Tracheoesophageal Fistula (Congenital)

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Risk

- Incidence of EA + TEF: 1:3500.
- EA/TEF in twins is 2.6 times higher than in singletons.

Perioperative Risks

- Respiratory distress
- Stomach distention, rupture (1%) possible
- Dehydration and hypoglycemia

Worry About

- Respiratory status
- Congenital heart disease (23%): PDA, VSD, TOF, ASD, right-sided aortic arch
- Location of fistula (typically unknown)
- Fluid and metabolic status
- Prematurity
- Low birth weight (common)

Overview

- Association with vertebral abnormalities (17%), anal and/or duodenal atresia (12%), cardiovascular anomalies (23%), tracheoesophageal fistula, esophageal atresia, renal (16%) and/or radial anomalies, and limb defects (10%) (VACTERL association).
- Respiratory compromise is possible from aspiration pneumonitis, tracheomalacia (usually present but clinically significant in 10–20% of cases), gastric distention, prematurity, and congenital heart disease.
- Gross classification: (A) EA without TEF (8%); (B) proximal TEF with distal EA (3%); (C) distal TEF with proximal EA (85%); (D) proximal and distal TEF (<1%); (E) TEF without EA or "H"-type TEF (4%). Gross type A is the only type without TEF and the attending risk of aspiration but the most likely to have associated anomalies (up to 65%).
- Preop echocardiography is useful for identifying cardiac anomalies and the presence right aortic arch (2.5–5%); if present, left-sided approach is required.
- In Gross C TEF, the fistula may be below the carina (11%) or within 1 cm proximal to the carina (22%), making the classic recommendation of positioning the ET tip between the carina and the fistula impossible or difficult. If margin between ET tip and carina is low, inadvertent endobronchial intubation of the compressed right lung may occur due to tube migration, and inadvertent ventilation of the fistula can occur due to tube migration. Repositioning of ET may lead to inadvertent endobronchial or fistula intubation.
- Most Gross C type do not have a long esophageal gap.
- Some fistula openings may be ≥3 mm with increased risk of stomach insufflation and inadvertent fistula intubation.
- Having the ET bevel facing posteriorly reduces the risk of inadvertent intubation in the beginning and subsequent ET position adjustments, if required.
- Avoiding high positive pressure ventilation reduces stomach insufflation.
- The fistula subtends an angle not unlike those of the mainstem bronchi, not orthogonally as illustrated in many textbooks. This makes it plausible on the one hand to accidentally intubate with the ET, but makes it easy to insert a balloon-tipped Fogarty cath (Plan A, discussed later).

Etiology

- Genetic syndromes associated with EA/TEF include all full trisomies (Down syndrome, Edwards syndrome, Patau syndrome), single gene disorders (CHARGE syndrome, Feingold syndrome, Opitz syndrome, and Fanconi anemia).
- Environmental factors implicated include maternal exposure to methimazole, exogenous sex hormones, alcohol, tobacco, diethylstilbestrol, infectious diseases, advanced maternal age, and working in agriculture or horticulture.
- An adriamycin-induced EA/TEF rat model facilitates embryologic study of the disease.

Usual Treatment

- Prevention of aspiration (NPO, continuous suctioning of esophageal pouch, elevation of head).
- Urgent surgery but 24–48 h for optimization with antibiotics and fluids is acceptable. Emergent if positive pressure ventilation is required with progressive distention of stomach; consider placement of a blocker in the fistula in NICU if stomach distention is a problem.
- Primary surgical correction via right posterolateral extrapleural thoracotomy below the tip of the scapula.
- Alternatively, use right thoracoscopic approach with thoracoscope inserted just caudad to the right scapula tip through which CO_2 is insufflated at a rate of 0.5 L/min to a pressure of 6 mm Hg into the right pleural cavity to collapse the right lung gradually.
- Extrapleural approach does not lead to empyema or mediastinitis if anastomosis leak occurs (up to 20%) but only an esophagocutaneous fistula.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	PDA, VSD, ASD, right-sided aortic arch	Cyanosis, tachypnea, respiratory distress	Murmur, cyanosis, enlarged liver, hypotension, bounding pulses	CXR, ECHO
RESP	Pneumonia, tracheomalacia, prematurity	Respiratory distress	Decreased breath sounds, tachypnea, cyanosis	CXR, ABG, bronchoscopy
GI	Gastric distention, anal and duodenal atresia	Enlarged abdomen, respiratory distress	Enlarged and tympanic abdomen	CXR, KUB series
RENAL	Dysplastic/dysfunction	UO	Palpation for kidneys	Abdominal US

Key References: Ho AMH, Dion JM, Wong J: Airway and ventilatory management options in congenital tracheoesophageal fistula repair, *J Cardiothorac Vasc Anesth* 30(2):515–520, 2016; Pinheiro PF, Simões e Silva AC, Pereira RM: Current knowledge on esophageal atresia, *World J Gastroenterol* 18(28):3662–3672, 2012.

Perioperative Implications

Preoperative Preparation

- NPO
- Placement of an orogastric cath into esophageal pouch for low continuous suction of secretions.
- Elevation of the head of the bed to minimize reflux.
- Consider antireflux and antacid medications.
- IV fluids and glucose.
- If intubation is needed in NICU, consider intubating the fistula with a blocker first.

Monitoring

- Arterial line is usually not required.
- Preductal (right upper limb) and postductal (lower limbs) oximetry.
- Left chest stethoscope for right-sided approach and vice versa.

Airway

- Plan A: Insert 3-Fr balloon-tipped Fogarty-type cath prior to ET. Immediate fiberoscopy via ET to ascertain location of fistula. If fistula is too distal to block by the ET, intubate the fistula with balloon cath. The surgeon is informed of the presence of a blocker in fistula, and the blocker is withdrawn prior to ligation.
- Plan B: Position the ET tip between the fistula and carina. This is the plan of choice if fiberoscopy after ET placement confirms the ET tip near carina and fistula is not in view. If carina-fistula distance is very small, or fistula is at or below carina, consider switching to Plan A.
- Plan C: If Plan B is impossible and Plan A is not in place, ventilate with the lowest pressure possible. There is significant risk of stomach insufflation; emergency decompressive gastrostomy may be required in rare situations.
- Plan D: Intubate the left mainstem bronchus with the ET (with bevel facing right and ET slightly concave to left, 92% chance it will go into the left mainstem).
- Choose an ET without Murphy eye to reduce exposure of fistula to positive airway pressure during surgery and in NICU.
- Cuffed versus uncuffed ET (without Murphy eye) based on individual preference.
- Intubate with bevel facing posteriorly to reduce chance of inadvertent fistula intubation.
- Subglottic stenosis may be present.

Induction

- Maintain spontaneous ventilation; minimal positive pressure ventilation if necessary.
- Topicalization of airway with lidocaine prior to balloon cath and ET placements.

Maintenance

- Pt in left lateral (thoracotomy) or left semi-prone position (thoracoscopic approach).
- Adjustment of ET depth quite difficult during surgery because of impeded access to airway.
- Positive pressure ventilation necessary once the right lung space is entered.
- Intraop O_2 desaturation is common. Most commonly this is due to right lung compression and/or mediastinal retraction by surgeon; temporary resumption of two-lung ventilation or easing of retraction may be necessary. Communication with surgeons is crucial. Other causes include inadvertent right mainstem intubation; ventilation of or inadvertent migration of ET into fistula; ET obstruction due to kinking, secretions, or blood; severe cardiovascular compromise; or bleeding while accessing the fistula, which is deep to the azygos vein.
- Intraop low or disappearance of $ETCO_2$ is common. Causes include migration of the ET tip into the right mainstem bronchus or fistula, cardiovascular compromise, or ET blockage. A left lung stethoscope helps monitor for inadvertent loss of ventilation to the left lung.
- In severely hypoxic and hypercarbic cases, use high frequency ventilation.
- Dopamine infusion may be required to maintain blood pressure.

Extubation

- Respiratory distress after surgery is common due to pain, tracheomalacia, prematurity, and aspiration prior to fistula ligation.

- High positive airway pressure from mask or ET ventilation puts stress on the fistula stump. ET tip distal to fistula (if possible) and use of ET without Murphy eye reduces such stress.
- Consider sedating and paralyzing the infant for several days as laryngoscopy puts stress on the esophageal anastomosis; intubation risks inadvertent intubation of the fistula stump.
- Surgeons may request placement of a trans-anastomotic tube via the mouth.
- Discuss with surgeon, including the degree of anastomotic tension.

Adjuvants

- Fiberoptic bronchoscopy to position the ET and fistula blocker (if deployed) and to ascertain the location of the fistula. Rarely, more than one fistula may be present.
- Have dopamine drawn and hooked up.
- May consider having NO and standby drugs for reducing pulm Htn.
- Emergency gastrostomy rarely indicated but could be lifesaving.
- High-frequency ventilator.

Postoperative Period

- Sedate and paralyze for several days unless all favorable conditions are present for early extubation and chance of requiring re-intubation is very low.
- Postop elective ventilation may protect against the development of leak in primary repair.
- Keep neck flexed to reduce tension on the esophagus.
- Respiratory distress after extubation poses a dilemma. Consider the possibility of inadequate pain control, tracheomalacia, pneumonia, and prematurity as causes. Pain may be a concern even after several days, especially with a chest drain in situ, and during handling, a neuraxial block (e.g., caudal entry thoracic epidural) may help avoid respiratory distress and the need for reintubation. Be wary of dynamic airway closure caused by tracheomalacia during rigorous crying. Caudal morphine may provide both analgesia and sedation. Fentanyl/adjunct analgesia infusion is an alternative but finding that sweet spot can be challenging.
- Antibiotics for 48–72 h or longer as required.
- Avoid tracheal and esophageal suctioning (transanastomosis cath is in place).
- A contrast study prior to oral feeding may be performed at 7–10 d postop.
- Minor leaks may heal spontaneously.
- Major leak requires surgical repair.

Anticipated Problems/Concerns

- ET not in ideal location, especially with pt movement and surgical manipulation. Correction by sliding the ET proximally and distally has associated risks (noted previously).
- Stomach insufflation.
- Respiratory distress.
- Cardiovascular anomalies which may affect hemodynamics and respiratory parameters.
- Low birth weight.
- Risk of prematurity.
- TEF recurrence (3–14%).
- A history of EA/TEF repair is associated with gastroesophageal reflux (50%).
- Tracheomalacia (an obstruction that exceeds 50% of the anteroposterior diameter of the trachea during inhalation) is found in 60% of children between 2–3 y.
- Respiratory symptoms during the first 5 y of life (>66% of cases), during adolescence (40%), and during adulthood (10%).

Transfusion-Related Acute Lung Injury

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Risk

- Can occur in any pt receiving blood or blood products, including platelets, plasma, cryoprecipitate, and rarely, IVIG.
- Overall incidence probably <1%; increasing awareness of the syndrome has resulted in improved recognition.
- Use of leukodepleted blood has decreased the incidence of packed red cell-related lung injury.

Perioperative Risks

- Noncardiogenic pulm edema, usually within 6 h of transfusion.
- Mortality reported as 5–10%.

Worry About

- O_2 toxicity
- Barotrauma or volutrauma secondary to PPV
- May be confused with transfusion-associated circulatory overload (TACO)

Overview

- Classic presentation is acute development of resp compromise indistinguishable from ARDS.
- Symptoms usually begin within 1–2 h after transfusion and may be manifested by 2–6 h.
- Severe hypoxemia and bilateral infiltrates are always present, while hypotension, fever, and pink frothy sputum may be present in some.
- Dx is clinical and one of exclusion.

Etiology

- Classically, has been attributed to the presence of leukocyte antibodies in the plasma of multiparous donors directed against recipient WBCs.
- Alternatively, may be effect of biologically active lipids in stored cellular blood components.
- Pulm edema arises from capillary injury rather than volume overload.

Usual Treatment

- Supportive care: Ventilation, if required, or supplemental O_2. There are no clear indications for steroids. Generally resolves within 1–4 d with appropriate care and no supervening complications.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
CV	Pulm edema		S_3, S_4	PA cath, ECHO
RESP	Pulm edema	Recent transfusion	Rales, hypoxemia	CXR: Bilateral infiltrates, SpO_2
HEME	Leukoagglutination			Agglutination of recipient leukocytes by donor plasma; contact blood collection agency

Key References: Triulzi DJ: Transfusion-related acute lung injury: current concepts for the clinician, *Anesth Analg* 108(3):770–776, 2009; Silliman CC, Ambruso DR, Boshkov LK: Transfusion-related acute lung injury, *Blood* 105(6):2266–2273, 2005.

Perioperative Implications

Preoperative Preparation

- Acute respiratory compromise may occur within 6 h of a transfusion, usually with FFP. Unlike RBCs, FFP is not leukodepleted; the presence of WBC in FFP is associated with an inflammatory response similar to that for large volumes of plt transfusion.
- Usually related to massive transfusion, although on occasion may happen after a single unit transfusion.

Monitoring

- PA cath may aid in the exclusion of cardiac etiology (i.e., normal wedge pressure).
- Beta-natriuretic peptide level may be checked to differentiate TRALI from TACO.

Postoperative Period

- Most pts require ventilatory support for several d.
- Ventilator management may be appropriate for ARDS.

Anticipated Problems/Concerns

- O_2 toxicity and barotraumas
- Hemodynamic instability