Tracheomalacia and the TOF cough

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The trachea

In the common type of TOF/OA there is a blind ending upper oesophagus or foodpipe (the atresia) and a connection between the lower end of the oesophagus and the trachea or windpipe (the fistula). Other forms of TOF/OA may be slightly different, but the effects on the trachea at the site of the fistula are the same:

i) the supporting cartilage (gristle) framework, which keeps the airway open, is not fully formed.
ii) specialised lining cells (goblet cells and ciliated cells) are replaced by less specialised cells (squamous cells) that are less efficient at keeping the airway protected against infections.

In spite of these differences, in almost all cases the body recovers well so that there are no long term serious problems by the time the child has grown up.

What is tracheomalacia?

Any problem with the cartilage framework which normally supports the trachea will mean that the wall making up the airway is softer than it should be.

If there is a long section without cartilage, this can cause a 'floppiness' of the trachea, and this is called tracheomalacia.

The 'TOF cough'

Mucus is a sticky fluid which is found on the surfaces of the normal airway. Its main function is to trap dust particles in the air, before they travel deep into the lungs. The specialised lining cells of the airway have mechanisms both to produce this mucus, and then to transport it – together with the trapped dust particles – up the airway to the throat, where it is either coughed out or swallowed.

The mechanism is normally so efficient that most people hardly notice that they have mucus to clear and just automatically clear their throat from time to time.

In a TOF child, the gap in the specialised protective lining cells at the site of the fistula can make the clearance of mucus less efficient and/or the mucus drier than normal. The child may therefore have to cough quite hard in order to clear this dry and therefore extra-sticky mucus.

The 'TOF cough' results from the need to clear this dry mucus by coughing through a trachea with a slightly floppy section that makes the flow of air less smooth during the cough. Anything that increases mucus production will make the TOF cough worse, e.g. colds, liquids or food getting into the airways (aspiration), or asthma.

The noise of the TOF cough is quite characteristic and can be alarming to others. Most TOF families find their own ways of dealing with any comments which may arise, however it can often be useful to tell people who will be looking after a TOF child (friends, playgroups, schools) about the cough in advance. Letting them know that it is not distressing for the child (as is usually the case) and that it does not mean that the child is unwell in any way can prevent worry and embarrassment.

How important is tracheomalacia?

Most babies who have had a TOF repair do not have major difficulties coping with tracheomalacia and it becomes less and less significant as they get older.

There are however a small number who have problems, which can be serious.

When are problems worst?

The problems with the respiratory tract following TOF repair tend to be at their worst in the first two years.

This may be because the trachea of normal infants contains immature cartilage which is quite soft. The area of absent cartilage from the TOF trachea has a much greater effect when surrounded by a softer cartilage framework; when there has been more growth of the normal section of the trachea and the tracheal cartilage is firmer, the deficit is easier to cope with.

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 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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Related leaflets from TOFS which you might like to read:

1. What is TOF/OA?
2. Chest infections and 'wheeze'

These are both 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.

Severe tracheomalacia

The infant with severe tracheomalacia usually becomes symptomatic around the age of 4-6 months when there may be an exaggeration of the TOF cough, excessive wheezing, or cyanosis ('blue attacks') during feeding.

In extreme cases, the infant may experience acute life-threatening episodes often called 'near death episodes,' when the baby seems to be choking and unable to breathe. The trachea actually collapses, so that no air can pass through it.

Because these extreme situations are so rare, many doctors have never witnessed one of these episodes; the child is in any case often perfectly well by the time they get to hospital or are seen by a doctor.

The story of a baby going blue with hard crying may suggest 'breath holding attacks' - which are relatively common. However, these tend to occur in older toddlers who are either angry or very upset, and happen at the end of a big breath in or out. The 'near death episodes' usually happen in the midst of a normal crying spell, and happen near the start of either breathing in or out (depending on where the tracheomalacia is). Observers who can describe these kinds of detail about episodes can help greatly in making a correct diagnosis.

The attacks can be very distressing. Nonetheless, when the child passes out, their relaxed state helps to open up the airway, bringing recovery; gently pulling the tongue forward may help as a first aid measure. If the baby is unconscious and not breathing, blowing gently into the airway by mouth to nose-and-mouth respiration is the correct course of action.

What other things happen?

A soft trachea can also be squashed from the outside. For example, a narrowed oesophagus can cause a 'hold up' to food (an obstruction), so that the upper oesophagus fills up and stretches so much that it pushes against the neighbouring trachea. Without a strong enough cartilage framework to keep it open, the trachea can become closed off until the oesophagus empties again.

The same situation can arise if the lower oesophagus fills up with refluxed stomach content. The latter episodes can be hard to explain as they may not seem to be linked to any reason for a breathing problem.

Will my baby have problems?

Most TOF babies have some floppiness in the trachea, but the majority are not troubled by it and any problems improve with age. In assessing children, it is therefore not enough just to look for 'floppiness' – the oesophagus may actually be the origin of the symptoms.

Sometimes there may be more than one problem; in these cases it can be difficult to work out which is the most important, and then to decide on the best treatment.

Tests for tracheomalacia

RADIOGRAPHY

The diagnosis can be suspected on a lateral (side-on) radiograph of the neck taken either as the baby breathes in and out, which will show a collapsing trachea.

A barium meal study is often useful to check for any oesophageal malfunction.

BRONCHOSCOPY

Bronchoscopy (looking into the airway via an endoscope) is the most reliable method of reaching a diagnosis.

RESPIRATORY FUNCTION TESTS

Tests of the baby's breathing can measure how much work the baby has to do in breathing in and out, but cannot give information about the length of the trachea which is abnormal.

Treatment

Most TOF children do not require treatment for tracheomalacia. A few, however will require surgery, which aims to give the trachea extra support.

Tracheopexy and aortopexy are two such procedures; which is used depends on the individual baby. If the floppy part of the airway stretches down into the smaller airways (bronchi) then these procedures may not be so effective; in such cases, a tracheostomy tube may be inserted.

TOF children with severe tracheomalacia have a good chance of improving greatly with treatment and age.