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TRACHEOESOPHAGEAL FISTULA REPAIR

Surgical Specialty:	General Surgery
Authors:	Holly Snyder MD, Seattle Children's Hospital, Seattle, WA Debnath Chatterjee MD, Children's Hospital Colorado, Aurora, CO

Background:

- General Considerations
  - Esophageal atresia (EA) includes a group of congenital anomalies characterized by an interruption of the continuity of the esophagus, with or without a trachea-esophageal fistula (TEF), which is an abnormal communication between the esophagus and the trachea.[1]
  - Incidence ranges from 1:2500 to 1:4000 births.[1]
  - EA is primarily diagnosed post-natally in newborns with the classic clinical picture of difficulty feeding, choking with feeds, inability to pass an orogastric tube, and the radiographic evidence of a coiled tube in the esophageal pouch. In addition, the radiographic presence of air in the stomach with EA indicates the presence of a TEF.
  - Several classification systems, such as the Vogt and Gross classifications, have been used to describe the anatomical variations. The most common presentation in 80-85% of patients is EA with a blind esophageal pouch and a distal TEF (Gross type C, Vogt 3B) (Figure 1).

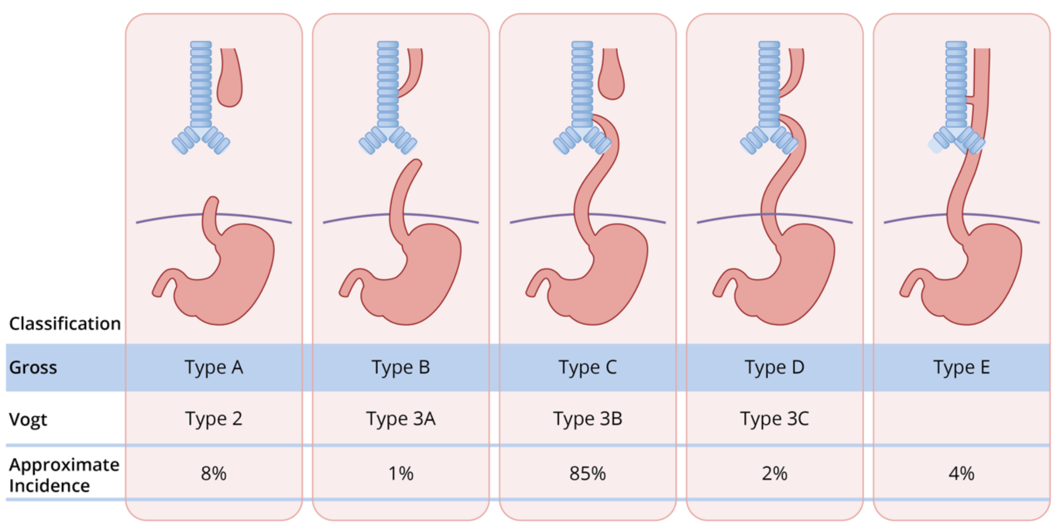

Figure 1: Gross and Vogt classification of EA and TEF. Image courtesy of Debnath Chatterjee, MD. Link.

  - Surgical repair is performed to establish gastro-esophageal continuity and to separate the tracheobronchial tree from the gastrointestinal tract by ligating the TEF. Repair is considered urgent and is usually performed in the first few days of life.

- Patient Considerations
  - About 20-30% of these patients are born prematurely, and 50% of the patients have additional congenital defects.[1]
    - Congenital heart disease -- 30% (ventricular septal defect, atrial septal defect, tetralogy of Fallot, atrioventricular canal, and coarctation of the aorta).[1]
    - Genitourinary disorders --14%, gastrointestinal disorders - 13% (duodenal atresia, imperforate anus, malrotation), skeletal anomalies --10%, and respiratory -- 6%.[1]
  - **VACTERL** association refers to a group of anomalies commonly coexisting in patients with EA and TEF. At least three VACTERL lesions are seen in 20-30% of patients with EA and TEF.[1,2] The VACTERL lesions include:
    - **V**ertebral anomalies: hemi vertebrae, scoliosis, vertebral dysgenesis.
    - **A**norectal anomalies: imperforate anus.
    - **C**ardiac anomalies: VSD, ASD, TOF, AV canal, coarctation.
    - **T**racheo-**E**sophageal fistula
    - **R**enal anomalies: renal agenesis, hydronephrosis, horseshoe kidney, urethral atresia, hypospadias.
    - **L**imb anomalies: radial aplasia, syndactyly, polydactyly, proximally placed thumbs.
  - **CHARGE** syndrome is another group of anomalies that may be present in patients with EA and TEF. CHARGE is an acronym for **c**oloboma, **h**eart defect, choanal **a**tresia, mental and growth **r**etardation, **g**enital anomalies, and **e**ar anomalies.
  - Patients at high risk for perioperative complications are premature infants with a birth weight <2 kg, associated cardiac anomalies, poor pulmonary compliance, and large pericarinal fistulas.[3]

Anesthetic Planning:

- Pre-Anesthetic Evaluation
  - Preoperative assessment includes a thorough evaluation of the cardiopulmonary system and a search for associated anomalies.[1, 2] Common preoperative investigations include:
    - Echocardiography to rule out cardiac defects and a right-sided aortic arch, which is present in about 2.5% of cases.[1, 2]
    - Blood tests should include blood cell counts, electrolytes, and crossmatch.[1]
    - A chest radiograph shows the classic upper mediastinal position of the orogastric tube. Air in the stomach confirms the presence of a TEF.
    - Ultrasound examinations to evaluate the presence of intracranial hemorrhage, rule out renal anomalies, and search for potential vertebral anomalies.[1]
    - Discussions with the surgeon should include surgical approach (open vs. thoracoscopic), mitigation strategies for accidental gastric insufflation during induction, and unplanned positive pressure ventilation (PPV). It is also imperative to discuss the plan for initial fistula evaluation via bronchoscopy, who is performing the bronchoscopy, and options for intraoperative occlusion/isolation of TEF.
- Specific or Unique Room Set-up Requirements
    - Airway
      - ETTs- uncuffed 2.5/3.0/3.5, cuffed or Microcuff ETT-3.0/3.5, if available.
      - An appropriately sized supraglottic airway should also be available.
      - Laryngoscope- Miller 0 and 1 blade or video laryngoscope.

        - Rigid or flexible bronchoscopy is often performed by the general surgery, ENT, or pulmonology team before surgical repair to evaluate the presence, location, and size of the TEF and rule out additional fistulas.[1,2]
        - Appropriately-sized flexible fiberoptic bronchoscope (FFB).
        - Balloon-tipped Fogarty catheter (4Fr) if the plan is to occlude the TEF.
    - Drugs/Infusions
        - Propofol infusion (especially during bronchoscopy) +/- remifentanil infusion or fentanyl, and/or ketamine boluses to help allow for spontaneous ventilation during airway evaluation. Attention must be paid to avoid boluses of medications that may cause apnea.
        - Lidocaine 1-2% to topicalize the vocal cords before bronchoscopy.
        - Sevoflurane or TIVA for induction and maintenance of anesthesia. Recommend paralytic when appropriate.
        - Optional: propofol or remifentanil infusion for bronchoscopy.
        - Emergency medications – succinylcholine, epinephrine bolus and infusion, and atropine**.**
        - Dextrose-containing maintenance IV fluids should be continued intraoperatively.
    - Monitors
        - Standard American Society of Anesthesiologists monitors.
        - Pre and post-ductal oxygen saturations and continuous hemodynamic monitoring with an arterial line are encouraged. If an umbilical arterial catheter is not present, a femoral or radial arterial catheter may be inserted.[2]
        - As in all neonatal cases, temperature monitoring and intraoperative measurement of blood gases, electrolytes, and glucose are imperative.
    - Access
        - Adequate venous access is essential; an in-situ umbilical venous catheter can be a good option.
    - Blood Availability
        - Minimal blood loss is anticipated, but blood should be available as the surgical repair is near large vessels in a small neonate.
    - Confirm the availability of a NICU bed for post-operative care.
    - ECMO standby for high-risk cases, such as patients with significant cardiopulmonary comorbidities.

Intraoperative Considerations:

- General
    - Anesthetic and Physiologic Implications of the Surgical Procedure:
    - Several patient factors and the planned surgical approach influence the anesthetic for TEF/EA repair. The primary goals of the anesthesiologist are to facilitate surgical exposure while optimizing pulmonary ventilation and minimizing or avoiding gastric insufflation through the TEF. This is often done by maintaining spontaneous ventilation with hemodynamic support through a bolus of pressors or an infusion. Some providers choose to use neuromuscular blockade after demonstrating minimal gastric insufflation with low-pressure PPV.[4] Communication and vigilance are paramount to successful anesthetics in these complex surgical repairs as these neonates are at high risk for hypercarbia and hypoxemia with inadequate ventilation and oxygenation.

- Induction
  - Obtain pre-induction vascular access.
  - Perform either an intravenous or inhalational induction and maintain spontaneous ventilation.
  - Topicalize the vocal cords with a local anesthetic.
  - Obtain an adequate plane of anesthesia for rigid or flexible bronchoscopy, often performed by the surgical, ENT, or pulmonary team.
- Intubation
  - Intubation is typically performed by the surgical or ENT team following visualization of the TEF. Some patients may come to the OR with an ETT in situ from the NICU.
  - The goal is to separate the TEF from pulmonary ventilation by one of the following methods. [2,4,5]
    - Position a cuffed ETT distal to TEF and proximal to the carina.
    - Intubate the main stem bronchus and achieve one-lung ventilation.
    - Occlude the TEF with a Fogarty catheter placed outside the ETT.
  - If a right-sided endobronchial intubation is chosen for one-lung ventilation, there is a high likelihood of occluding the take-off of the right upper lobe since it is closer to the carina.
  - In patients with high pulmonary compliance, consider positioning a cuffed ETT proximal to the TEF and using gentle PPV to avoid gastric insufflation.[5]

- Positioning
  - Right or left lateral decubitus position for thoracotomy on the side opposite to the aortic arch. Since most patients have a left-sided aortic arch, the left lateral decubitus position with a right thoracotomy approach is most common.
  - If a thoracoscopic approach is planned, the patient is often positioned in a modified 45° prone position (between prone and lateral position).
- Maintenance
  - Bronchoscopy (rigid vs. flexible)
    - Avoid PPV and maintain spontaneous ventilation.
    - Oxygenation insufflation during the bronchoscopy can be achieved via the side port of the rigid bronchoscope, ETT in the oropharynx, or a nasal cannula.
    - Following topicalization of the airway with lidocaine, a propofol infusion is commonly used to maintain an adequate depth of general anesthesia.
    - Some providers choose a remifentanil infusion titrated to respiratory rate; however, great care must be taken to avoid apnea.
  - Surgical Repair
    - Avoid excessive PPV before the TEF is ligated.[2]
    - Consider maintenance with volatile anesthetics and short-acting opioids
    - Neuromuscular blockade may be used if there is minimal gastric insufflation through the TEF.[4]
    - Once the TEF has been ligated, neuromuscular blockade and conventional PPV may be used.
- Hemodynamic/Physiologic goals
  - Optimize oxygenation and ventilation, which is often limited by surgical retraction, single lung ventilation, and lateral positioning.
  - The goal is maintaining $SpO_2 > 88\%$; the patient may require gentle hand ventilation with 100% oxygen.

  - Closely monitor physiologic tolerance of permissive hypercarbia due to single lung ventilation or surgical retraction of the non-dependent lung.
  - Maintain hemodynamics with gestational age-appropriate mean arterial pressures
  - The use of pressors may be necessary.
- Surgical Considerations
  - The expected blood loss is minimal, but since the surgical approach often requires ligation of the azygous vein and the surgeons are working near large vessels, there is a potential for rapid blood loss.
  - Often, the surgical repair is performed via a thoracotomy on the side opposite the aortic arch (usually right thoracotomy) via a posterolateral, extra-pleural approach.[1-3]
  - In carefully selected patients, a thoracoscopic approach may be attempted. Advantages of a thoracoscopic approach include improved surgical exposure and reduced long-term musculoskeletal comorbidities such as scoliosis, winged scapula, and rib deformities.[2,3]
  - With either approach, the TEF is first ligated, followed by a primary anastomosis of the proximal and distal esophageal segments. In patients with a long-gap EA, a staged repair is performed by shortening the gap between the segments, followed by a definitive repair at 3-6 months.
- Emergence/Disposition
  - Most neonates remain intubated for 24-48 hours postoperatively.
    - Considerations include preoperative pulmonary status, ease of surgical repair, prematurity, associated comorbidities, and issues with intraoperative ventilation.[1]
    - Patients with long-gap EA repairs will remain intubated for more extended periods to minimize tension at the repair site.[1,2]
- Post-op Care
  - Postoperative analgesia
    - Multi-modal analgesia and opioids.
    - Regional Anesthesia
      - A mid-thoracic epidural catheter placed directly at the thoracic level or threaded from the caudal or lumbar level may be considered if the spinal ultrasound is normal.
      - Alternatives to epidural analgesia include local anesthetic infiltration, unilateral intercostal blocks, paravertebral catheter, erector spinae block, or intrapleural infusion of local anesthetics.[2]

Case-Specific Complications/Pitfalls

- Complex TEF isolation:
  - Particular attention must be paid to avoid direct intubation of the TEF. Ideally, the ETT cuff should be positioned distal to the TEF and proximal to the carina. However, given the relatively common incidence of TEFs just above or at the carina, options for ETT positioning include:[2,4,5]
    - Intubation of the main stem bronchus and one-lung ventilation.
    - Occlusion of the TEF with a Fogarty catheter placed extraluminal to ETT.
    - ETT is positioned proximal to TEF, and gentle ventilation is used to avoid gastric insufflation.
- Accidental gastric insufflation
  - Complication secondary to PPV, especially in neonates with low pulmonary compliance.
  - Discuss with the surgical team if significant gastric insufflation is suspected before TEF

ligation. This can warrant an emergent surgical intervention even before the TEF is ligated. Significant gastric insufflation may result in gastric rupture.[5]

- Accidental displacement of the ETT into the main stem bronchus.
    - This typically presents with increasing peak inspiratory pressures (PIP) and desaturation.
    - Auscultate for bilateral breath sounds, confirm the position of the ETT with a FFB, and reposition the ETT.
- Inadvertent intubation of the TEF
    - Indicated by loss of $ETCO_2$ and desaturation.
    - Communicate with the surgical team immediately.
    - Auscultate for bilateral breath sounds, confirm the position of the ETT with a FFB, and reposition the ETT.
- Obstruction or kinking of the ETT
    - Indicated by increased PIP and difficulty ventilating.
    - ETT obstruction is relatively common at the end of the case and is secondary to dried blood or secretions.
    - Suction ETT and auscultate for bilateral breath sounds.
- Postoperative migration of the ETT through the tracheal suture line
    - Avoid manipulation of ETT after repair.
    - This typically presents as loss of pulmonary ventilation and $EtCO_2$.
    - Consider slightly withdrawing the ETT until breath sounds are heard, ideally with fiberoptic guidance.
    - Communicate immediately with the surgical team and convey emergency.
    - The most experienced provider must re-intubate the trachea, ideally under fiberoptic guidance.

References

1. Spitz L. Oesophageal atresia. Orphanet J Rare Dis. 2007; 2:24. PubMed link.
2. Knottenbelt G, Skinner A, Seefelder C. Tracheo-oesophageal fistula and oesophageal atresia. Best Pract Res Clin Anaesthesiol 2010; 24: 387-401. PubMed link
3. Holcomb GW, et al. Thoracoscopic repair of esophageal atresia and tracheoesophageal fistula. A multi-institutional analysis. *Ann Surg* 2005; 242:1-9. PubMed link
4. Broemling N, Campbell F. Anesthetic management of congenital tracheoesophageal fistula. Paediatr Anaesth 2011; 21: 1092-99. PubMed link
5. Ho AM, Dion JM, Wong JCP. Airway & ventilatory management options in congenital TE fistula repair. J Cardiothorac Vasc Anesth 2016; 30: 515-20. PubMed link

Other References:

1. Snyder H, Chatterjee D. Esophageal Atresia and Tracheoesophageal Fistula. OpenAnesthesia. Published January 25, 2023. Accessed October 30, 2024. Link

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Reviewed by:

Reviewer #1: Leah Webb, MD, Children's Hospital Colorado

Reviewer #2: Anushree Doshi, MD, Children's Hospital of Philadelphia

Reviewer #3: Tracy Wester, MD, University of North Carolina

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