If your baby has not been born at a specialist hospital, he or she will be transferred and if you are fit to be discharged from your local hospital you may be able to go there along with, or soon after, your baby.

There is a possibility that you may not be able to sleep at the specialist centre initially. If you are unable to be transferred with your baby, the father, or a close relative may be able to go along with the baby and keep you informed.

The need for surgery

OA/TOF babies need surgery within the first few days of life. Your baby will first be checked to ensure that there are no other problems which might affect his or her ability to undergo surgery. When it comes to the operation, the surgeon aims to close the connection between the oesophagus and the trachea (the TOF) and then join the two ends of the oesophagus.

VACTERL

Sometimes OA/TOF babies have additional problems, most commonly those described as 'VACTERL' – a group of anomalies that often occur together. The incidence of VACTERL is about 1 in 20,000 - 35,000 births. VACTERL babies have a combination of three or more of the following:

- **V** = Vertebral (spinal) defects
- **A** = Anal (back passage) defects
- **C** = Cardiac (heart) defects
- **T** = Tracheo-oesophageal fistula
- **E** = Esophageal atresia (American spelling)
- **R** = Renal (kidney) defects
- **L** = Limb defects

It is usual to check for heart or kidney problems using ultrasound scanning; medical staff will explain any findings to you. More information about VACTERL is available from TOFS.

What causes OA and TOF?

Although there can be genetically inherited causes for OA/TOF, most babies with OA/TOF are an isolated case within a family and have not inherited the condition. This is an important fact for new parents to appreciate.

Very rarely, OA and TOF can feature as part of a more complex genetic condition.

The risk to future pregnancies is very small (between 1% and 3%). You can ask your GP to refer you to a genetic counsellor for further advice.

The operation

Correction of OA/TOF involves an operation on the chest to divide the fistula and then repair the atresia. This operation is performed by a paediatric surgeon. Your baby will be in theatre under a general anaesthetic with an experienced paediatric anaesthetist continuously monitoring the baby's vital signs; heart rate, ECG (the electrical activity in the heart), blood pressure, oxygen saturation (the oxygen-carrying capacity of the blood) and body temperature.

In most cases, a primary anastomosis (or primary repair of OA) is possible, ie, the two ends can be joined up immediately. The aim is to repair the oesophagus with as little tension as possible at the site of the join.

Sometimes, the gap in the oesophagus is too great to close. If this is the case, the surgeon will divide the fistula and place a gastrostomy tube into your baby's stomach for feeding. The oesophagus may grow so that it can be joined later, or an alternative way of repairing the gap will be used.

After the operation, the surgeon will report back to you. If tension at the anastomosis is high, (ie, it is a tight join), it is standard practice to protect the repair by keeping the baby sedated and paralysed (using anaesthetic medications) and ventilated (using a machine to breathe for the baby) for five to seven days after surgery. If the anastomosis was made with minimal tension, the baby will usually be woken soon after the procedure and returned to the neonatal intensive care unit (NICU) breathing on their own.

Dealing with your feelings

To be told that your baby has been born with a condition that requires immediate surgery is frightening.

People react differently in such situations. Support from those around you gives you the time and space to come to terms with things. Being aware of your feelings and communicating them to others will help too.

Having an unwell baby puts an extra strain on your life, but you – like many before you – will cope. TOFS can offer you contact with parents who have been through it all before, to lend a friendly ear and offer information and support.

On the ward

The special unit where babies are cared for after surgery can be unnerving to many parents. Other very sick babies will be on the ward and there is a lot of medical equipment, which can initially be very frightening. You will be encouraged to touch your baby and they will be reassured from hearing your voice as you speak to them.

The nurse responsible for your baby will explain to you what all the tubes and leads are for, and you should soon become more comfortable with the environment. As progress is made, less monitoring will be required and you will become increasingly involved in your baby's care.

Getting information

In the first days and weeks you may find it hard to take in all the information you are given. Never be afraid to ask questions. It is important that you understand everything you need to know and that you are comfortable with the care of your baby before you go home.

Some people may be better at explaining things than others, and nursing staff often have more time to talk to you than the doctors.

Making a list of queries may help, so that you remember what to ask when the opportunity arises.

There is a an easy-to-follow short video animation for new parents on the TOFS website, that gives a good overview of what OA/TOF might mean for your child.

What to tell others?

Friends and family will share your shock and concern, but may show this in different ways – or they may simply not know what to say or do.

Asking for what you want from them – be it time alone, someone to talk to, or perhaps some practical help – can be hard, but will be better for you, and in fact may help them to feel of some assistance at this difficult time.

Taking your baby home

Taking your baby home from hospital is a big step, but one that you should look forward to. After the initial trauma, you will need privacy and the chance to enjoy parenting in your home.

The medical staff will work with you to prepare you and your baby for discharge. This will give you the knowledge and confidence to handle any difficulties that may arise.

It's easy, whilst under stress, to forget what you have been told, so don't worry about asking staff to repeat information.

You should leave hospital feeling reassured that you know who to contact for support, help, advice and/or supplies.

Attendance at clinics for vaccinations, developmental checks and routine health care is as important for an OA/TOF baby as for any other child.

Although your baby may well need some specialised care, in other respects he or she needs exactly the same attention as any other baby.

Your GP, health visitor and other advisors in your local area have probably never looked after an OA/TOF baby before.

The surgical unit will contact them before you go home to provide information about any potential problems and any medicines, equipment or special supplies.

Follow-up appointments will be required to check on your baby's progress and to answer any questions you may have. These may either take place at the surgical unit or at a local hospital.

TOFS offers a range of leaflets and a comprehensive book about aspects of OA, TOF and VACTERL. These may be useful to you, your doctor and anyone involved in the care of your baby as he or she grows up.

Feeding your baby

From a medical point of view there is no reason why OA/TOF infants cannot be breastfed once oral feeding is permitted. In fact, breastfeeding is encouraged, and can take many forms including exclusive breastfeeding or expressing breastmilk (EBN) to bottle/tube/peg feed. With time, your baby will move on to puréed food, lumpy textures and eventually solid food – like any other baby – however this process may take a little longer.

After surgery for OA, the oesophagus does not function totally normally, which means that food is not transported to the stomach as efficiently as it might be. Holding your baby relatively upright during feeds will help.

Weaning can begin in children born with OA/TOF at the same age as other infants. Children are particularly receptive to new tastes from around four to six months of age. Difficulty coping with different textures and lumpy food may mean that soft puréed baby foods have to be given for longer. When lumps are introduced, plenty of liquid to drink should be given with meals, and the child must be reminded to chew food well and not to hurry.

The social aspect of meal times is important to all of us, and eating with others has an effect of increasing food intake, both quantity and variety.

If your child coughs and splutters, help them calmly to recover and then continue eating.

If this is a recurring problem you might need to make food softer and it is something you should explain to your consultant next time you visit clinic.

Many OA/TOF children have specific 'problem foods' during their early years. Often these are peculiar to the individual and can only be found out by trial and error, but there are useful tips... for example, adding sauce or gravy to make food moist and avoiding foods with skins such as sausages and apples.

Further information on weaning is available from TOFS.

Reflux

There are a few common problems which can occur after surgery, of which reflux is one.

Gastro-oesophageal reflux – to use its proper name – describes a condition where acidic stomach contents are pushed back into the lower oesophagus, causing feeding problems and other troublesome symptoms. Most babies reflux but OA/TOF babies are particularly prone to it.

Your follow-up appointments will monitor your child for reflux, which is usually managed using a combination of simple practical measures and prescribed medicines.

Strictures

OA/TOF children may develop oesophageal strictures - a narrowing of the food tube at the site of the join. Strictures are more likely to occur in the early weeks/ months following the repair, though can occur beyond this period too. Strictures can be made worse by reflux.

Strictures cause a deterioration in feeding. Ask your consultant if you suspect this is happening. Strictures are treated by dilating (stretching) the narrowing under a general anaesthetic.

Respiratory problems

Many OA/TOF children have some degree of tracheomalacia (caused by a floppy trachea) which causes a characteristic harsh, barking cough, often referred to as 'the TOF cough'. The noise associated with the cough can come as a surprise to others.

Tracheomalacia can also make it more difficult for children to shift secretions during a viral illness such as a cold. This can also cause OA/TOF children to be more susceptible to chest infections.

Mild tracheomalacia, and associated problems, invariably gets better as the child physically grows. Rarely, severe cases need further surgery.

Long-term outlook

Some OA/TOF children will go home and have few problems following surgery. Other children experience feeding difficulties and chest problems, and a few may require further surgery. Problems generally get less with time - some issues disappear and children learn to cope with any restrictions.

The results of long-term follow-up studies suggest that the majority of children born with OA/TOF are able to participate fully in sport and fulfil their academic potential.

Most adults born with OA/TOF enjoy normal lifestyles and careers, and many become parents themselves. However a significant minority experience chronic health problems which continue into adult life. Those born with OA/TOF often suffer a higher-than-normal level of respiratory infections and other respiratory difficulties.

Some feeding problems remain, for example, eating slowly and needing to drink with meals to help food pass down. Reflux can remain an issue throughout life and quite often those born with OA/TOF need to stay on anti-reflux medicine long term.

Lifelong follow up is essential for maintained good health.

Membership of TOFS

If you have concerns about your child's condition, the first people you must talk to are the staff at the surgical unit.Follow-up for OA/TOF children is important and appointments at the hospital must be kept to ensure continued improvement.

By joining the charity TOFS (Tracheo-Oesophageal Fistula Support) parents/ carers get the chance to talk to others who have seen their TOF child grow up; and adults have the support of others sharing similar experiences. This can be very reassuring.

TOFS offer a number of member services:

- A comprehensive range of leaflets and support information for parents, carers, adults, teachers and medical professionals.
- Our newsletter "Chew" where parents and adults can share their stories, and where we can keep you in touch with information, news and events.
- Regular online and in person events, to meet other members and hear from healthcare professionals who work with OA/TOF/VACTERL patients and families.
- Access to TOFS Local Contacts, (TLCs) - parents who volunteer in areas across the UK and provide support for members.

TOFS needs funding to help children, adults, and families to sponsor research into the cause and treatment of OA, TOF and VACTERL. Please remember us when you are organising fundraising events.

Some parents/carers do not feel ready to join TOFS straight after their baby's birth. We will be here whenever you choose to join and will be happy to hear from you. More information about TOFS membership, or making a donation, can be found on our website at: tofs.org.uk

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tofs.org.uk/oa-tof-information/resources/about-oa-tof-and-vacterl

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tofs lifelong support for those born unable to swallow

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