

# About TOF, OA and VACTERL

This leaflet provides information for parents of babies born with tracheo-oesophageal fistula (TOF) and oesophageal atresia (OA).

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

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



*Freddie Clamp-Gray, who was born with TOF, enjoying his 2nd birthday party with the help of Wotsits, thin crust pizza and potato salad.*

*A leaflet provided courtesy of TOFS (Tracheo-Oesophageal Fistula Support) – helping children born unable to swallow, with thanks to the Round Table.*



**TOFS**

## What is TOF? What is OA?

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

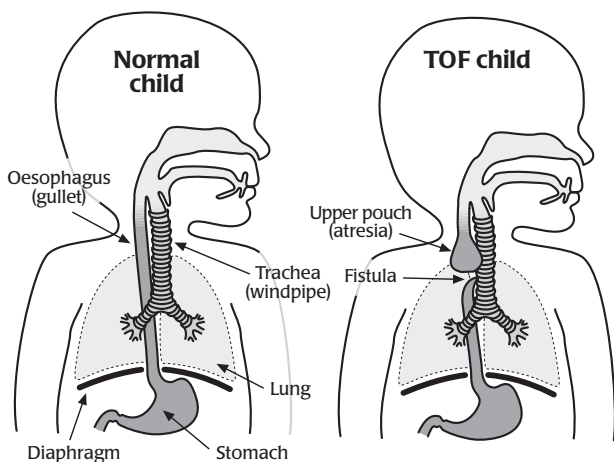
A normal baby has two distinct and separate tubes running from the mouth into the body: the oesophagus (gullet) is the food tube, taking food from the mouth to the stomach, and the trachea (windpipe) is the airway, through which air moves when we breathe in and out.

‘Atresia’ is taken from the Ancient Greek word meaning ‘no way through.’ A baby with oesophageal atresia (OA) has a gap in his or her oesophagus, so that milk – or anything else swallowed – cannot reach the stomach.

‘Fistula’ is from the Latin meaning ‘pipe.’ In tracheo-oesophageal fistula (TOF), there is a join between the food tube and the airway.

These conditions can occur either on their own or together; most so-called ‘TOF babies’ have both TOF and OA, with the fistula (join) occurring below the atresia (gap) in the oesophagus, as shown below.

Some other variations are shown opposite.



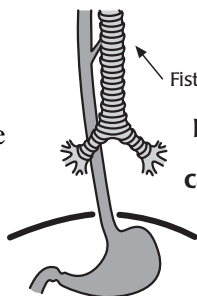
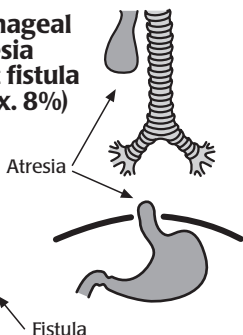
Staff at the hospital will explain what the problem with your baby is to you. You might like to draw a circle around the picture relevant to your baby.

The news of this condition can be very alarming for parents. We hope that this leaflet will give you some basic information, so that you know what to expect and do not worry about things which may never happen.

For many, the first concern is whether or not their baby will survive.

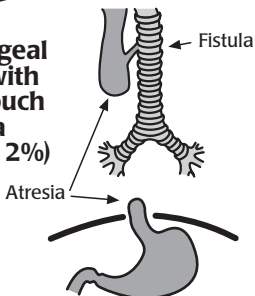
It should be reassuring to hear that most babies with TOF or OA survive – only prematurity or the presence of other, more serious anomalies will affect their chances.

**Oesophageal atresia without fistula (approx. 8%)**



**Fistula without atresia; often called "H" fistula (approx. 5%)**

**Oesophageal atresia with upper pouch fistula (less than 2%)**



## Transfer to another hospital

Your TOF baby needs treatment at a specialist centre where the staff are experienced in the treatment of TOF and OA. This is likely to mean that mother and baby are separated, since such hospitals generally cannot care for mothers immediately after giving birth. However, the father, or a close friend or relative, may be able to go with the baby and can then keep the mother informed about progress.

## The need for surgery

**T**OF babies need prompt surgery. The surgeon aims to connect up the two ends of the oesophagus, and to close the connection between the oesophagus and the trachea.

Your baby will first be checked over to ensure that there are no other problems which might affect his or her ability to undergo surgery.

## VACTERL

**M**any TOF babies are also born with other problems, most commonly those described as 'VACTERL' – a group of anomalies which often occur together. VACTERL babies have a combination of three or more of the following:

- V = vertebral (spinal) defects
- A = anal (backpassage) 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.

## The operation

**A**fter the operation, the surgeon will report back to you. Your baby will be taken to a special unit so that he/she can receive extra care.

Very occasionally the gap in the oesophagus is too great to close. If this is the case, the surgeon will place a tube into your baby's stomach for feeding, and will then explain to you what will happen next. The oesophagus may grow so that it can be joined up later, or an alternative way of closing the gap will be used.

## What causes TOF and OA?

It is not known what causes TOF and OA, although we do know that these problems are the result of a disturbance in the division of the top end of the primitive gut into a digestive tract (oesophagus) and a respiratory tract (trachea).

Extremely rarely, TOF and OA are associated with specific genetic defects, but this is not the normal situation. Usually TOF and OA occur as a 'one-off' case. Nobody is 'at fault' – it just happened. It is very important to recognise and acknowledge this, so we'll say it again: this was not your fault, you could not have prevented it.

The risk to future pregnancies is very small (between 1% and 3%). If you are worried, seek genetic counselling to put your mind at rest.

## Dealing with your feelings

To be told that the eagerly anticipated joyful event – the birth of a perfect baby – has not been fulfilled, is very frightening.



People react differently in such situations, but often there is so much going on, with decisions to be made and other day-to-day matters to be dealt with, that somehow the initial stressful period passes. 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

**T**he 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.

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

**I**n 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 with him/her.

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 don't forget what to ask when the opportunity arises.

## What to tell others?

**F**riends and family will share your shock and concern, but may show this in different ways – or 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

**T**aking 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 which may arise. You should leave happy 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 a 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 a 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. The hospital's neonatal surgical outreach nurse will then provide information and assistance to your local health-care team on an ongoing basis.

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 about aspects of TOF, OA 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 TOF infants cannot be breast fed once oral feeding is permitted. With time, your baby will move on to pureed 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 (‘food tube’) 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 need not be delayed and solids can be introduced from 3 to 4 months. Introducing variety between the ages of 4 and 6 months particularly helps to stimulate an interest in food and in different flavours and textures.

Difficulty coping with lumpy food may mean that soft, pureed baby foods must be given for a while longer. When lumps are introduced, fluid should be given with meals. Older children may need reminding to chew food well and not to hurry.

Eating with others is an important social event; a child who is having problems should be allowed to recover and continue eating – rather than being excluded from the rest of the meal.

Many 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 is valuable for swallowing food trouble free.

Hospital staff will offer you advice on feeding TOF children, and further information 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. Many babies reflux, however 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

TOF children may develop oesophageal strictures – a narrowing of the food tube at the site of the join which occurs during healing. Strictures can also be made worse by reflux.

Strictures cause eating problems, and are treated by stretching or ‘dilating’ the narrowing while the child is under a general anaesthetic.

## Respiratory problems

Many TOF children have a characteristic cough, often referred to as ‘the TOF cough.’ This is due to a weakness in the wall of the trachea at the site of the tracheo-oesophageal fistula and usually causes no problem to the child – although the noise associated with the cough can come as a surprise to others.

TOF children can also be susceptible to chest infections and asthma.

## Long term outlook

Some TOF children will go home and have very little in the way of problems following surgery; however others may experience feeding difficulties and chest problems, and a few may require further surgery. The problems are always at their worst in the first two years of life, and problems after five years of age are uncommon.

The results of long-term follow-up studies are very reassuring; the majority of TOFs are able to participate fully in sport and to fulfil their academic potential. Their achievements as adults are not impaired by their earlier problems. Some symptoms of feeding problems may remain, however any difficulties are generally mild – for example, requiring a drink with meals in order to wash down any slight obstructions – and adults generally do not feel their lifestyle is restricted.



*ABOVE: Vicki Martin was born in 1966 with TOF/OA. Her parents experienced many of the problems common to TOF families today.*

*As an adult, Vicki has few eating problems – although prefers to have a drink to hand at meals. She enjoys an active lifestyle and is not restricted by having had TOF/OA as a child.*

*Vicki works as a medical illustrator/graphic designer, and created the book 'The TOF Child' for TOFS.*

## Membership of TOFS

Never forget that if you have concerns about your child's TOF condition, the first people you must talk to are the staff at the surgical unit. Follow-up for TOF children is important and appointments at the hospital must be kept to ensure continued improvement.

By joining TOFS (Tracheo-Oesophageal Fistula Support) parents get the chance to talk to others who have seen their TOF child grow up and move on. This can be very reassuring.

TOFS offers a number of member services:

- reduced prices on our book *The TOF Child*.
- a comprehensive range of leaflets for parents, carers, teachers and medical professionals.
- a quarterly newsletter where parents can voice their worries, and where we can keep you in touch with news and events.
- a biennial conference.

TOFS constantly seeks further funds to help TOF parents and to sponsor research into the cause and treatment of TOF, OA and VACTERL.

Many parents do not feel ready to join TOFS straight after their baby's birth. We will be here whenever you choose to contact us and will be happy to hear from you.

More information about membership of TOFS, or making a donation, can be gained from from the TOFS web site [www.tofs.org.uk](http://www.tofs.org.uk) or from the TOFS office.



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*For additional copies of this booklet, or for more information about TOFS, please contact:*

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