

# FACTSHEET

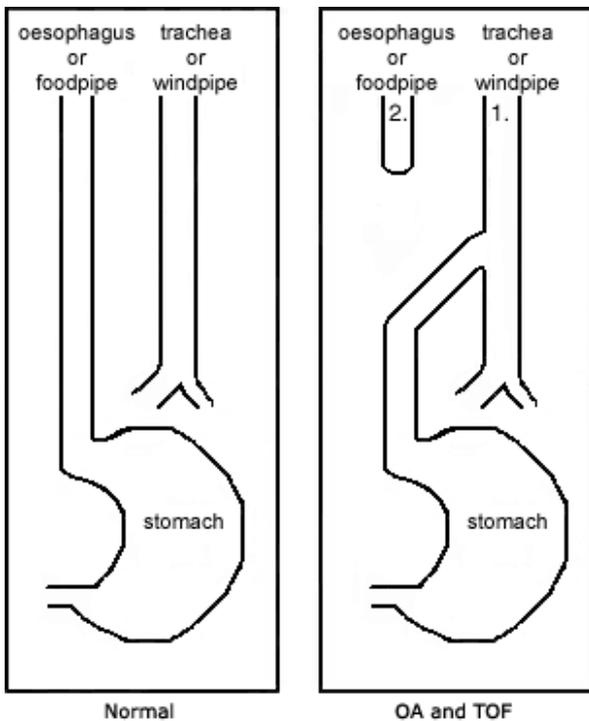


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## Oesophageal atresia and/or tracheo-oesophageal fistula

By now you will know that your baby was born with a condition called oesophageal atresia (OA). OA usually occurs with tracheo-oesophageal fistula (TOF), but not always. This fact sheet has been prepared to give you some basic information about the condition and to help you care for your child.

The **trachea** is the **wind pipe**, which passes air to the lungs when we breathe. The **oesophagus** is the **food pipe**, which carries food from the mouth to the stomach.



1. Trachea (Wind Pipe) connected to Oesophagus (Food Pipe) by Fistula – TOF
2. Atresia (“Blind” end) of oesophagus-OA

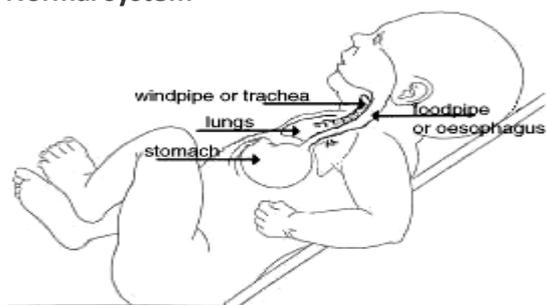
### What is oesophageal atresia (OA)?

In oesophageal atresia a section of the oesophagus (food pipe) has not formed properly and so the oesophagus is not one continuous tube but has two unconnected parts; an upper pouch and a lower segment. This means that saliva and food cannot pass from the mouth into the stomach as it becomes stuck in the upper blind-ending pouch (see diagram). Most babies will present at birth with an inability to swallow their secretions.

### What is tracheo-oesophageal fistula (TOF)?

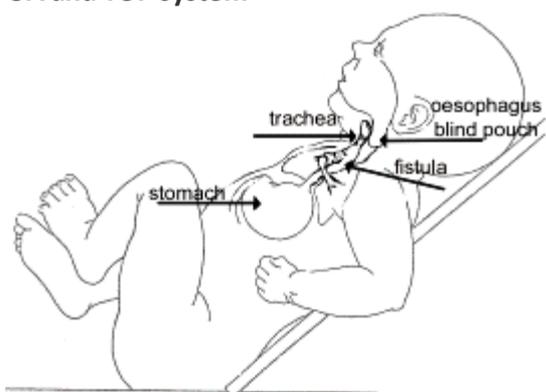
TOF is a rare condition. About 1 in 4,500 children are affected. If your baby is a reasonable size when born with OA/TOF, and if your baby is born without any other serious life threatening conditions, the outlook is extremely good. While your baby may have some difficulties in the early years of their life, it is important to remember that they will be able to lead fully normal lives.

## Normal system



Normal digestive system

## OA and TOF system



OA and TOF digestive system

## What is tracheo-oesophageal fistula (TOF)?

Most babies with OA will also be born with an abnormal connection or “fistula” between the food pipe (oesophagus) and the windpipe (trachea) (see diagram above). This causes air to pass from the windpipe to the stomach and stomach juices to reflux from the stomach to the lungs both which cause breathing difficulties and dusky episodes soon after birth.

There are 5 types of OA/TOF. Most babies born with TOF are of the common type, often referred to as Type C. This is where there is a connection between the distal part of the oesophagus and the trachea. Check with your baby's doctor which type of TOF your baby has.

## How are children with OA /TOF diagnosed?

Both OA and TOF are diagnosed soon after birth. The midwife may try to pass a tube (nasogastric tube or NG tube) through your baby's nose into their stomach, and find that it is not possible to do so. Sometimes, OA/TOF might be discovered in an ultrasound scan during pregnancy, particularly if the stomach is not very clear in the scan.

## What causes the condition?

No one really knows! What we do know is that the usual development does not occur at the normal time, very early in the pregnancy - sometime during the first three to six weeks.. Boys and girls are equally affected. It is believed that it is caused by a number of factors acting together, but just what they are we have yet to learn.

We do know that one of the contributing factors may be hereditary but this is not certain. It is highly unlikely to happen in following pregnancies or in your grandchildren. An important point to keep in mind is that you have not done anything wrong. The good news is that the majority of OA/TOF children grow up to be normal, active young people if they are born with no other major problems.

## How are OA and TOF repaired?

Until your baby has the operation to repair the OA/TOF, a tube called a “reple tube” is passed through the nose into the food pipe (oesophagus). This will drain off any saliva which is in the oesophagus, so that your baby does not choke. Your child will be nursed in the intensive care unit during this period, depending on their general condition.

For most TOF babies it is possible to repair the food pipe immediately with surgery. During the operation the connection between the windpipe and the food pipe (fistula) is closed off (ligated) and the two ends of the food pipe (oesophagus) are joined together. The join is called anastomosis.

There are two ways of carrying out the operation: using thoroscopic (keyhole) or open (thoracotomy) surgery. The surgeon will discuss the most appropriate method with you. The method used to repair the OA depends on the distance between the ends of the oesophagus. In most cases, the surgeons will be able to join the two ends of the oesophagus together to form a continuous passage from the mouth to the stomach.

In rare cases (around 5%) called “Long Gap OA” where the distance between the two ends of the oesophagus is too large for the surgeon to be able to join them straightaway, a different treatment is needed. One of the ways in which “Long Gap OA” is repaired is by a technique called the “Foker Technique”. The principle behind this technique is the fact that in a foetus inside the uterus, the normal oesophagus develops because of the tension placed on it by the growing tissues of the baby. This tension has to be created for children with OA to help lengthen the oesophagus. After birth, traction sutures are attached to the tiny oesophageal ends and the tension on these sutures is increased, bit by bit.

These traction sutures stimulate the upper and lower ends of the oesophagus to grow. At least two operations are needed in the “Foker Technique”. The first operation attaches traction sutures to both ends of the oesophagus. This tension is then increased on the sutures over the following days or weeks (depending on length of the gap), by your surgeon. Another operation then removes the sutures and joins the oesophageal ends together. In some cases while waiting for the two ends of the oesophagus to approximate, a special feeding tube or button (gastrostomy tube) is placed in the stomach of your child so milk can be given directly into the stomach. A small operation is needed when the gastrostomy tube is first put in place.

Another way to repair a “Long Gap OA” involves nursing your baby in hospital until the oesophagus (food pipe) grows enough to make it possible to be joined together. During this time a repleg tube is used. Also a special feeding tube (gastrostomy tube) is placed in the stomach so milk can be given directly into the stomach. A small operation is needed when the gastrostomy tube is first put in place.

For some children with “Long Gap OA”, the upper part of the food pipe (oesophagus) is brought out into an opening at the side of the neck. This is called an “oesophagostomy”. This opening is left for 1-2 years before another operation is done to join the food pipe together. The opening in the neck allows saliva to drain out onto a small pad so your baby does not choke. It is important to feed your baby with milk into the mouth so that they remember how to suck when the feeding tube is repaired/joined. This is called “sham” feeding. The milk will flow out the opening in the neck so your baby does not choke. A special feeding gastrostomy tube or button is also placed in the stomach so milk can be given directly into the stomach.

Once you are confident with the special care your baby needs, you can be discharged to go home until the next operation.

### Are there any risks?

All surgery carries a small risk of bleeding during or after the operation. Even if the gap between the ends of the oesophagus is quite small, it can still be difficult to join together. After the operation, the join may leak or it may narrow over time, but these can both be treated. Your child will be reviewed regularly for a long time after the operation.

Every anaesthetic carries a risk of complications, but this is very small. Your child’s anaesthetist is a very experienced doctor who is trained to deal with any

complications. For more information read our factsheet on [Anaesthesia and risk in infants](#).

### What happens after surgery?

Your baby will go to the intensive care unit (ICU) to recover. For a while after the operation, your baby will need help with breathing and will be connected to a ventilator. All babies are closely monitored after the operation. Your baby will be connected to monitors to check breathing, heart rate and oxygen levels. Pain relief will also be given through a ‘drip’.

Your baby will be transferred to a hospital ward when they no longer need intensive care. While your baby’s intestines recover and start to work, they will have an intravenous drip of fluids. Feeding usually starts a few days after the operation, with breast or bottled milk given through a naso-gastric tube (tube passed through a nostril, down the oesophagus and into the stomach). As your baby recovers, you will be able to breast or bottle feed.

Some babies’ intestines take a little longer to recover, so need to be fed through a tube into the veins (total parenteral nutrition or TPN). Naso-gastric feeding is then tried when your baby’s intestines start to show signs of recovery.

Once your baby is feeding properly and gaining weight, you will be discharged home.

### What is the outlook for children with OA/TOF?

If your OA and TOF baby has no other associated problems, the outlook is excellent. Most children grow up to live normal lives. The outlook for children with OA and TOF who have other difficulties, such as extreme prematurity and heart conditions will vary depending on how severe these other problems are.

There are a few problems which may happen after surgery that you should know about. They happen most often in the first few years after the operation and improve as your child grows older.

### Care of your TOF baby

Caring for your baby does not begin when you take your baby home. You can share in your baby’s care while still in hospital. Don’t be afraid to ask questions, even the same question over and over again. It is often hard to fully understand things when you are worried or anxious. People caring for babies and children in hospital

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understand that all parents are worried and anxious when their child is not well and needing medical attention. Confused feelings are normal at this early time.

Many parents speak of their deep disappointment that they were unable to take their baby home from the maternity hospital in the usual way. Sometimes people can feel confused and angry. Most parents feel frightened to see their baby connected up to tubes and machines. Some parents may not want to see their baby until they are well on the road to recovery and free of equipment. If that is the way you feel, remember that these feelings are perfectly normal. The sooner parents are involved with their baby the better. The more you know your child and care for them in hospital, the easier it will be when you take your baby home.

Parents are the real experts in the care of their child. Every TOF child, like every other child, is different. Though you need the help of other experts for a time, you are the most important people in the team caring for your child. A multidisciplinary team of experts will help you look after your baby by monitoring their progress during regular clinic visits

### Problems you may have in the early years

It is important to know that your child may experience problems in the early years. Problems include swallowing difficulties and a higher number of childhood coughs and colds. It is important to keep in close touch with your baby's doctor about anything that you are worried about. The following gives you an idea of problems that can occur:

#### Swallowing problems

Many children with OA/TOF experience some form of feeding difficulty, however the amount of difficulty varies between children. Feeding will improve as your child gets older.

When you take your baby home you will be given a feeding program suitable for your baby and their specific needs. You should start feeding orally as soon as your child is allowed. Many children can breastfeed. As many children with OA/TOF suffer from reflux, they often cope better with smaller more regular feeds.

Solids should be introduced around 6 months, as they would with any other baby, unless you are told otherwise by your doctor.

As your baby grows you will add solids and increased varieties of foods. Often children with OA/TOF have difficulty progressing with their food textures from very smooth runny purees to more solid or lumpy foods. OA/TOF children may take longer than usual with the

introduction of solids into their diet. In order to give your baby a good start to feeding, your child needs to stay on smooth purees and "mushy foods" for a longer period of time to reduce the risk of choking. Feeding should be done under close supervision to reduce the risk of choking. A blender or small mincer is really helpful when preparing "mushy" food.

We recommend that you give your baby a wide range of food. Frequent and early exposure to a wide range of tastes has been shown to play a major role in what food children will eat.

Common problem foods for children with OA/TOF include:

- meat (unless pureed or minced)
- bread/doughy foods
- hard raw vegetables (carrots, apples)
- foods with skins (grapes, tomatoes).

When you introduce new foods, always blend, mince or chop them finely. When your child gets used to eating the new food, gradually make the sizes bigger. Also let your child watch you cut it up. Give them a nibble while you are doing it. As they grow, they won't be afraid to eat new things if you have been introducing them to food gradually.

Some children complain of problems with swallowing and need to have a drink with all food. This is often caused by the oesophagus not being coordinated. Try to make sure that meal times are quiet and peaceful (this is not always easy with a young, energetic family). When children are in a hurry or excited, swallowing problems are more likely to occur. During social occasions keep an eye on your child. Things like nuts and potato crisps left at the child's level can be a significant problem.

The Speech Pathologist and Dietician can give more individualised advice on feeding. They can help you with advice on weaning and suggest the types of foods that are most suitable for your child as they grow older. The Speech Pathologist can help with problems with swallowing including troubles with textures and also help with exercises to help with chewing and swallowing at meal times. The Dietician can help with ensuring your child's intake is meeting their nutritional needs and recommend higher energy options or additions if there are concerns with growth.

#### Stricture

Sometimes the oesophagus becomes narrow at the site of the join (anastomosis) - this is called a **stricture**. Babies with a stricture have problems swallowing and may have increased vomiting or may choke or gag with feeds. Stricture is treated with dilation (stretching the

narrowed food pipe). This procedure is done under a general anaesthetic either in the radiology department or with the help of an endoscope in the operating theatre. The purpose is to widen the narrowed oesophagus to help with feeding.

### **Reflux**

Children with OA/TOF often develop a problem with gastro-oesophageal reflux disease. This is where the contents of the stomach flow back up the oesophagus causing pain and irritation. This may cause a baby to vomit, but not always. As acid reflux can irritate the oesophagus and increase the chances of stricture, your child will be started on an anti-reflux medication after the initial operation to reduce the risk of this developing. Your child will need regular surveillance endoscopies to monitor their reflux.

### **Tracheomalacia**

In addition to a possible abnormal connection (fistula) between your baby's windpipe (trachea) and food pipe (oesophagus), the windpipe in children with OA/TOF is unduly floppy due to a lack of properly developed cartilage to support its walls. This is known as "tracheomalacia" (or soft trachea) and may cause noisy breathing at rest or with activity, a loud "barking" cough, reduced ability to bring up phlegm/sputum and breathing difficulties.

Many, but not all children with OA/TOF suffer from what has become known as the "TOF" cough. This is a harsh, brassy, "barking" cough, which sounds as if the child is very sick. Remember, this does not mean that your child is sick and often family and friends need to be reassured that your child is fine.

Sometimes due to their floppy windpipe, your child might experience a sudden breath-holding episode or "blue spell". This usually happens during eating, if a piece of food becomes stuck in the food pipe. The lump of food puts pressure on the windpipe significantly narrowing it so that air cannot move in and out. As a result, breathing becomes extremely difficult and your child may change colour, becoming a dusky red or even blue. Please contact your doctor immediately if this happens.

### **Respiratory problems**

Children with OA/TOF are more likely to experience prolonged colds and even pneumonia in the first few years of life. This is because they cannot clear their secretions effectively due to their floppy windpipe. Although this is likely to improve by the early school years, chest physiotherapy and measures to increase mucus clearance can help. Physiotherapy exercises can be taught at the hospital by a trained physiotherapist. These can be done at home quite easily and help when

your baby has a cold. We would recommend that your child also have the Influenza Vaccine every year to prevent serious pneumonia.

In time most respiratory symptoms improve. It is important though that your child be regularly reviewed throughout childhood so that any ongoing problems can be identified and managed so that your child's wellbeing is maintained.

### **Acknowledgments**

Some of the information for this fact sheet was gathered in a long-term study of TOF children undertaken at The Children's Hospital at Westmead in conjunction with the [Children's Medical Research Institute](#). Sixty-two children over the age of ten who were successfully operated on when they were babies were interviewed along with their parents. We are very grateful to these young people and their parents for giving us the benefit of their experience to help others in similar situations.

### **Remember:**

- Most babies with TOF are able to lead a normal life by the time they get to school.
- As your baby grows you will need to add solids and increased varieties of foods.
- There really is no reason to delay the introduction of solids (always in consultation with your baby's doctor).
- TOF children may take longer than usual with the introduction of solids into their diet.