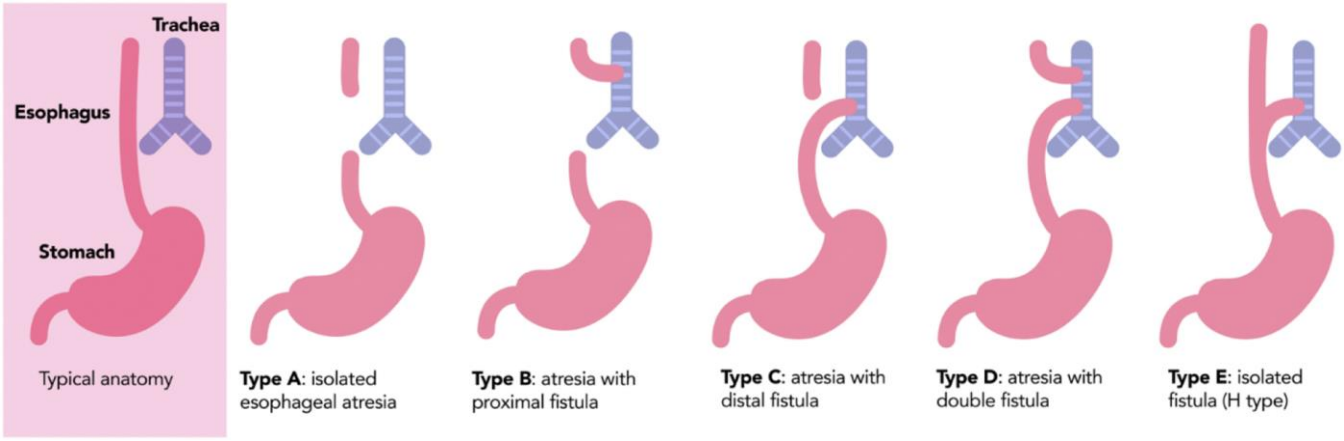
Newborn Critical Care Center (NCCC) Clinical Guidelines

Esophageal Atresia / Tracheoesophageal Fistula (EA/TEF) Guideline

BACKGROUND

- Incidence: 1 per 2500-4000 live births with a slight male predominance.[1, 2]
- There are various types of esophageal (EA)/ tracheoesophageal fistula (TEF):



Type A: The upper and lower segments of the esophagus both end in a pouch. Type A does not include tracheoesophageal fistula (TEF), which is an abnormal connection between the trachea and esophagus that can allow swallowed food or liquids into the lungs.

Type B: The lower segment ends in a pouch, and the upper segment has a TEF. This type of esophageal atresia is rare.

Type C: The upper segment ends in a pouch, and the lower segment has a TEF. This is the most common type of esophageal atresia (accounts for ~85% of cases).[3]

Type D: Both the upper and lower segments have a TEF. This type of esophageal atresia is the rarest.

Type E: TEF only ("H-type", usually not diagnosed in newborn period).

ASSOCIATED CONDITIONS

Approximately 50% of infants with EA/TEF have other associated congenital anomalies.[1,2] One of the most common associations is VACTERL association (Table 1). Approximately 10-30% of EA/TEF cases are part of the VACTERL association.[4]

Table 1: VACTERL Association

Presence of at least 3 of the following congenital malformations:

Vertebral – vertebral defects

Anus – anorectal malformations

Cardiac – cardiac defects (such as VSD, TOF, PDA)

Trachea – TEF

Esophagus – esophageal atresia

Renal – renal anomalies (such as horseshoe kidney, polycystic kidney, absent kidney)

Limbs – limb abnormalities (such as MSK problems or polydactyly)

Other associated conditions/findings include[4]:

- Charge Syndrome
- AEG Syndrome (anophthalmia/microphthalmia, genitourinary anomalies)
- Digestive tract anomalies such as congenital diaphragmatic hernia or duodenal atresia
- Trisomy 13, Trisomy 18, and Trisomy 21

PRESENTATION

EA/TEF is confirmed after birth. However, prenatal features that can be concerning include small or absent stomach, dilated or enlarged esophagus (pouch), or polyhydramnios. These features should raise your suspicion for EA but are not specific to EA and may be seen with other birth defects.

Postnatal presentation can include:

- Inability to pass an OG/NG
- Inability to manage secretions: signs of drooling, choking, coughing
- Cyanosis with feeding
- Respiratory distress

X-ray confirmation will demonstrate a gastric tube in the proximal esophagus pouch. A gasless abdomen suggests EA without fistula (Type A). Gas in the abdomen suggests a distal fistula is present (Type B ,C, D).

PRE-OPERATIVE MANAGEMENT FOR TYPE C EA/TEF
(Management for other types may vary and should be discussed with surgical team)

- Consult Pediatric Surgery at time of admission.
- Avoid positive pressure ventilation when possible (sometimes may be needed during resuscitation).
- If at any point a baby with Type C EA/TEF is intubated, the surgery team should be aware.
- Place 10Fr Replogle in term infants (preterm infants may require smaller size) to upper pouch to low continuous suction.
    - In general, the Replogle should maintain patency since it is suctioning oral secretions; however, the Replogle can be flushed with 3 mLs of air if needed to maintain patency.
    - Nursing may flush and replace Replogle as needed in the pre-op period unless otherwise specified.
- Obtain CXR to confirm Replogle and/or ETT placement.
- Discuss timing and type (UVC vs PICC) of central access with the primary NCCC & Pediatric Surgery teams.
    - This should be discussed prior to the OR for EA/TEF repair so that appropriate planning and consent can be obtained.
- Admission/pre-op labs:
    - CBCd, CMP, Type and Screen
    - Karyotype and Microarray (if not already sent with cord blood and testing is desired by parents)

    - Blood culture if clinical concern for infection
- Start broad spectrum antibiotics (ampicillin/gentamicin) with plan to continue through post-operative period, but may be stopped post-operatively in the absence of infection or absence of a tracheoesophageal fistula.
    - If the patient has a long gap esophageal atresia and will not be repaired in the immediate postnatal period, but requires surgical feeding access, discuss duration of antibiotics with Pediatric Surgery team.
- STAT echocardiogram to evaluate for congenital heart disease and aortic arch orientation.
    - An echo must be obtained before the patient goes to the OR to ensure the proper anesthesia team involved. This does not usually affect the sequence of surgery even in the presence of congenital heart disease.
    - This also provides information for anatomic planning with right vs left sided aortic arch and the presence/absence of an aberrant right subclavian artery.
- Start TPN/SMOF as early as possible given prolonged NPO period.
- If time allows prior to surgery, obtain additional studies to assess for VACTERL features (see Table 2).
- **REMEMBER**: anorectal malformation with a TEF is a ***surgical emergency!***
    - No stool output could indicate an occult anal obstruction/atresia.

Table 2: VACTERL Evaluation:

- ECHO prior to OR
- RENAL US after 48 hours life
- SACRAL US to evaluate for tethered cord, non-urgent
- CXR/KUB to evaluate for vertebral anomalies
- Karyotype / microarray

PRE-OP HANDOFF WITH ANESTHESIA / PEDIATRIC SURGERY

- Communicate the plan that the patient is to return from OR intubated.
- Anesthesia to call 4 NCCC RT via Vocera when preparing for intubation. NCCC RT will go to the OR and secure the ETT.
- OG/NG should be secured per attached instructions (Appendix 1).
- Discuss with Pediatric Surgery prior to OR whether they anticipate the need for paralysis.
    - If paralysis is anticipated, the medication should be ordered and on the unit upon the infant's return from the OR.
    - Sometimes paralysis is not initially anticipated but becomes necessary based on the operative course. Paralysis need should be confirmed during post-op handoff.

POST-OP HANDOFF

Ensure all points are addressed during surgical handoff at bedside, if not, page Pediatric Surgery for clarification.

- Location of fistula & repair, degree of tension at anastomotic site
- Need for paralysis or heavy sedation in immediate post-op period
    - If paralysis was not originally anticipated preoperatively but is now necessary, order the paralytic bolus stored in the NICU and order the paralytic infusion STAT from pharmacy
- Confirm whether ETT position needs to be intentionally maintained at a certain depth based on repair site
- Extubation plan:
    - If extubating, what amount of respiratory support would be acceptable (i.e. 2LPM)
    - If not extubating and ETT was not taped by NCCC RT in OR, Pediatric Surgery and Pediatric Anesthesia team should be asked to stay at the bedside while ETT is retaped
    - Discuss possibility of pre-extubation steroid use and duration with Pediatric Surgery team prior to ordering
- Type and length of antibiotic coverage
- If a trans-anastomotic feeding tube is present, confirm insertion depth and add depth to bedside diagram.
- Discuss plan for chest tube/chest drain including suction vs water seal. If using suction, confirm level of suction with surgery team.
- Status of family update: if the family has not been updated by the Pediatric Surgery team, provide the surgery team with the family location and offer interpreter iPad as needed.
- Update TEF/EA bedside sign with relevant details. Attach sign to warmer bed.
- The above details should be included in the post-operative significant event note written by the NCCC team.

POST-OPERATIVE MANAGEMENT

- Obtain post-op labs, chest x-ray, and continue broad-spectrum antibiotics per discussion with Pediatric Surgery team.

Respiratory Status

- If intubated, the goal is to wean towards extubation by ~48-72 hours post-op.
    - Avoiding retaping ETT if anticipated extubation is soon.
    - Confirm whether ETT position needs to be intentionally maintained at a certain depth based on repair site(s).
    - Obtain daily CXR while intubated.
- If requiring respiratory support following extubation:
    - Confirm surgery's preference for maximum post-op positive pressure (i.e. maximum 2L HFNC).

- Discuss administration of pre-extubation steroids (possibility of steroid use and duration) with Pediatric Surgery team prior to ordering.
- Maintain chest tube to waterseal until esophagram demonstrates no leak *(unless otherwise specified by surgery).*
- Reintubation:
    - Reintubation should be performed by the most experienced provider using a CMAC to avoid trauma to the surgical site.
    - Positive pressure should be avoided for preoxygenation if possible. If positive pressure is needed, maintain PIPs as low as possible while providing effective ventilation.
    - If RSI is required, the infant should be intubated expeditiously after the paralytic is given to avoid positive pressure.

NCCC Nursing Considerations

- **DO NOT MANIPULATE THE NG/OG TUBE.** An NG/OG tube in a post-op EA patient is a trans-anastomotic feeding tube, which means that it passes the esophageal repair site. Any movement of this feeding tube increases the risk for causing damage to the site. If a tube is incidentally removed replacement carries even higher risk of injury.
- If the tube is dislodged, contact the Pediatric Surgery team immediately using the Pediatric Surgery Chief Resident Pager, if no response in 15 minutes, page attending surgeon on call.
- Feeding tube depth should be written on the bedside sign.
- Secure the feeding tube per the steps in Appendix 1.
- No deep suctioning through the GI tract or beyond the end of the ETT.
    - Suction oral/nasal cavity only to posterior pharynx to avoid damage to anastomotic site in esophagus and fistula repair site in trachea.
- Post-op positioning *(may vary case-by-case depending on tension, type of approach, etc.)*
    - HOB elevation
    - Neck positioning – maintain neutral position, do not hyperextend neck
- Bedside sign (see Appendix 2) should be attached to the patient's warmer bed.

Pain and Sedation

- Post-op pain / sedation should focus on adequate pain control while avoiding over-sedation.
- Schedule IV Tylenol for 72h post-op
    - IV Tylenol must be re-ordered every 24 hours
- If anticipating a prolonged period of intubation, the patient should be on a continuous infusion of pain/sedation medications with the shortest-lasting effective agent possible.

Nutrition

- Start PPI to protect surgical site from stomach acid and decrease risk of strictures.
- Esophagram to be obtained on POD #5 (occasionally on POD #7 depending on case and

radiology availability).

- Consult SLP for oral stimulation.
- If G-tube is present (in pure EA patients), ensure it is to gravity post-op, and discuss timing of feeding initiation.
- If a transanastomotic feeding tube is present, feeds should be held until POD #2. At the time of feeding initiation, they should be started using a continuous rate.
- Gastric residuals do not need to be monitored with continuous feeds.
- Timing of initiation & advancement of continuous vs bolus enteral feeds should be discussed with the surgical team.
- Infants with an EA and small stomach may require a slower advancement to full volume, bolus feeds.
- Venting the g-tube:
    - **Continuous feeds:** the feed should be paused to vent the g-tube every 4-6 hours
    - **Bolus feeds:** vent the g-tube before the feed

References:

1. O'Shea D, Schmoke N, Porigow C, et al.. Recent Advances in the Genetic Pathogenesis, Diagnosis, and Management of Esophageal Atresia and Tracheoesophageal Fistula: A Review. *Journal of Pediatric Gastroenterology and Nutrition.* 2023; 77 (6): 703-712. doi: 10.1097/MPG.0000000000003952.
2. Pinheiro PF, Simões e Silva AC, Pereira RM. Current knowledge on esophageal atresia. *World J Gastroenterol*. 2012;18(28):3662-3672. doi:10.3748/wjg.v18.i28.3662
3. Gutierrez RS, Guelfand M, Varela Balbontin P. Congenital and acquired tracheoesophageal fistulas in children. *Seminars in Pediatric Surgery*. 2021;30(3):151060.
4. de Jong EM, Felix JF, de Klein A, Tibboel D. Etiology of esophageal atresia and tracheoesophageal fistula: "mind the gap". *Curr Gastroenterol Rep.* Jun 2010;12(3):215-22. Doi:10.1007/s11894-010-0108-1
5. Salik I, Paul M. Tracheesophageal Fistula. StatPearls. StatPearls Publishing LLC. 2025.

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APPENDIX 1

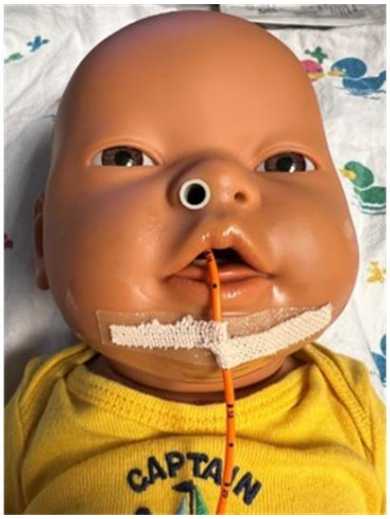
Securement of Orogastric Tubes for TEF/EA Patients

(Disregard the nasal trumpet in model, these patients do not require a nasal trumpet.)

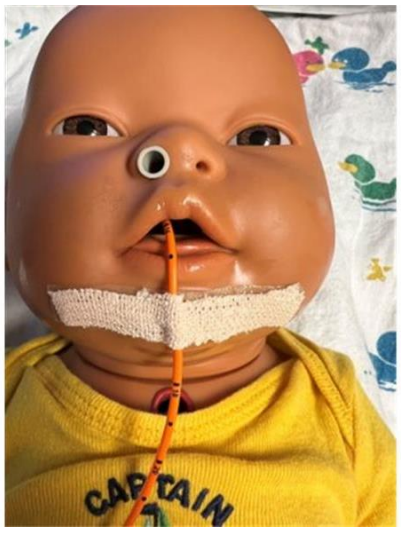
1. Prepare skin with no sting barrier.

2. Apply appropriately sized extra thin Duoderm for skin protection.

3. Chevron one piece of Elastikon tape around the OG tube. Elastikon is recommended due to possibility of excess secretions interrupting integrity of silk tape. Make sure to keep Elastikon within the boundaries of the Duoderm.



4. Apply second piece of Elastikon tape over chevron for additional securement, keeping Elastikon within the boundaries of Duoderm.



APPENDIX 2

Bedside Sign

(Laminated copies will be kept on 4NCCC)

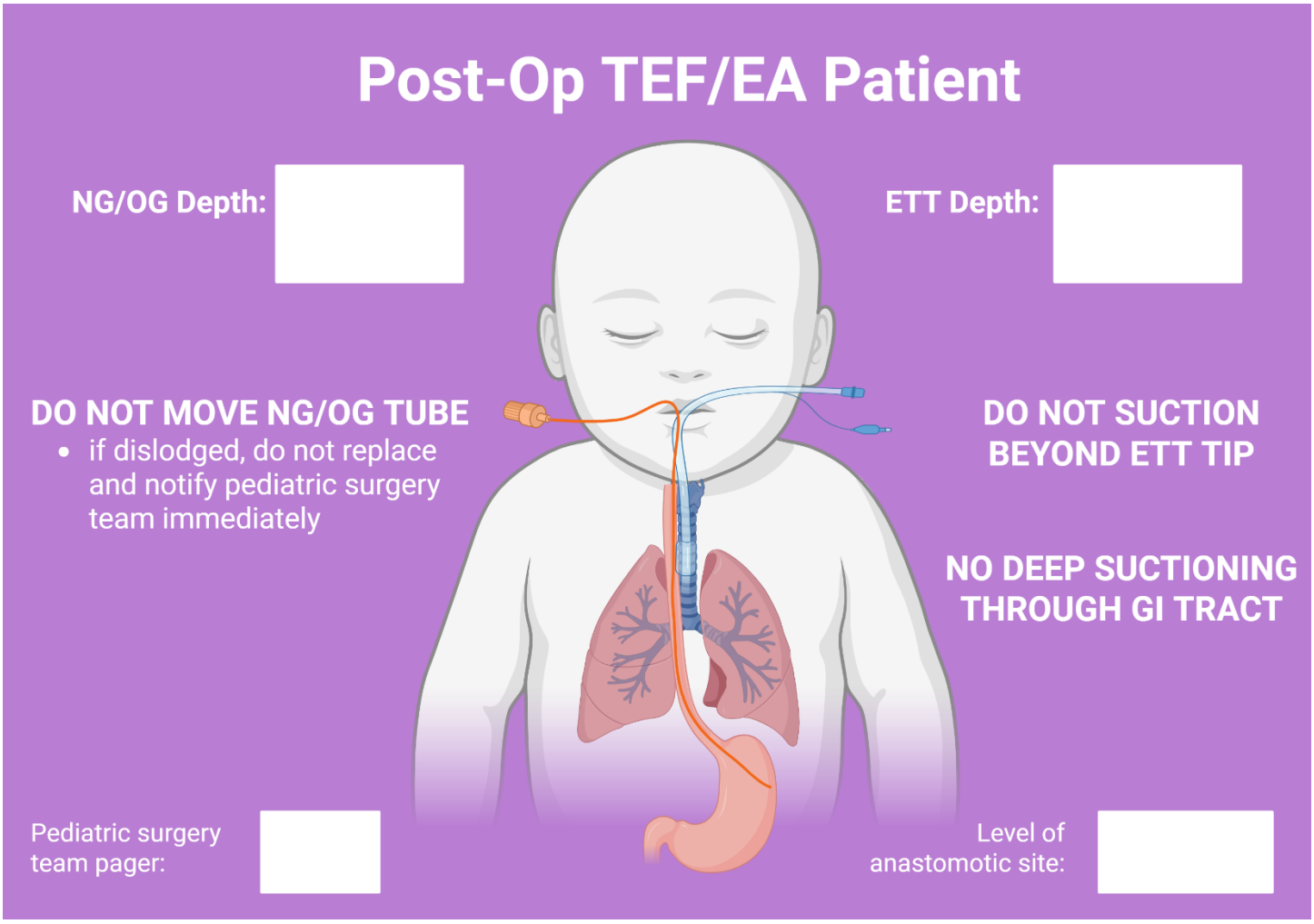


Image created with BioRender.