NTS Clinical Guideline

Title:	Oesophageal atresia and Tracheoesophageal fistula		
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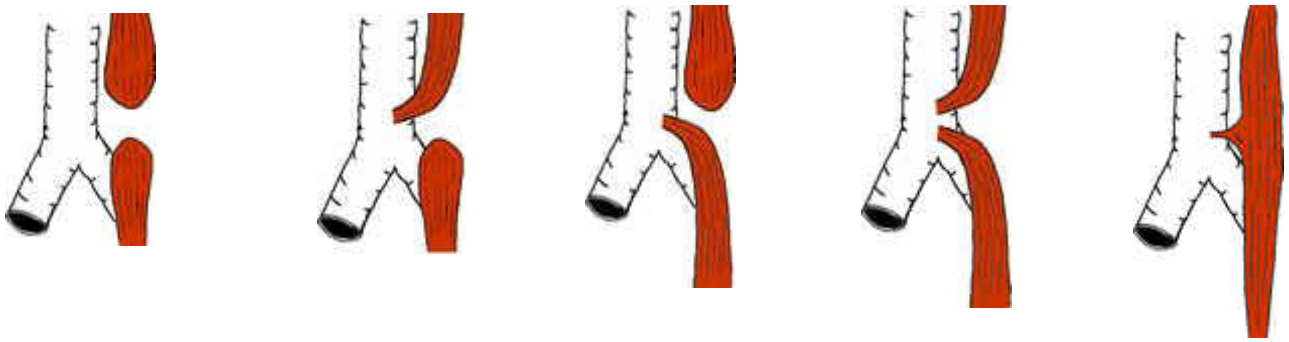
Description

Oesophagus and trachea develop from foregut around the 4th week of foetal development. Incomplete formation of oesophagus and failure in the complete separation of oesophagus and trachea results in oesophageal atresia and its variants with or without fistula between them. It can occur as an isolated anomaly or can be associated with other malformations.

Incidence approximately 1:2000 to 4000 livebirths.

High likelihood of premature delivery because of the high association with polyhydramnios.

Types



A **B** **C** **D** **E**

A Oesophageal atresia with no fistula and blind pouches 7-10%
B Proximal fistula and distal blind pouch <1%
C Oesophageal atresia with a distal fistula 85-87%
D Proximal and distal fistulae <1%
E 'H' type tracheo-oesophageal fistula 4%

Associations

- Atresias elsewhere in gastrointestinal tract, imperforate anus
- As part of VACTERL association: Vertebral, Anal, Cardiac, Tracheal, Esophageal, Renal and Limb (skeletal) anomalies. (Shaw-Smith, 2006)
- Can be associated with CHARGE syndrome: Coloboma of the eye, congenital Heart disease, Atresia of nasal choanae, Retardation of growth and development, Genital anomalies, Ear anomalies.

Presentation

Antenatally

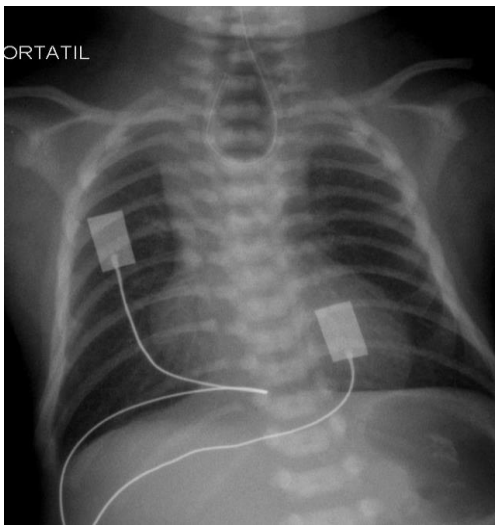
- Excess amniotic liquor volume / polyhydramnios, due to obstruction
- Small / absent stomach on antenatal ultrasound.

Postnatally

- Baby may have **excessive secretions** which pool in the proximal oesophageal pouch.
- **Respiratory distress** on feeding
  - The baby will choke, cough, or regurgitate unchanged milk
- **Inability to advance nasogastric tube** beyond 8-10 cm
- **Abdominal distension** is common if there is a distal fistula, as air moves through the fistula into the stomach. This may be exacerbated by positive pressure ventilation and cause further respiratory deterioration.

X-ray chest and whole abdomen with NG tube / Replogle 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 fistula. Gasless abdomen indicates pure oesophageal atresia.
- For distal pouches or H-type fistulae, the diagnosis can be more difficult.



Oesophageal atresia with Tracheooesophageal fistula

Acute Management

- **AIRWAY**
  These babies can be unstable – airway management is the main key
  Nurse supine in head up position, as high as restraints allow.
  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 relevant guideline), place on low suction (5-10kPa) flushing with 0.5ml saline every 15 minutes.
  - If not possible to use low suction, pass a size 10Fr tube and aspirate every 5-10 minutes.

  If intubation is indicated for respiratory distress aim for a normally positioned tube T1-T2. Be mindful of the inflation of the stomach with a distal fistula.

- **BREATHING**
  If the baby is spontaneously breathing with good oxygenation, intubation should be avoided.
  Mask ventilation should be avoided – leads to upper pouch distension and gastric distension if a lower fistula is present, leading to respiratory compromise.
  If respiratory support is needed, avoid non-invasive ventilation – intubation should be first line.

- **CIRCULATION**
  Nothing specific.

- **FEEDS AND FLUIDS**
  Nil by mouth, on IV fluids.

- **OTHER**
  Antibiotics if any evidence of aspiration pneumonia. Aspiration pneumonitis has accounted for up to 50% of the perioperative morbidity and mortality in this population (Taneja B., 2014)
  Examine for imperforate anus as this may worsen gastric distention.
  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.
  Intubated OA/TOF is a time critical transfer.

References

Shaw-Smith C. Oesophageal atresia, tracheo-oesophageal fistula, and the VACTERL association: review of genetics and epidemiology. J Med Genet. 2006 Jul;43(7):545-54. doi: 10.1136/jmg.2005.038158. Epub 2005 Nov 18. PMID: 16299066; PMCID: PMC2564549.

Taneja B, Saxena KN. Endotracheal intubation in a neonate with esophageal atresia and tracheo-esophageal fistula: pitfalls and techniques. J Neonatal Surg. 2014 Apr 1;3(2):18. PMID: 26023489; PMCID: PMC4420322.