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 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.
- 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.

**Acute Management**

- **These babies can be unstable** – airway management is the main key
- Keep the oro-pharynx / upper pouch clear of secretions to prevent risk of aspiration of fluids into the lungs by frequent suctioning
  - Pass Replogle tube (see relevant guideline), place on low suction (5-10 kPa) flushing with 0.5ml saline every 15 minutes.
  - If not possible to use low suction, pass a size 10 Fr tube and aspirate every 5-10 minutes.
- **Mask ventilation should be avoided** - leads to upper pouch distension and gastric distension if a lower fistula is present, leading to respiratory compromise.
- **Examine for imperforate anus** as this may worsen gastric distension.
- If the baby is spontaneously breathing with good oxygenation, intubation should be avoided. If intubation is indicated for respiratory distress, ET tube should be positioned close to carina to avoid gas flow through any fistula.
- Nil by mouth, on IV fluids.
- Nurse supine in head up position (approximately 30–60 degrees)
- 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.