

# Long-gap Oesophageal Atresia

A guide for parents



oara  
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Nate (6 weeks old)



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Research Association

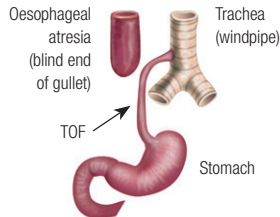
# Are there different types of OA?

There are four main types of oesophageal atresia with or without TOF. These include the following:

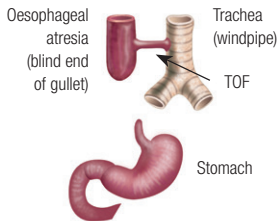
## Pure OA (no TOF):



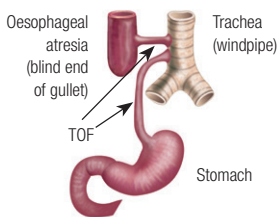
## OA & distal TOF:



## OA with proximal TOF:



## OA with double TOF:



## What is “long-gap” oesophageal atresia?

As you can see from the diagram, babies with pure OA or OA with a proximal TOF usually have a long gap between the two ends of their oesophagus. This means that immediate surgery to repair the oesophagus is not usually possible. Most babies with “long-gap” OA have pure OA. Occasionally a proximal TOF may also be present.



**Pure OA**

or



**OA with proximal TOF**

(abnormal connection  
between upper  
oesophagus and trachea)

Babies with OA and distal TOF may also have a large gap between the two ends of their oesophagus; these babies will be treated the same as those with a “long-gap” OA.

## What is oesophageal atresia?

*Oesophageal atresia (OA) is a congenital obstruction in the oesophagus in newborn babies. In this condition the oesophagus ends in a blind upper pouch. In babies born with oesophageal atresia the oesophagus, also known as the gullet (or foodpipe), is not connected to the stomach. As a result of this obstruction, food and saliva can't pass into the stomach. In most cases, the baby with oesophageal atresia also has a tracheo-oesophageal fistula (TOF).*

## What is a tracheo-oesophageal fistula?

*Tracheo-oesophageal fistula is the condition where a section of the baby's trachea (windpipe) is abnormally connected by a small tube (fistula) to the baby's oesophagus.*

## How does TOF/OA occur?

*The oesophagus and trachea actually form from the one tube. OA and TOF are believed to occur when the oesophagus and trachea fail to differentiate and separate properly. Research is continuing into the cause of TOF/OA; at this stage the cause is not known.*

## Are there other abnormalities associated with TOF/OA?

*Approximately half the babies born with TOF/OA have other abnormalities. This is why a number of investigations are undertaken to see if your baby has any additional problems. Some babies are born with abnormalities of the spine, heart, kidneys, anus and/or radial bone (a bone in the lower arm). If a baby has three or more of these problems, it has what is known as "VACTERL Association". VACTERL is an acronym where the letters stand for vertebral, anal, cardiac, tracheal (TOF), esophagus (oesophageal atresia), renal and limb.*

## How common is TOF/OA?

*TOF/OA occurs once in every 3500 live births.*

## What special care does my baby with long-gap OA require?

Due to the obstruction in your baby's oesophagus, your baby will not be fed initially, but will receive fluid containing water and sugar through an intravenous line. It is not possible to feed your baby orally as the milk and saliva will pool in the oesophageal pouch and may spill over into your baby's windpipe (trachea) and lungs resulting in pneumonia.

For this reason your baby will require their oesophageal pouch to be kept clear of saliva and secretions that can spill into their lungs. The oesophageal pouch is kept clear of saliva by suctioning your baby intermittently or with a Replogle tube. Intermittent suction involves inserting a suction catheter into your baby's oesophageal pouch every 10 – 30 minutes and applying suction to remove the saliva. A Replogle tube is a soft plastic tube that sits near the bottom of the oesophageal pouch and is connected to continuous low pressure suction to keep your baby's oesophageal pouch clear of secretions. The Replogle tube is flushed frequently to ensure it is not blocked with secretions. Suction of the upper oesophageal pouch continues until your baby is taken to theatre for surgical repair of their oesophageal atresia.

Your baby will also continue to have their heart rate, breathing, oxygen saturation levels and temperature monitored.

## How do the doctors know that my baby has a long-gap OA?

On admission your baby will have an abdominal and chest X-ray performed. If there is no air in your baby's stomach on X-ray this usually means that there is no distal TOF i.e. no connection between their windpipe and the bottom part of their oesophagus and therefore no way for air to get from your baby's mouth to their stomach. This usually means that they either have a pure OA or an OA with a proximal TOF and these two types of OA generally have a very large gap between the two ends of the oesophagus.

## My baby also has a TOF, when will this be repaired?

If your baby has a TOF, this will be repaired within the first few days of life. The TOF is divided and the tracheal end of the fistula is repaired with sutures. The oesophageal end of the TOF will be oversewn and closed. The site of the TOF will determine what incision the surgeons will make to reach the TOF. If your baby has a proximal TOF the surgeon will usually repair the TOF through a small incision in your baby's neck. If there is a distal TOF present, a thoracotomy will usually be performed; this involves an incision usually on the right side of your baby's chest toward their back.

## How will my baby be fed?

In most cases, your baby can't be fed orally until their oesophagus has been repaired.

An operation will usually be performed in the first few days of life so that your baby can be fed. A tube called a gastrostomy will be inserted into your baby's stomach through their abdomen, so that milk feeds can be given. Milk feeds are slowly introduced through the gastrostomy tube a few days after the gastrostomy tube is inserted. The feeds will be increased in volume slowly until your baby is receiving full milk feeds and will no longer need intravenous fluids.

During gastrostomy feeding your baby's dummy can be dipped in breast milk or formula to enable them to experience the taste of milk and encourage sucking on the dummy whilst their stomach is filling with their feed.

Occasionally, in some cases involving a delayed repair of the OA, sham feeds may be offered to your baby before the OA is repaired. This involves breast or bottle feeding baby a few times a day, with the milk removed continuously from the oesophageal pouch with a Replogle tube connected to suction. The milk removed from the oesophageal pouch is then given to your baby via their gastrostomy tube. Sham feeding is not offered in all children's hospitals and is not appropriate for all babies with long-gap OA. You will need to discuss sham feeding with your baby's surgeon to see if this is possible for your baby.

Following surgical repair of your baby's oesophagus your baby should be able to breast feed. If you are planning on breastfeeding it is important to begin expressing breast milk regularly after birth. A lactation consultant is available for support and advice.

## When will surgery to repair the oesophageal atresia be done?

The surgeon will decide on when is the best time to repair the oesophageal atresia. Your baby's surgeon will be able to talk to you about this and the timing and type of surgery to repair the oesophageal atresia.

## Does my baby require specific care before surgical repair of their oesophagus?

Your baby needs to stay in hospital until their oesophagus is repaired by surgery. This is because the oesophageal pouch needs to be continually cleared of secretions to prevent pneumonia. We encourage parents to be as involved as much as possible in their baby's care. Parents are encouraged to learn to give their baby feeds through the gastrostomy tube and to learn how to change the gastrostomy dressings. In most cases, your baby will be discharged home with their gastrostomy tube in place, so it is important that you know how to look after it.

## Surgical repair of the oesophageal atresia

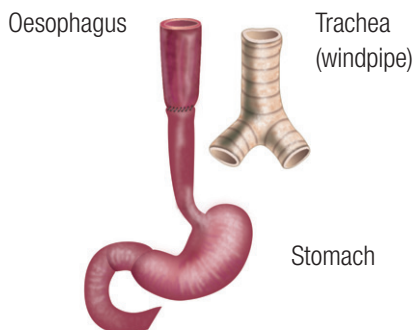
There are different types of surgery available to repair the oesophageal atresia including:

- Primary join (anastomosis) of the oesophageal ends.
- Foker procedure for oesophageal lengthening (this involves sutures placed in both ends of the oesophageal pouches to pull the ends closer together so that the oesophageal ends can be joined).
- Oesophageal replacement procedure. (This may occasionally be necessary in some cases, if the gap between the two ends of the oesophagus is still too large to enable repair of the oesophagus).

In most cases the surgeon is able to do a delayed surgical repair involving a primary join (anastomosis) of the two ends of the oesophagus. The surgical repair of the OA is not usually done immediately, as the ends are usually too far apart. This surgery is usually delayed for a number of weeks until the ends of the oesophagus are closer together to enable the repair of the OA to be done. Surgery to repair the OA is usually done via a thoracotomy which involves an incision usually made on the right side of the chest, under your baby's arm, towards their back. The upper oesophageal pouch and lower oesophagus may be mobilised to enable the surgeons to get the two ends of the oesophagus together with minimal tension on the anastomotic join site. The upper oesophageal pouch is then opened and joined to the lower oesophagus.

Your baby's surgeon will discuss with you the type of surgery that they recommend for your baby.

### Repaired oesophageal atresia:



## What special care will my baby receive following repair of their oesophageal atresia?

After surgery your baby will return to the neonatal intensive care unit for ongoing care. Your baby will come back from theatre with a breathing tube (an endotracheal tube) in their windpipe (trachea) and will be attached to a ventilator to assist with breathing. Your baby will receive analgesia to relieve the pain from the operation and may remain on the ventilator for a number of days after surgery. The neonatal doctors will decide when to take your baby off the ventilator. Your baby will continue to have their heart rate, breathing, oxygen saturation levels, blood pressure and temperature monitored and will be assessed regularly for any signs of pain.

The doctors and nurses will continue to monitor your baby after surgery for any signs that the join between the two ends of the oesophagus has come apart (this is known as an anastomotic leak). If this happens the surgeons usually wait for the join in the oesophagus to heal by itself without further surgery. If your baby develops an anastomotic leak a chest drain will usually be inserted to drain saliva from the chest, antibiotics will be given and your baby will continue to receive an intravenous nutrition solution. It may take a number of weeks for the leak to heal.

## When can my baby start feeding?

Your baby will continue to receive intravenous fluids until milk feeds can begin. The surgeon will decide when this will happen. Some surgeons prefer to do a contrast study of your baby's oesophagus to ensure that there are no problems with the oesophageal join before they allow your baby to start feeding. This contrast study usually involves your baby drinking some contrast solution to outline the oesophagus in an X-ray to see if the anastomosis has healed properly.

Initially the feeds may be given through the gastrostomy tube and then oral feeds will be commenced. You will then be able to give your baby breast feeds or bottle feeds depending on how you have decided to feed your baby.

Sometimes it may take many weeks or months for your baby to be able to take all feeds orally.

## When can my baby be discharged home from hospital?

Your baby can be discharged home from hospital following repair of their OA when they are taking some of their feeds orally, with the rest via their gastrostomy tube and they are gaining weight.

On discharge babies are usually having their feeds every three to four hours. It is important to continue to encourage your baby to feed orally after discharge. These babies vary with how long it takes them to take all of their feeds orally.

Your baby's gastrostomy tube will be removed when the surgeon is happy that the gastrostomy tube is no longer needed. For this to happen your baby needs to be taking all feeds orally and to be gaining weight.

## What sort of problems may happen once the oesophageal atresia has been repaired?

Most babies with OA grow up to live perfectly healthy, normal lives and may be no different from any other child. Whilst some children are fortunate not to have any further complications, others may have some problems. (Please refer to the brochure Repaired Long-Gap Oesophageal Atresia – A guide for parents).





Larni (3 Months old)



Riley (6 Months old)

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## Is there a support group for parents of babies with TOF/OA?

OARA (Oesophageal Atresia Research Association) is a support group for parents and families of babies/children with TOF/OA. A member of OARA may be available to visit you while your baby is in hospital.

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