Esophageal Atresia: Tracheaoesophageal Fistula

Esophageal atresia is a blockage of the oesophagus occurs with an incidence of 1 in 3000-4500 live births.

Epidemiology

- Incidence: ~1 in 3,000 to 4,500 live births.
- About 33% of affected infants are premature.
- Approximately 85% of EA cases are associated with a tracheoesophageal fistula (TEF).
- EA with TEF is a congenital malformation characterized by incomplete separation of the esophagus and trachea during embryogenesis.

Embryology

- Occurs due to abnormal division of the **tracheoesophageal septum** during the 4th to 6th weeks of gestation.
- Failure of the foregut to properly separate into the trachea and esophagus leads to a blind-ending esophagus (atresia) and/or abnormal connection (fistula).
- Alternative theory: failure of recanalization of the esophagus in the 8th week due to defective endodermal cell growth.

Classification (Gross Classification)

Type	Description	Incidence
C	Proximal esophageal atresia with distal TEF (most common)	>85%
A	Isolated esophageal atresia (no fistula)	~8%
B	Proximal TEF with distal esophageal atresia	~2%
D	TEF with atresia of both upper and lower segments	Rare
E (H-type)	Isolated TEF without atresia	~2%

Clinical Presentation

In Utero:

- Polyhydramnios due to fetus's inability to swallow amniotic fluid.

At Birth:

- Initially appears healthy.
- Excessive frothy saliva and drooling.
- Respiratory distress caused by aspiration of saliva.
- Recurrent coughing, choking, cyanosis, especially during feeding.
- Inability to pass a nasogastric (NG) tube into the stomach.
- Associated anomalies seen in VACTERL/VACTREL syndrome (Vertebral, Anal, Cardiac, Tracheoesophageal, Renal, Limb defects).

Diagnostic Workup

1. **Chest X-ray** with NG tube in situ: tube coils in upper esophageal pouch.
2. **Abdominal X-ray:**
    - Presence of gas indicates distal TEF.
    - Absence of gas indicates isolated EA (no fistula).
3. **Bronchoscopy:** helps locate fistula position.
4. **Water-soluble contrast esophagogram:** rarely used due to aspiration risk.
5. Screening for associated anomalies (VACTERL) via echocardiogram, renal ultrasound, spinal X-rays.

Management

Initial Stabilization

- **NPO (nil per os) status** — no oral feeding.
- Insert a soft, double-lumen orogastric catheter into the upper esophageal pouch for continuous low suction to prevent saliva aspiration.
- Position infant with head elevated at 30–45° to reduce reflux and aid secretion drainage.
- IV fluids to maintain hydration and electrolyte balance.
- Prophylactic antibiotics to prevent aspiration pneumonia.
- Maintain thermal environment (humidified incubator).

Definitive Surgical Treatment

- Timing and approach depend on:
    - Gestational age and weight.
    - Presence of pneumonia or other complications.
    - Length of esophageal gap.
    - Associated anomalies.

Primary Repair:

- Preferred if infant stable with a short gap (<2 cm).
- Closure of fistula and end-to-end esophageal anastomosis within first 24–72 hours.

Staged Repair:

- For long-gap atresia (>4 cm), severe prematurity, or unstable infants.
- Initial gastrostomy for feeding and cervical esophagostomy to drain saliva.
- Esophageal replacement (colonic interposition, gastric transposition) considered later.

Surgical Details (Primary Repair)

- Right thoracotomy at the 4th intercostal space (usually right side).
- Ligate azygous vein.
- Identify and protect vagus nerve.
- Dissect upper pouch, place traction sutures.
- End-to-end anastomosis with interrupted sutures to reduce stricture risk.
- Transanastomotic NG tube for postoperative feeding and suction.

Postoperative Care and Complications

Postoperative Care:

- NG tube feeding starts around day 4 post-op.
- Contrast swallow study on day 10 to check anastomotic integrity.
- Gradual oral feeding if no leak.

Complications:

- **Anastomotic leak:** occurs in up to 50%, presents with respiratory distress and sepsis; managed conservatively.
- **Stricture formation:** common weeks to months post-op; requires serial dilatations.
- **Dysphagia:** due to impaired peristalsis.
- **Gastroesophageal reflux disease (GERD):** may lead to esophagitis, Barrett's esophagus; fundoplication sometimes needed.
- **Respiratory complications:** pneumonia, pneumothorax.
- **Recurrence of fistula** or **tracheomalacia**.

Nursing Interventions

- **Airway management:**
  - Maintain patent airway; suction secretions from upper pouch to prevent aspiration.
  - Monitor for signs of respiratory distress and cyanosis.
- **Positioning:**
  - Elevate head of bed (30–45°) to prevent reflux and aspiration.
- **Nutrition:**
  - NPO until surgery or feeding guidelines from surgeon.
  - Administer IV fluids and monitor hydration status.
  - After surgery, manage feeding via NG or gastrostomy tube per protocol.
- **Infection prevention:**
  - Strict aseptic technique with suction and IV lines.
  - Administer prophylactic antibiotics as prescribed.
- **Monitoring:**
  - Vital signs closely monitored for respiratory distress or sepsis.
  - Check for abdominal distension or signs of anastomotic leak (tachypnea, fever).
- **Family support and education:**
  - Prepare parents for surgery and potential complications.
  - Teach parents about feeding modifications and signs to report.
- **Postoperative care:**

- Monitor wound and chest tube sites if present.
- Assist with pain management and comfort.
- Encourage developmental positioning and stimulation as appropriate.

High-Yield Summary

- **EA with distal TEF (Type C) is the most common form (>85%).**
- **Polyhydramnios in pregnancy and inability to pass NG tube in newborn are key diagnostic clues.**
- **Early diagnosis and surgical repair improve outcomes; staged repair for long-gap cases.**
- **Postoperative complications are common; slow feeding advancement and vigilant monitoring are crucial.**
- **Nursing care focuses on airway protection, fluid management, infection prevention, and family education.**