

What is TOF/OA?

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What is wrong in TOF/OA? Variations

THE NORMAL OESOPHAGUS

The oesophagus (gullet or 'foodpipe') is the passage through which food moves on its route from the mouth to the stomach. It starts in the neck, just behind the larynx (Adam's apple), and ends below the diaphragm where it joins the stomach at an acute angle.

OESOPHAGEAL ATRESIA (OA)

The word 'atresia' is taken from ancient Greek and means 'no passage / no way through.' Thus in oesophageal atresia there is a break in the continuity of the oesophagus. The end nearest the mouth is not attached to the end which enters the stomach, the gap usually occurring high up in the chest. The presence of a blind-ending pouch in the upper oesophagus means that food is unable to reach the stomach; any swallowed milk or saliva instead returns to the mouth.

THE NORMAL TRACHEA

The trachea (windpipe) starts at the larynx (the voice box, seen from the outside as the 'Adam's apple' in the neck) and passes in front of the oesophagus before it enters the upper chest, where it divides into two tubes, the main bronchi, which go to the right and left lungs.

TRACHEO-OESOPHAGEAL FISTULA (TOF)

A fistula, from the Latin meaning 'a pipe,' is an abnormal connection running either between two tubes or between a tube and a surface. In tracheo-oesophageal fistula it runs between the trachea and the oesophagus. This connection may or may not have a central cavity; if it does, then food within the oesophagus may pass into the trachea (and on to the lungs) or alternatively, air in the trachea may cross into the oesophagus.

RIGHT:
illustrations depicting
the arrangement of the
oesophagus and trachea in a
normal baby, and in a baby
born with TOF/OA.

ATRESIA WITH LOWER POUCH FISTULA

In 85% of cases the upper oesophagus ends blindly, usually in the upper part of the chest. The lower part starts at the fistula from the trachea, which is located near the point where it divides into the main bronchi.

ATRESIA WITHOUT FISTULA

In 8% of cases there is no fistula. These babies are often more difficult to treat because the gap between the two ends of the oesophagus is usually very wide and the stomach very small.

FISTULA WITHOUT ATRESIA

Here the continuity of the oesophagus is not affected, there being no atresia, however there is a fistula joining the oesophagus and the trachea. This variation occurs in about 5% of cases and is often called an 'H fistula,' although the fistula is nearly always oblique and therefore more like an 'N.'

ATRESIA WITH UPPER POUCH FISTULA

Here the fistula runs from the upper sac of the oesophagus to the trachea. This is one of many variants making up the remaining 2% of cases.

DOUBLE FISTULA

Occasionally there may be more than one fistula present.

LARYNGEAL CLEFT

In this very rare condition the normal separation between the air passage and the oesophagus fails to form, so that food spills over into the windpipe on swallowing.

This information has been written for the parents of TOF children by TOFS (Tracheo-Oesophageal Fistula Support) – helping children born unable to swallow.

If you have any feedback on this leaflet, please use our leaflets feedback form which is available from either the TOFS office or our web site.

TOFS relies on money from membership fees, voluntary donations and other sources of charitable income to fund its activities.

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TOFS does not offer specific medical advice to parents. We work

only in a supportive role, offering emotional and practical support to meet the needs of parents and providing a source of information which complements that given by the specialist hospital.

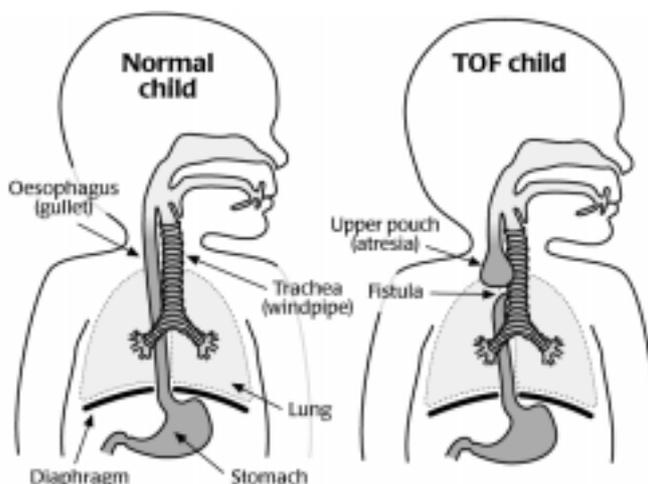
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"One of the nurses said to me "Don't worry – we get plenty of TOFs in here." Strange as it seems now, at that time I had never heard the term 'TOF,' I had just been told there was a pouch in Colin's oesophagus. So I thought she had said 'We get plenty of tofs in here', meaning that they treated a lot of rich kids... I thought this was a very odd thing to say, and replied 'But he's just an ordinary little boy' which she must have found strange. It wasn't until several days later that I realised what she had said."

"I was frightened to begin with as the child was so ill after birth. As I found out more about the condition I became less worried, but those first few months were terrifying."

Related leaflets from TOFS which you might like to read:

1. The news that your baby is a TOF
2. What causes TOF?
3. Surgery for TOF/OA
4. TOF: long term follow up
5. Conditions occurring with TOF/OA
6. VACTERL: an overview

These are all available from the TOFS web site (www.tofs.org.uk) or from TOFS office.

TOFS also publishes a book, 'The TOF Child,' which is suitable for both parents and medical professionals. Details are available from TOFS.

Diagnosis of TOF/OA

Oesophageal atresia may be suspected before birth, however with most TOF/OA babies there is no warning before birth that anything is wrong.

The main characteristic of oesophageal atresia in a newborn baby is the inability to swallow saliva – frothing at the mouth and 'blowing bubbles.' Fluid gaining entry to the trachea and lungs, either through a tracheo-oesophageal fistula or directly from the mouth down the trachea, may cause problems with breathing with the baby making exaggerated efforts to breathe and/or showing blueness of the lips and fingertips. Any air passing into the stomach from the trachea via a tracheo-oesophageal fistula (often as a result of the respiratory movements) may also distend the tummy.

If an oesophageal atresia is suspected, the passage of a tube down the oesophagus from the mouth will either confirm the diagnosis (if it is held up in the blind-ending sac) or exclude it (if the tube reaches the stomach).

Radiographs (X-rays) of the chest are essential to see where the tube is being held up and to examine the lungs and heart. Radiographs of the abdomen may also be taken; the presence of air in the stomach means that the lower oesophagus is attached to the trachea (i.e. a tracheo-oesophageal fistula).

Radiographs using barium to further examine the oesophagus are not usually necessary. Once the diagnosis has been confirmed, many surgeons will start the operation by examining the trachea and oesophagus using endoscopy (i.e. bronchoscopy and oesophagoscopy) to identify the length of the upper oesophagus and position of any fistulas.

The need for surgery

TOF/OA REQUIRES SURGERY;

- i) the two ends of the oesophagus must be joined together to allow the baby to swallow food such that the nutrients it contains can be digested and absorbed.
- ii) any connection(s) with the trachea must be closed off to prevent swallowed food/fluids passing from the oesophagus into the lungs – also to stop air passing from the trachea to the oesophagus and then into the stomach.

Other problems

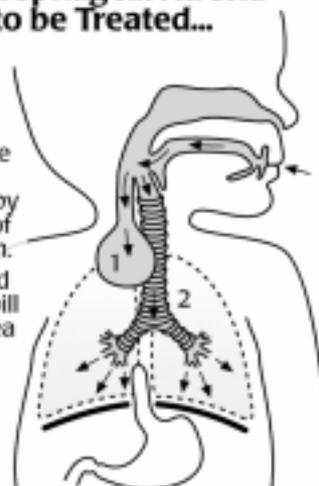
Other associated congenital problems may also be seen in the abdominal radiograph.

The commonest are gut problems either in the rectum (the last part of the large bowel) and/or in the duodenum (the part of the small bowel next to the stomach). These have all been successfully corrected even in very small babies. Rarely all three are seen in the same baby.

It is usual to examine the heart and kidneys using ultrasound scanning; defects in these important organs are common because of the associations between TOF/OA and two groups of abnormalities – VATER syndrome (more recently termed VACTERL) and CHARGE.

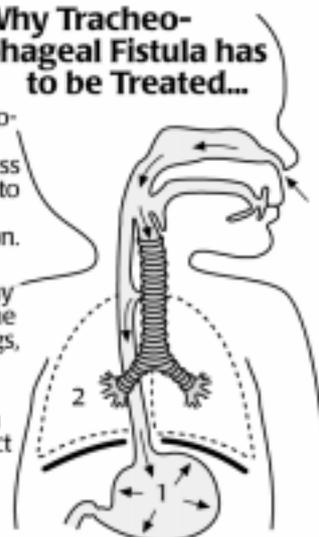
Why Oesophageal Atresia has to be Treated...

1. With an oesophageal atresia, food cannot enter the stomach and therefore the baby has no means of gaining nutrition.
2. Unswallowed fluid may also spill down the trachea into the lungs, causing pneumonia.



Why Tracheo-Oesophageal Fistula has to be Treated...

1. With a tracheo-oesophageal fistula, air can pass from the trachea to the stomach, causing distension.
 2. Food in the oesophagus may also pass into the trachea and lungs, causing pneumonia.
- Presence of an atresia will affect the relative likelihood of these events.



IF YOU'RE NOT ALREADY A MEMBER OF TOFS, WHY NOT JOIN US?
Information available from either TOFS office or the TOFS web site.

