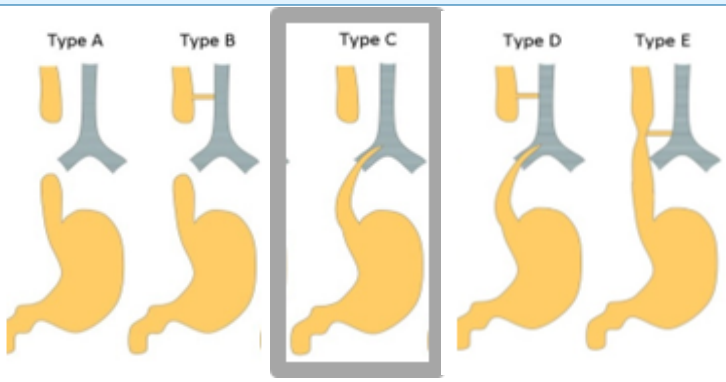
Tracheoesophageal Fistula / Esophageal Atresia (Short Segment TEF/EA) Care Guideline



Available Order Sets
- TEF/EA Pre-Op Order Set
- TEF/EA Post-Op Order Set

Parent/Family Education
- TEF and/or EA Family Teaching Handout

Inclusion Criteria: Any infant admitted to the NICU for suspected short segment TEF/EA except those that meet Small Baby Unit criteria (< 1000 grams or 28 weeks gestation).

Pre-Operative Assessment

- Polyhydramnios and/or prenatal ultrasound diagnosis.
- Inability to manage secretions, including signs of drooling, choking, and coughing.
- Inability to pass OG/NG tube beyond approximately 10 cm with coiling of tube in esophagus.
- X-ray: A gastric tube may end in the proximal esophagus. A gasless abdomen suggests pure EA (Type A). Gas in the abdomen suggest tracheoesophageal fistula.

Admission and Pre-Operative Interventions

- Provide respiratory support if necessary.
  - If mechanical ventilation is required attempt to use low mean airway pressures or high-frequency oscillatory ventilation (HFOV) to minimize gastric distention.
  - Maintain endotracheal tube (ETT) close to level of carina to bypass TEF. If possible turn ETT to have blue line facing posterior.
  - Avoid use of continuous positive airway pressure (CPAP) and bag mask ventilation if possible.
- Obtain chest x-ray (CXR) to verify ETT placement.
- Gently insert 10 Fr Replogle to predetermine length or as far as possible (usually 9 – 13 cm) and connect to low continuous suction.
  - Maintain patency with irrigations or infusion of 3 mL air q4h.
  - Use sterile water for irrigations – ***DO NOT*** use saline.
  - May require use of double chamber set-up if difficulty in maintaining output.
- Elevate HOB 30 – 45°
- Broad spectrum antibiotics (ampicillin and gentamicin) ***ARE NOT*** routinely indicated for TEF/EA diagnosis – may be started for other clinical indications per clinical teams discretion.
- Make NPO and order milk oropharyngeal (MOPs)
- Consult Pediatric Surgery and ENT on admission.
- Obtain echocardiogram (echo) ASAP – Must be obtained prior to OR.
  - Include in comment: Evaluate for left or right-sidedness aortic arch.
  - After echo has been read, evaluate if cardiac anesthesia indicated. If yes, alert surgical team to contact.
  - Determine if patient requires a high risk anesthesia consult (h/o anesthetic complications, high risk airway, significant/multiple comorbidities) if yes place order.
- Insert PICC and at least 1 peripheral intravenous (PIV) and provide maintenance IV fluids. Place peripheral arterial line (PAL) prior to OR – plan for the day prior to surgery.
- Obtain routine admission and pre-op labs:
  - Transport work-up (Blood Type and Screen), MRSA & VRE surveillance
  - CBC with diff, CMP
  - Blood culture (if indicated and not previously done)
  - Chromosomal microarray for genetic work up
- Prior to OR:
  - Surgical NICU attending to discuss with operating surgeon regarding patient's candidacy for intra-operative placement of transanastomotic nasogastric tube.
  - Please consider placement of NG if baby is born at ≤ 35 weeks or if has an additional neurologic, cardiac or genetic diagnosis as these patients often have feeding/swallowing challenges that may impact time to full oral feeding.
- Place 20 mL/kg PRBC's on hold for OR.
- If intubated, CXR the morning of surgery.
- Bring unopened feeding time to be used as a stent.

Approved Evidence Based Medicine Committee
Original: 3/16/2016
Review/Revised: 11/20/2024; 9/20/2023

Reassess the appropriateness of Care Guidelines as condition changes and 24 hrs after admission. This guideline is a tool to aid clinical decision making. It is not a standard of care. The physician should deviate from the guideline when clinical judgment so indicates.

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Post-Operative Interventions

- Check blood gas, blood sugar and temperature upon return to NICU.
- CXR to confirm ETT placement.
  - Maintain ETT at precise location to prevent trauma to surgical site.
  - ETT suction only to precise length of ET tubes to prevent damage to tracheal repair
- Suction oral/nasal cavity only to posterior pharynx, ***DO NOT*** deep suction.
- Elevate HOB 30 – 45°
- Maintain neck in a neutral position.
  - Do not hyperextend neck to avoid surgical site trauma.
- Maintain chest tube to water seal drainage unless surgical preference is to –20cm H_2O suction.
- Gastric tube (G-tube), if placed, to gravity drainage.
- NPO, maintain central IV access, and provide maintenance TPN and lipids
- Surgically placed nasogastric tube (NGT), if placed during OR, to gravity drainage. It acts as a stent to the anastomotic site.
  - ***DO NOT MANIPULATE OR REPLCE NG TUBE!*** Ensure tubing is secure at all times. Page surgery if tube becomes dislodged.
  - Some patients may return from OR without NGT per surgeon discretion.
- Administer post-operative cefoxitin for 24 hours after surgery (alternative regimen: ampicillin and cefepime).
- Post-operative labs on POD #1:
  - Blood gas, BMP, Mg, Phos, T/D bilirubin, CBC
- Proceed with remainder of VACTERL work up.
- Aim for extubation around 48-72 hours post-op.
  - Order peri-extubation dexamethasone (0.2 mg/kg q6h x 4 doses) to start the day/ evening prior to extubation → can extend course if concern for airway edema.
  - Notify anesthesia about planned extubation if concern for airway management → anesthesia ***DOES NOT*** need to be at bedside for extubation.
  - ***DO NOT*** extubate patient with neonatology attending and surgeon approval. Re-intubation by attending only due to risk of anastomotic rupture.
  - No CPAP or noninvasive positive pressure ventilation (NIPPV) after extubation.
- Once extubated, contact ENT for bedside scope to evaluate vocal cords within 72 hours of extubation and ***PRIOR*** to stating PO feeds.
- Begin and maintain patient on proton pump inhibitor (PPI) to protect surgical site from stomach acid and decrease risk of strictures.
- With surgeon approval, may consider early (continuous post-pyloric) trophic continuous feeds after extubation and before esophagram.
- Esophagram on POD #7 to assess for healing of anastomotic site and signs of leakage.
- If no leak on esophagram, initiate oral feeds and place order for 'Feeding/Swallow Team Evaluation'.
  - Start feeds on Surgical Guideline 2 with human milk.
  - Surgery to remove chest tube once feeds are tolerated for 24 hours.
- If requires esophageal dilation, generally must wait at least 6 weeks from initial repair.

Pain Management

- Use Pain Guideline 2 for post-operative pain management
  - Titrate and wean per guideline

Follow Up Studies / Consults

Obtain:

- Sacral ultrasound (US) to evaluate for tethered cord
- Renal US on DOL #2 or later
- CXR and KUB to evaluate for vertebral anomalies
- Genetics consult: Obtain chromosomal microarray analysis

VACTERL Criteria – Inclusion of at least 3 of the following:

Vertebral – hemi vertebrae
Anus – imperforate anus
Cardiac – TOF, VSD, right sided arch
Trachea – tracheoesophageal fistula
Esophagus – esophageal atresia
Renal – solitary kidney or reflux
Limbs – absence of radius

Considerations for Management

- Aspiration
- Gastroesophageal Reflux
- Infection from pneumonia, central line, chest tube, or surgical site
- Anastomotic leak or stricture
- Esophageal dysmotility
- Vocal cord paralysis
- Tracheomalacia
- TEF cough and stridor

Discharge Planning

- PPI home prescription.
- Surgical follow up 2 – 3 weeks after discharge.
- Pediatrician 1 – 3 days after discharge
- Bridge Clinical follow–up.
- Specialist follow up: GI (mandatory), other services as needed if consulting.

Shafer, Ahmad, Kabeer, O'Toole, Lawrence and Haymond, October 2024

Tracheoesophageal Fistula / Esophageal Atresia (Short Segment TEF/EA) Care Guideline References

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