Guideline

Stabilisation and transfer of an infant with oesophageal atresia +/- tracheo-oesophageal fistula

1 Scope

For use within the Paediatric and Neonatal Decision Support and Retrieval service (PaNDR) for the East of England.

2 Purpose

To provide safe, efficient and practical guidance for the management of neonates with oesophageal atresia +/- tracheo-oesophageal fistula during stabilisation and transfer.

3 Abbreviations

OA	oesophageal atresia
TOF	tracheo-oesophageal fistula
NGT	naso-gastric tube
HFNC	high flow nasal cannula

4 Introduction

Oesophageal atresia (OA) is a rare birth defect in which the oesophagus (the tube that connects the oral cavity to the stomach) does not develop normally (within 4-8 weeks of conception). In infants with OA, the oesophagus is usually separated into two parts, an upper and lower segment. These two segments do not connect. One or both segments (usually the upper) end in a blind pouch. Consequently, the normal passage between the mouth and stomach does not exist.

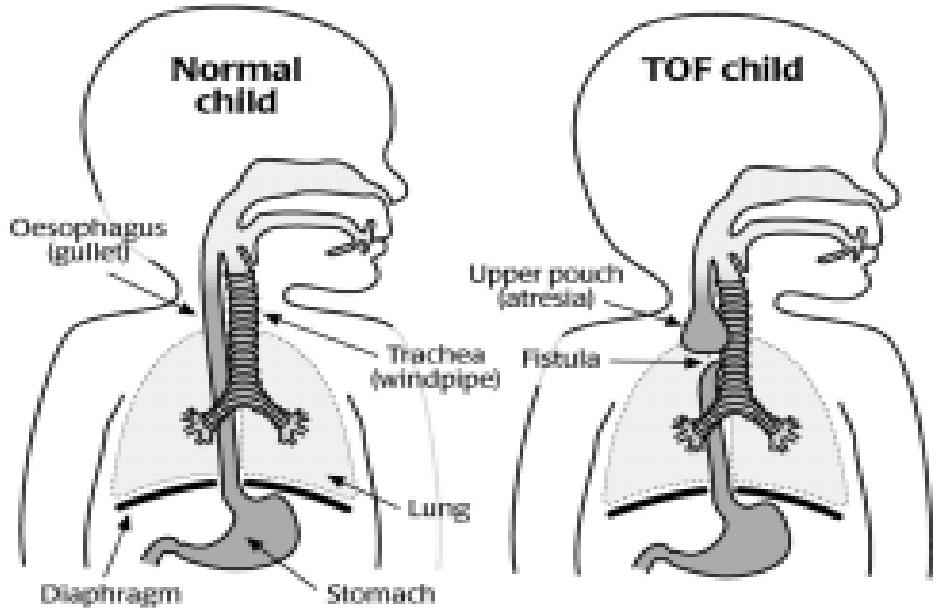
Almost 90% of infants born with OA also have a tracheo-oesophageal fistula (TOF), in which the oesophagus and the trachea (windpipe) are abnormally connected, allowing fluids from the oesophagus to get into the airways and interfere with breathing. A small number of infants have an isolated TOF, without OA.

It is estimated to affect 1:3000-5000 live births, with an equal rate in males and females.

OA +/- TOF is a life-threatening condition, however the majority of affected infants will recover fully with prompt recognition of the defect and appropriate surgical management.

The exact underlying causes of OA +/- TOF are not fully understood. OA +/- TOF can occur as isolated findings, be associated with other birth defects, or occur as part of a larger syndrome.

Figure 1: Comparison of the normal anatomy vs that of a child with OA + TOF (Type C)



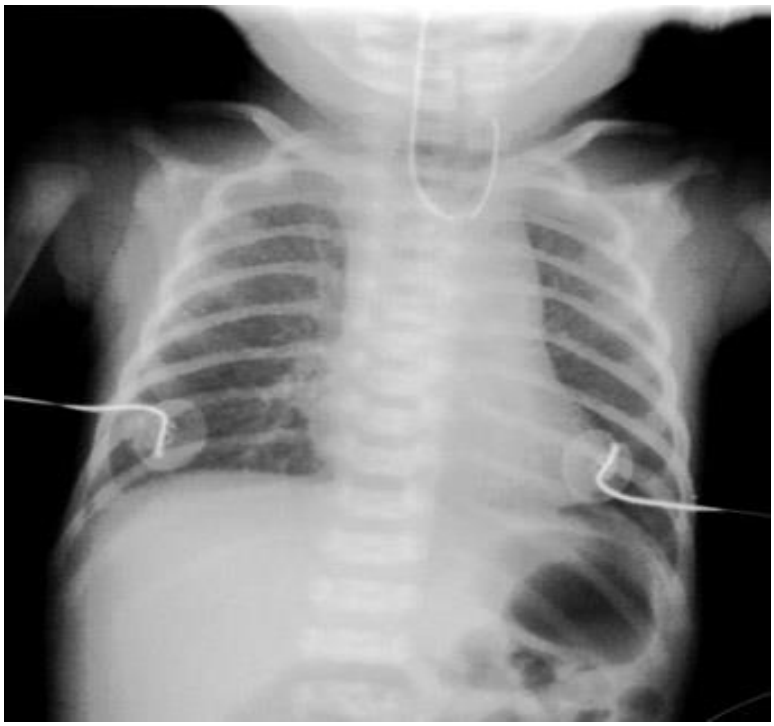
5 Diagnosis

Diagnosis of OA may be suspected before birth because of the presence of a small or absent stomach bubble on a routine ultrasound examination performed after the 18th week of pregnancy. The presence on ultrasound of excessive amounts of amniotic fluid (polyhydramnios) raises further suspicion of OA. Polyhydramnios alone is a poor indicator of OA however, as it has numerous, varied causes.

If not diagnosed antenatally, OA is usually detected shortly after birth, either by copious bubbly oropharyngeal secretions (which are not able to be swallowed) or when feeding is attempted and the infant coughs, chokes, and becomes cyanosed. As soon as the diagnosis is suspected, a health care professional should attempt to pass a large bore (size 8F or ideally size 10F) nasogastric tube through the mouth or nose into the stomach. The feeding tube will not pass

all the way to the stomach (beyond 10-12 cm) in a baby with OA and may curl back up into the oropharynx.

Figure 2: X-ray chest and abdomen with NG tube in situ:

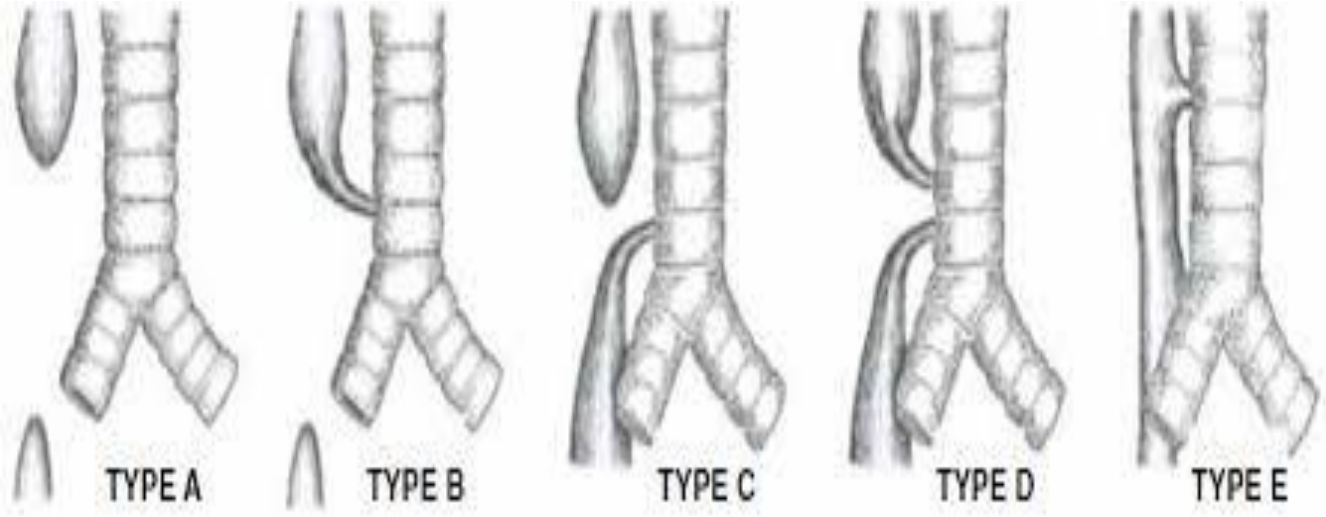


- The nasogastric tube is seen coiled in the upper oesophageal pouch
- The presence of air in the abdomen indicates the presence of a distal tracheo-oesophageal fistula (a gasless abdomen would be suggestive of OA **without** TOF)
- For distal pouches or H-type fistulae, the diagnosis can be more difficult.

6 Clinical features

- Bluish coloration to the skin (cyanosis) with attempted feeding
- Coughing, gagging, and choking with attempted feeding
- Drooling
- Inability to advance NGT beyond 10-12 cm
- Poor feeding

7 Types



7.1 Type A

OA is present but there is no TOF. The oesophagus is split in two with both the upper and lower segments ending in blind pouches. This is sometimes referred to as pure oesophageal atresia and accounts for approximately 8% of cases.

7.2 Type B

The lower segment of the oesophagus ends in a blind pouch and the upper segment is connected to the trachea via a TOF. This type of OA + TOF is extremely rare, accounting for only 2% of cases.

7.3 Type C (the most common type)

The upper segment of the oesophagus ends in a blind pouch and the lower segment is connected to the trachea via a TOF. This is the most common type of OA + TOF occurring in approximately 85% of cases.

7.4 Type D

A TOF is present connecting both the upper and lower segments of the oesophagus to the trachea. This is the rarest type, accounting for less than 1% of cases.

7.5 Type E

The oesophagus is not split in two and connects normally to the stomach but a TOF is present connecting the oesophagus and the trachea. This is also known as an H-type fistula and accounts for 4% of cases.

8 Associated conditions

50% of infants with OA +/- TOF also have other birth defects. Associations include:

- Atresias elsewhere in the gastrointestinal tract (duodenal, jejunal, colonic, anal)
- VACTERL association: Vertebral, anal, cardiac, tracheal, esophageal, renal and limb (skeletal) anomalies.
- Genetic syndromes such as:
  - CHARGE syndrome (Coloboma of the eye, congenital Heart disease, Atresia of nasal choanae, Retardation of growth and development, Genital anomalies, Ear anomalies)
  - Feingold syndrome
  - Anophthalmia-esophageal-genital (AEC) syndrome
  - Pallister-Hall syndrome
  - Fanconi anemia
  - Chromosome 22q 11 deletion syndrome
  - Trisomies 13, 18 and 21 (Down syndrome)

9 Management during stabilisation and transfer

- Keep the oropharynx/ upper pouch clear of secretions to prevent risk of aspiration of fluids into the lungs by frequent suctioning.
- Pass a Replogle tube (see PaNDR guideline insertion of a Replogle tube) place on low suction (5kPa) flushing with 0.5ml saline every 15 minutes, or more frequently as required. The syringe should be removed between flushes so that 0.9% sodium chloride cannot be inadvertently sucked into the tube if the Replogle tube blocks.
- If not possible to use low suction, pass a size 8-10 Fr NGT and aspirate every 5-10 minutes, or more frequently as required.
- Prolonged mask ventilation should be avoided as it may lead to upper pouch distension and gastric distension if a lower fistula is present. The gastric distension will be very difficult to decompress (in the presence of OA) and can cause significant respiratory compromise.
- Infants with suspected OA and respiratory distress may deteriorate rapidly and require urgent referral for transfer to a surgical centre.

- CPAP or HFNC should be used with caution in OA + TOF due to the risk of problematic gastric distension. Multiple infants with the condition have now been successfully moved on HFNC by the PaNDR team, so these modes may be considered after careful risk assessment and discussion with the PaNDR consultant.
- If the baby is spontaneously breathing with good oxygenation, intubation should be avoided.
- If intubation is indicated for respiratory distress, it may be beneficial to position the ET tube close to the carina, beyond the likely position of a fistula to avoid gas flow through a TOF.
- If the child requires ventilation low pressures should be employed to prevent gastric distension (via preferential air flow through the fistula). Gastric distension can lead to diaphragmatic splinting. In rare cases this can lead to gastric perforation.
- Examine for imperforate anus as this may worsen gastric distension.
- Keep nil by mouth and start IV fluids.
- Nurse infant in whatever position minimises respiratory distress (supine, prone or side). A 'head up' position (approximately 30–60 degrees) may be beneficial to encourage secretions to collect at the bottom of the upper pouch, making them easier to aspirate/ drain.
- Consider antibiotics if any evidence of aspiration pneumonia.
- Examine to rule out any other anomalies. The baby will need cardiac and renal scans and genetic testing by the local unit on a non-urgent basis.

10 Sources of further information

The following support group may be useful for parents:
TOFS
St George's Centre
91 Victoria Road
Netherfield
Nottingham NG4 2NN
Tel: 0115 961 3092
Email: info@tofs.org.uk, Website: www.tofs.org.uk

11 Monitoring compliance with and the effectiveness of this document

The effectiveness of the document will be monitored by review of any reported incidents via the lead nurse for risk. These incidents will be shared with the team and consideration given to adjusting the guideline if concerns are identified.

12 References

- https://www.rarediseases.org/rare-disease-information/rare-diseases/byID/930/viewFullReport
- http://www.nlm.nih.gov/medlineplus/ency/article/000961.htm
- http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=EN&Expert=1199
- Esophageal atresia pictures

13 Associated documents

- PaNDR guideline Insertion of a Replogle Tube

  https://img1.wsimg.com/blobby/go/37474867-8297-4fbd-8acf-ee0d108337d6/downloads/Insertion%20of%20a%20Replogle%20tube.pdf?ver=1670411883640

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Document management

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