Advances in Complex Congenital Tracheoesophageal Anomalies

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KEYWORDS

• Esophageal atresia • Tracheoesophageal fistula • Tracheomalacia
• Tracheobronchomalacia • Esophageal leak • Esophageal strictures

KEY POINTS

- On diagnosis of EA/TEF, decompressing the esophageal pouch, minimizing positive pressure respiratory support, maintaining head of bed elevated, obtaining urgent echocardiogram, and preparing for surgical division of TEF with possible EA repair is essential.
- Various methods to achieve tension-induced esophageal lengthening exist but the ultimate approach to LGEA management should be tailored to the constellation of problems present in each individual patient.
- Incidence of tracheomalacia in patients with EA/TEF is high, for which patients benefit from preoperative dynamic tracheobronchoscopy and potential tracheopexy, if indicated. Separation of suture lines between the airway and esophagus helps reduce complications, such as recurrent or acquired TEF.
- Endoscopy has immense potential to assist in the management of esophageal leaks and strictures. Proactive endoscopic therapy may spare future esophageal stricture resections.
- Comprehensive longitudinal multidisciplinary care allows for durable patient outcomes in a highly complex subset of neonatal and pediatric patients.

INTRODUCTION

Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) (EA/TEF) is the most common anomaly of the esophagus (incidence 1/5000 births).[1] Due to significant advances in neonatal intensive care, anesthesia, nutrition, antimicrobial therapy, and surgical technique, survival associated with EA/TEF has improved to 91% to 98%,

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with mortality limited to cases involving coexisting life-threatening problems, such as extreme prematurity or complex congenital cardiac disease.[2–4] Because of improved survival, morbidity associated with EA/TEF warrants comprehensive, longitudinal multidisciplinary care.

Fig. 1 illustrates the most widely used EA/TEF classification scheme. Understanding the type of anomaly allows appropriate attention to relevant challenges and operative planning for the neonate.

DIAGNOSIS AND INITIAL MANAGEMENT

Although EA/TEF is usually diagnosed postnatally, it may be suspected on prenatal ultrasound or MRI (rates ranging from 16% to 36%).[5–7] Nonspecific signs, such as an absent or small stomach and polyhydramnios, raise prenatal suspicion. More specific findings include the presence of a blind-ending proximal esophageal pouch, which has been shown to have a high positive predictive value for EA and is seen in one-third of patients with prenatal diagnosis.[8,9] More recently, the presence of a dilated hypopharynx (DHP) has emerged as another sign suggesting EA.[10] Among 88 pregnant women who were evaluated prenatally for possible EA (of which 75 women had postnatal follow-up), DHP and/or dilated esophageal pouch was seen in 36% of those patients, 78% had postnatal EA diagnosis.[10] The authors of this study proposed an algorithm (Table 1) to predict EA risk based on a combination of prenatal findings. Accurately suspecting EA prenatally facilitates improved counseling regarding delivery plans, postnatal evaluation, and need for potential surgery; therefore, continued efforts to improve prenatal diagnosis remain essential.

Ultimately, EA/TEF remains a diagnosis made largely postnatally. Babies with prenatal suspicion or those with respiratory distress, increased oropharyngeal secretions, or feeding difficulties warrant further evaluation with attempt at passage of a naso/orogastric catheter. If unsuccessful, a plain radiograph is obtained to assess for coiling of the catheter in a blind ending upper esophagus (Fig. 2A). The presence of intestinal air suggests a TEF between the distal esophageal segment and the respiratory tract, consistent with a Gross Type C anomaly (Fig. 2B), whereas lack thereof corresponds to a Gross Type A anomaly (Fig. 2C).

On diagnosis, the proximal esophageal pouch is decompressed to avoid pooling of secretions or soiling of the respiratory tract from aspiration. Other important precautions include head of bed elevation, minimizing positive pressure respiratory support, and keeping the baby as calm as possible to avoid excessive swallowing of air. Although it may be tempting to acquire peripherally inserted central catheters in the Neonatal Intensive care unit in advance of anticipated surgery, we avoid this practice to minimize agitating the baby until the TEF is occluded or divided.

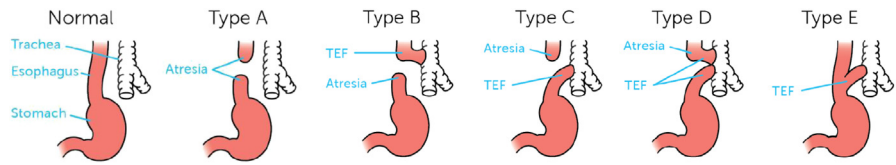


Fig. 1. Gross classification for EA/TEF. Type A: atresia only, no TEF (10% of cases); Type B: EA with TEF on proximal esophageal pouch (<1% of cases); Type C: EA with TEF on distal esophageal pouch (85% of cases); Type D: EA with TEFs on both upper and lower esophageal pouches (<1%); and Type E: TEF without EA (4%).

Table 1
Prediction algorithm for esophageal atresia based on presence or absence of primary and secondary signs

Condition	Percent Predicted to Have EA (95%)
No primary signs,[a] 1 or 0 secondary signs[b]	17 (7–35)
No primary signs,[a] Both secondary signs[b]	44 (22–69)
1 or both primary signs,[a] 1 or 0 secondary signs[b]	67 (42–85)
1 or both primary signs,[a] Both secondary signs[b]	89 (76–96)

[a] Primary signs: DHP, dilated proximal esophageal pouch.
[b] Secondary signs: polyhydramnios and small or absent stomach.
Adapted from Tracy S, Buchmiller TL, Ben-Ishay O, Barnewolt CE, Connolly SA, Zurakowski D, Phelps A, Estroff JA. The Distended Fetal Hypopharynx: A Sensitive and Novel Sign for the Prenatal Diagnosis of Esophageal Atresia. J Pediatr Surg. 2018 Jun;53(6):1137-1141.

EA/TEF is often associated with other anomalies, specifically of the VACTERL complex or CHARGE syndrome. Therefore, coordination of subsequent studies is essential, particularly an echocardiogram to assess for cardiac or vascular anomalies and to determine the aortic arch sidedness. These findings have significant anesthetic and surgical implications.

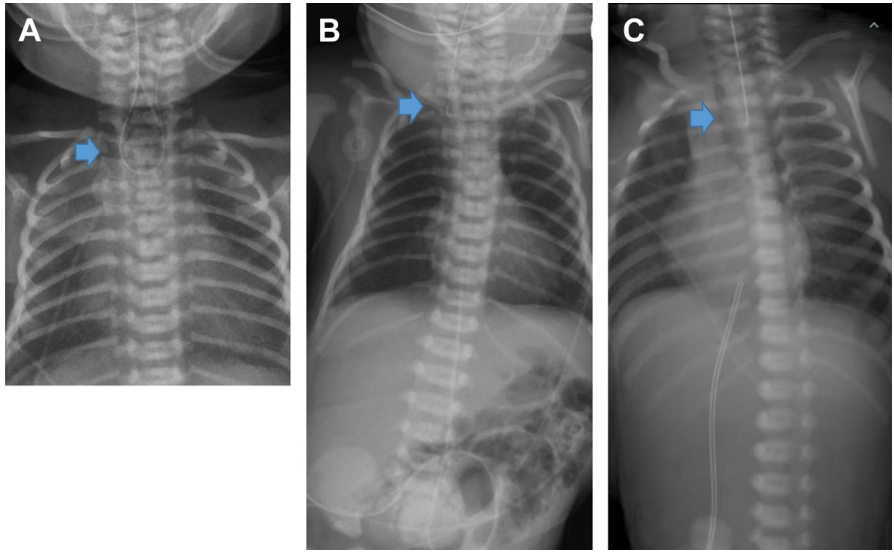


Fig. 2. Plain radiographs in newborns with EA. (*A*) Nasoesophageal tube coiled in atretic upper esophageal segment. (*B*) EA with distal TEF, as suggested by the presence of intraluminal intestinal air. (*C*) EA without distal TEF, as suggested by gasless abdomen. Arrows denotes nasoesophageal tube location.

SURGICAL REPAIR

We begin with rigid tracheobronchoscopy to aspirate airway secretions, assess associated tracheobronchomalacia (TBM), identify TEF location, and rule out any additional TEFs. After this, a Fogarty catheter is directed into the TEF with bronchoscopic visualization and then inflated to provide balloon occlusion. The patient is subsequently intubated over a rigid bronchoscope with the TEF in mind. If possible, the endotracheal tube (ETT) cuff is placed in the trachea beyond the TEF, thus providing an additional safety measure; not all TEFs are amenable to this strategy, however.

The anesthesia team places an arterial catheter. If the patient remains stable and successful temporizing control of the TEF (with balloon occlusion or cuffed ETT beyond TEF site) was achieved, we allow an attempt for central access. Otherwise, the case is performed with peripheral access only and central access is achieved on case conclusion. The patient's abdominal examination is monitored throughout. If the abdomen was distended preoperatively or if the patient would benefit from durable enteral access postoperatively (extremely premature patient, complex cardiac disease), then a gastrostomy is performed first with the gastric tube placed to water seal until the TEF can be surgically divided.

Depending on the aortic arch sidedness, a thoracotomy is performed. A thoracotomy is generally performed opposite the side of the aortic arch for improved exposure.We divide the azygous vein, except in cases where its caliber makes ligation prohibitive (eg, suspected interrupted inferior vena cava with azygous continuation). The distal esophagus is circumferentially dissected being mindful of the vagus nerves. A vessel loop is passed to control the TEF, at which point the balloon-based catheter is deflated and removed. The esophagus is dissected to its insertion into the airway, where the fistula is divided as flush with the airway as possible. Communication with the anesthesia team is essential because there is an open airway at this point.

The resultant tracheal wound is closed transversely using interrupted absorbable monofilament suture. This strategy results in essentially no tracheal diverticulum. If the patient's ETT can accommodate a flexible bronchoscope, the tracheal repair site is assessed endoscopically. Our practice has now evolved to include posterior tracheopexy at the time of newborn type C repair to separate tracheal and esophageal suture lines, support the otherwise-wide posterior membrane seen in EA/TEF patients, and preemptively address any future risk of dynamic airway collapse.

If the patient remains stable, we dissect the upper esophageal segment as far beyond the thoracic inlet as needed. The dissection is kept on the esophageal wall to minimize risk of injury to the recurrent laryngeal nerves (RLNs). We routinely use intraoperative nerve monitoring, even in newborns, to assess RLN function. Most newborn Type C EA/TEF procedures are amenable to primary esophagoesophageal anastomosis, which we perform with single-layer interrupted nonabsorbable monofilament sutures.

Once the anastomosis is complete, we perform a microvascular perfusion test using indocyanine green (ICG) SPY-PHY technology (Stryker, Kalamazoo, MI), even in newborns. A chest tube is placed. We do not use transanastomotic feeding tubes. The anesthesia team places a paravertebral catheter for analgesia. Postoperatively, patients are pharmacologically paralyzed for variable days based on the integrity, tension, blood supply, and overall assessment of the anastomosis. An esophagram is performed postoperative day 7 to 14, depending on the level of tension and overall clinical status. We have a proactive approach to esophageal stricture surveillance with most of our patients receiving endoscopy starting 1 month postoperatively if they are 3 kg or greater in size.

Indeed, many aspects of the procedure delineated above are innovative and evolve from within a high-volume referral-based practice encompassing various complex esophageal and airway disorders. Ultimately, surgeons and institutions should adapt the newborn EA/TEF repair in a manner that safely divides the fistula and adequately reconstructs the esophagus.

LONG GAP ESOPHAGEAL ATRESIA

The International Network of Esophageal Atresia defines long gap EA (LGEA) as "any esophageal atresia that lacks intra-abdominal air" or "all other types that technically prove difficult to repair."[11] This encompasses gap lengths ranging from one to many centimeters in length. Ultimately, LGEA management depends on various factors, such as surgeon experience, institutional resources, esophageal gap, associated airway symptoms, and more. Various techniques exist to achieve esophageal continuity. These include delayed primary anastomosis (after allowing natural growth), serial bougie dilation, esophageal myotomies, gastric pull-up, esophageal replacement, and tension-induced natural growth techniques (Foker process).

The authors' institution has been performing the Foker process since 2005, accumulating the world's largest experience. In a review comparing our historical cohort (2005–2013) of patients who underwent this technique to a more contemporary cohort (2014–2020), continued evolution of the procedure demonstrated improved outcomes, less morbidity, and increased esophageal preservation rates over time. Specifically, there were less leaks on traction, bone fractures, anastomotic leaks, or failed Foker procedures resulting in jejunal interposition in the contemporary cohort.[12] We also found that redo Foker procedures resulted in inferior outcomes compared with those initially performed at our institution.[12] This highlights the importance of appropriately planning the ideal LGEA operation or referring to a center with expertise when local resources and recently demonstrated experience are lacking.

With respect to our LGEA strategy, we comprehensively assess each patient and tailor the procedure to best address the constellation of problems present. This includes diagnostic laryngoscopy and rigid dynamic tracheobronchoscopy to assess for associated airway anomalies, such as laryngeal cleft, TBM, and TEF. We then perform contrast and endoscopic studies to assess the length and luminal quality of the upper and lower esophageal segments. A gap length is measured at rest and then again with pressure applied on each pouch (**Fig. 3**). These studies collectively guide operative strategy.

All patients undergo preoperative echocardiogram and contrast-enhanced chest computed tomography (CT) to evaluate aortic arch sidedness and great vessel anomalies. Preoperative vocal cord function is assessed with flexible nasolaryngoscopy (and is repeated postoperatively). Although we do not have an age threshold, we generally wait until size is 3.5 kg or greater before initiating surgical interventions for LGEA. In our experience, patients weighing less have had higher rate of traction system malfunction, including suture dislodgment.

At the time of LGEA repair, we explore the chest intending to achieve a primary anastomosis. We have a low threshold for cervical dissection to mobilize the upper esophagus. If a primary anastomosis cannot be achieved with acceptable tension and good tissue quality, the esophagus is placed on traction. If the patient has symptoms of severe TBM or bronchoscopy demonstrates greater than 50% tracheal collapse, a posterior tracheopexy is performed. For these patients, a right-sided approach is preferred in order to facilitate tracheobronchopexy.

If the patient has a short upper esophageal segment or a type B LGEA configuration, a cervical incision allows mobilization of the proximal esophagus and fistula division/

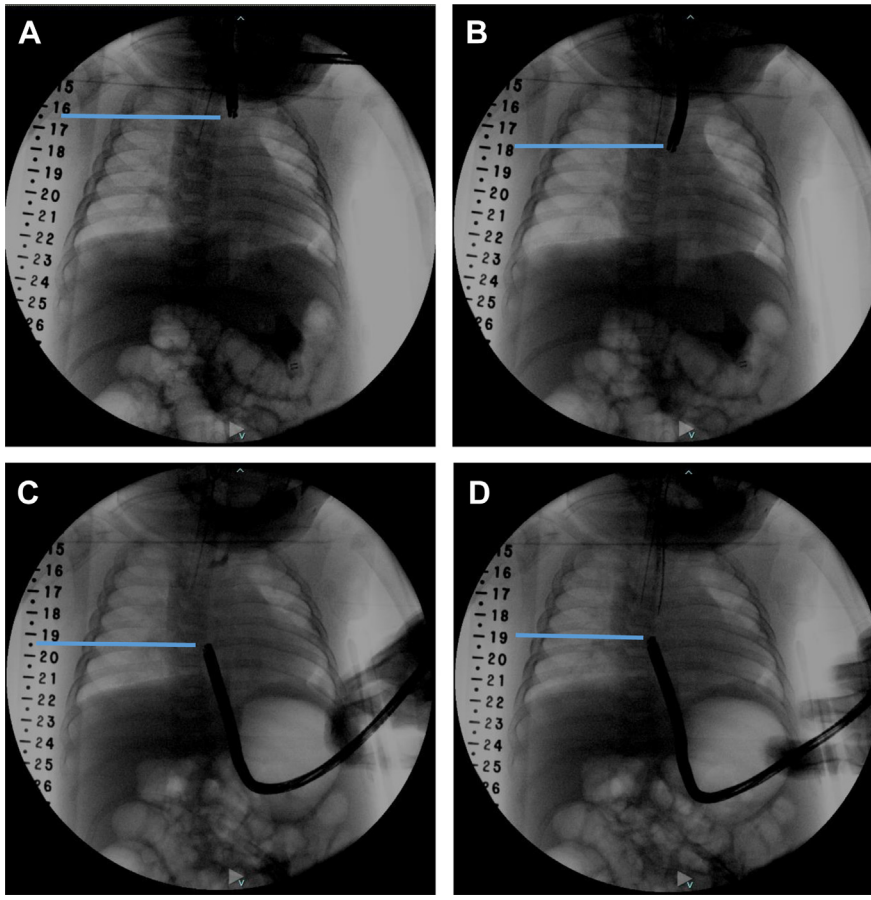


Fig. 3. Fluoroscopic assessment of esophageal gap length in LGEA. (*A*) Endoscope at the tip of upper esophageal pouch at rest and (*B*) with forward pressure. (*C*) Endoscope at tip of lower esophageal pouch at rest and (*D*) with forward pressure. In this patient, esophageal gap length at rest measured 3 cm and with forward pressure on each pouch reduced to 1 cm.

repair. Although others have approached proximal/cervical TEFs via minimally invasive surgery (MIS) (clipping/division strategies), we prefer the open approach. This allows full esophageal mobilization, identification/protection of RLNs, division of TEF as close to the airway with suture repair of the resultant tracheal wound, and placement of a silastic sleeve in the neck through which the esophagus passes thus minimizing its ability to adhere within the thoracic inlet.[12] The cervical incision does not preclude an MIS thoracic strategy, however. We consider MIS for patients without significant TBM and for those who have not previously had multiple thoracic operations. Either left-sided or right-sided MIS thoracic approach (regardless of sidedness of the aortic arch) is feasible. For example, patients with a large leftward upper esophageal pouch, minimal tracheomalacia, no TEF, and a history of previous right-sided surgeries could undergo a left-sided operation.[12]

The decision to pursue dynamic external versus static internal traction is based on esophageal gap length and patient comorbidities. If the gap is short, prolonged

postoperative paralysis would be detrimental to the patient, or there is a high likelihood the patient could be extubated between serial traction adjustments, we favor static internal traction. For longer gaps, external traction with frequent bedside adjustments to the traction system or internal traction via serial thoracoscopic traction adjustments are considered.[12] Neuromuscular paralysis is used throughout the external traction process. Ultimately, the operative strategy is customized to each patient. Regardless of strategy—external or internal, open or MIS—the median daily rate of esophageal growth measured radiographically is 1.1 mm per esophageal segment.[13]

Whenever possible, intraoperative endoscopy is used to guide traction suture placement to assure that no suture is full-thickness or intraluminal. We also place silastic sleeves around the esophageal pouches in order to minimize adhesion formation. **Fig. 4** demonstrates our external traction setup. For the MIS traction system, endoscopy also guides suture placement, which are placed in a bucket-handle configuration through which additional suture (typically fiberwire) is passed (**Fig. 5**). This is brought around a rib with the knot tied in the subcutaneous tissues to maintain traction. The tails are untied and the fiberwire retied under thoracoscopic visualization with each traction adjustment.

We have learned many lessons in the process of refining this procedure. Although at the start of our experience, traction adjustments were performed every other day, we have realized that tension-induced lengthening responds to less frequent adjustments, too. This led to the increased use of internal traction and MIS approaches over time.[12] Patients undergoing MIS approach also had reduced duration of neuromuscular paralysis, shorter intensive care unit and hospital lengths of stay, and no greater risk of complications.[12] For patients who undergo rescue Foker procedure after failed LGEA repair elsewhere, however, the hospital course remains longer and more complicated. Consequently, some patients with prior failed LGEA repairs are best served with esophageal replacement instead of rescue lengthening procedures. The jejunal interposition (JI) is our preferred replacement when the native esophagus cannot be reconstructed but this operation is not performed in the neonatal period (deferred until patients are >10 kg in size).

ASSESSMENT OF ANASTOMOTIC INTEGRITY

Due to our institution's vast experience with complex esophageal and airway disorders, we now systematically evaluate every esophageal anastomosis with respect to blood supply, tension, and tissue quality because they each contribute to healing. Microvascular perfusion is assessed with ICG SPY-PHY technology, looking at speed and intensity of perfusion, and degree of hypoperfusion near the anastomosis. We note whether the anastomosis is in a reoperative field or involves a prior failed anastomosis. We gauge degree of tension by overall appearance (eg, sutures pulling through) and how much effort was required to achieve the anastomosis (eg, putting patient in flexed position). Altogether, our assessment guides postoperative management in that there is a low threshold to pharmacologically paralyze a patient if the anastomosis is on severe tension or involves poor tissue quality. This allows initial anastomotic healing without active swallowing or excessive movement and cervical extension. We have observed higher anastomotic complication rates in settings of poor integrity due to impaired perfusion, high tension, or poor tissue quality. Ongoing research is necessary to continue understanding factors that influence anastomotic outcomes to further refine operative technique and postoperative management.

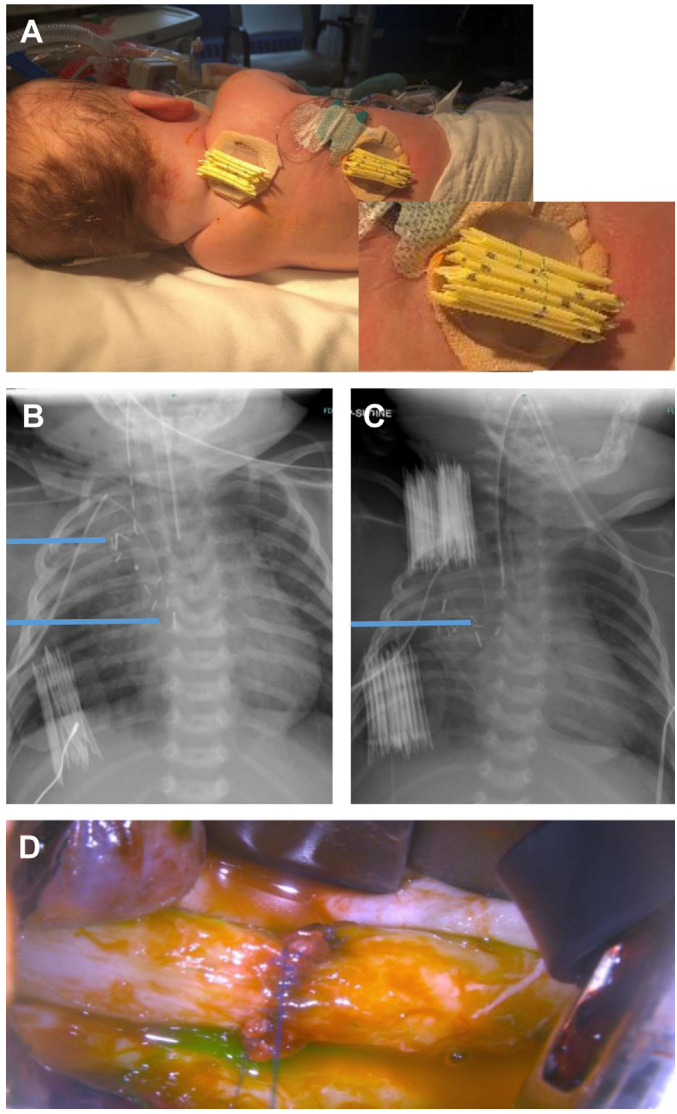


Fig. 4. External traction process for LGEA. (*A*) Traction sutures are brought out through the skin and tied to a silicone disk. Tension is transmitted to the esophageal segments by adding feeding tube fragments under the sutures. (*B*) Radiopaque clips on the traction system and the esophageal wall are tracked on plain radiographs until (*C*) the clips cluster together at which point esophago-esophageal anastomosis (*D*) is performed.

MANAGEMENT OF ESOPHAGEAL LEAKS AND STRICTURES

Although not standard practice within the field, we perform endoscopy on EA/TEF patients around 1 month postoperatively (if patients are >3 kg in size). Most LGEA patients receive a series of 3 planned dilations with intralesional steroid injections (ISI). Thereafter, stricture response to therapy and patient symptoms guide additional interventions, which typically include endoscopic incisional therapy (EIT) and/or stenting.

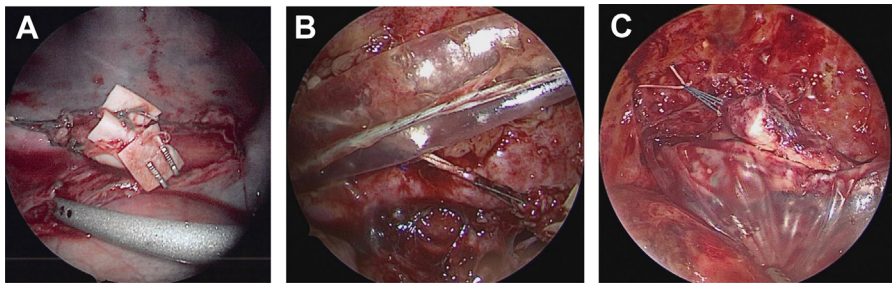


Fig. 5. Thoracoscopic internal traction process for LGEA. (*A*) Traction system sutures are placed on the esophageal pouches in mattress fashion with pericardial pledgets marked by radiopaque clips. Through the bucket-handle configuration, additional suture is passed. (*B, C*) Serial adjustments involve untying the knot and retying it under thoracoscopic visualization to set a new level of tension. In doing so, the esophageal pouches traverse the thoracic cavity.

We consider surgical resection for strictures refractory to endoscopic interventions. The point at which endoscopic therapy should be abandoned in favor of resection is unclear. In a retrospective study of 171 repaired EA patients who underwent serial endoscopies, factors associated with eventual need for stricture resection were discerned. The probability of remaining free from stricture resection decreased with increasing number of therapeutic endoscopies. A cutoff of 7 endoscopies discriminated between patients who needed stricture resection and those who did not.[14] Despite this, most patients remained free of stricture resection well beyond 7 therapeutic endoscopies.[14] Other predictors for needing stricture resection included esophageal leak, initial anastomotic diameter less than 3 mm, and need for advanced therapeutic endoscopic maneuvers (such as ISI, EIT, stenting).[15] Ultimately, we thought that proactive therapeutic endoscopies may spare stricture resections.

Patients who have undergone EA/TEF repair have a reported anastomotic leak rate of up to 38%.[16,17] Our institution routinely uses negative pressure wound therapy (also known as vacuum-assisted closure [VAC] therapy) for endoscopic management of esophageal leaks. This highly effective method helps close leak cavities and promote wound healing by stimulating angiogenesis, removing excess debris, and allowing granulation tissue formation.[18,19] Early experience with endoscopic VAC (e-VAC) therapy demonstrates technical feasibility, safety, and effectiveness in managing esophageal anastomotic leaks.[18] Additional centers have also reported on use of e-VAC therapy for esophageal leaks.[20–22] When institutional experience with advanced endoscopy is lacking, antibiotics and chest drainage or operative washout with tacking of esophageal segments to the prevertebral fascia should be considered.

MANAGEMENT OF RECURRENT OR ACQUIRED TRACHEOESOPHAGEAL FISTULA

Recurrent TEF (recTEF) complicates 5% to 10% of EA/TEF repairs.[23–27] Postoperative acquired TEF (acqTEF) can occur in addition to or even in the absence of prior congenital TEFs in the setting of esophageal anastomotic complications. These TEF variants rarely close spontaneously, and given the perceived high risks of operative intervention, they are often first approached endoscopically with reported re-recurrence rates approaching 63%.[23,28–31] Surgical techniques to address rec/acqTEFs include placing autologous tissue (pleural or muscle flaps) or prosthetic material (mesh) between the repair. Re-recurrence rates for these cases, although better than

endoscopic interventions, still range from 11% to 22%.[23,28,29] At our institution, posterior tracheopexy and rotational esophagoplasty is the backbone of surgical repair of rec/acqTEFs in order to completely separate suture lines without the need for interposing tissue.[32,33] Our published experience involving 62 patients who underwent rec-TEF repair reports 0 re-recurrences during a median follow-up of 2.5 years.[33] We prefer an upfront surgical approach as we think repeated endoscopies can be futile and/or harmful and surgery allows for addressing coexisting TBM, strictures, or other intrathoracic pathologic conditions.

TRACHEOBRONCHOMALACIA

Tracheomalacia is the most common congenital tracheobronchial anomaly (incidence 1/2100 children).[34] It refers to excessive compliance of the trachea, predisposing it to static or dynamic collapse. If the mainstem bronchi are also involved, TBM results. Tracheomalacia is common among EA/TEF patients (incidence of 10%–75%) due to shared embryologic origins of the trachea and esophagus.[35–38] Signs suggesting tracheomalacia include chronic barky cough, noisy breathing, exercise intolerance, expiratory stridor, frequent or more severe respiratory illnesses, feeding difficulties, development of bronchiectasis, or acute life-threatening events (ALTEs), including brief resolving unexplained events.

A detailed endoscopic airway assessment is essential to diagnosing TBM. The normal trachea and bronchi consist of C-shaped cartilages with a narrow posterior membrane that intrudes during cough without compromising overall airway patency. In TBM, malformed cartilages are often U or bow-shaped with a wider more pliable membrane that lends itself to significant posterior intrusion, thereby collapsing the airway lumen. This leads to impaired clearance of pulmonary secretions, ineffective cough, and insufficient air movement.

We perform rigid dynamic 3-phase tracheobronchoscopy to assess structure and function of the visible airways.[39] The first phase of assessment involves a spontaneously breathing patient. Anatomy is characterized with respect to tracheobronchial tree branching pattern, cartilage shape, posterior membrane intrusion at rest, and any fixed airway compression. Next, sedation is titrated to allow more vigorous breathing and cough. The degree of dynamic airway compression, particularly the extent of posterior membrane intrusion or anterior intrusion, is gauged at each portion of the visible airway, if possible. Finally, anesthesia provides additional sedation, and the airway is distended to inspect for tracheal diverticulums, occult TEFs, aberrant bronchi, and other abnormalities. Lesions suspicious for TEFs are probed with a catheter for passage into a tract or contrast is instilled to delineate communication with the esophagus. This assessment can also be performed with flexible bronchoscopy to evaluate small airway collapse in premature infants, those with bronchopulmonary dysplasia, or older patients for whom the rigid bronchoscopes are not long enough.

The combination of symptoms with abnormal tracheobronchoscopy supports further intervention. In our experience, children with symptomatic TBM often demonstrate greater than 75% narrowing of the airway during forced exhalation or coughing.[38,40] However, bronchoscopic findings must correlate with concerning symptoms to warrant the risks of any proposed interventions.

MANAGEMENT OF TRACHEOBRONCHOMALACIA

Many think that children with TBM will outgrow their symptoms but this is a common misconception. Although milder cases may become less symptomatic because the airway diameter enlarges with the child's growth, TBM will not simply resolve on its

own and, in fact, can exacerbate with age. A graduated approach to management is therefore essential. We begin with medical management. Optimizing mucociliary clearance involves decreasing quantity of secretions without thickening them (using ipratropium bromide [atrovent]) and loosening secretions (with normal saline or hypertonic saline nebulizers). Chest physiotherapy enhances clearance. Low-dose inhaled corticosteroids help reduce mucosal inflammation but should be used cautiously to minimize any negative effects on cartilage development. Early initiation of antibiotics during an active infection is also part of our strategy to decrease severity and length of symptoms. Patients with documented severe collapse on tracheobronchoscopy and failure of maximal medical management are considered operative candidates.

Surgical management of TBM depends on the type and location of disease and the airway's relationship to major blood vessels and the esophagus. For this reason, preoperative evaluation also includes multidetector CT (MDCT) with 3-dimensional reconstruction.

Historically, TBM was addressed with anterior aortopexy. This entails sternotomy for thymectomy, after which the innominate artery and ascending aorta or aortic arch are pulled anteriorly by suturing it to the posterior aspect of the sternum, thereby relieving anterior airway compression. Because the vessels remain attached to the airway through areolar tissue, pulling the vessels anteriorly effectively opens the airway as well. This strategy, however, does not address posterior membranous tracheal intrusion, which is the more common finding seen in patients with a history of EA/TEF.

In this patient population, we begin with posterior tracheopexy. This involves a right thoracotomy for esophageal mobilization, being mindful of the vagus nerves, the RLNs, and the thoracic duct. The aorta is mobilized if a descending aortopexy is anticipated. If a recTEF or tracheal diverticulum from prior TEF repair is noted, it is corrected by dividing and repairing the TEF or resecting the tracheal diverticulum flush with the tracheal wall under bronchoscopic visualization. The resultant tracheal defect is primarily closed using interrupted absorbable monofilament suture. An air-leak test is performed in coordination with our anesthesiologists.

After this, posterior tracheopexy is performed by passing autologous-pledgeted polypropylene sutures into, but not through, the posterior membrane of the trachea using bronchoscopic guidance to assure no bite is full thickness. These bites are mattress fashion and taken to the anterior longitudinal spinal ligament, thus securing the posterior membrane there. All stitches are placed first and then sequentially tied with no retractors in place. A negative-pressure suction test is typically performed after the completion of tracheopexy to assess for residual posterior intrusion and airway patency. The end result is opening of the airway and rotation of the esophagus laterally such that variations in luminal size with feeding no longer cause intrusion into the posterior tracheal membrane.

If a descending aortopexy is indicated, it is performed before posterior tracheopexy. We do this procedure if the descending aorta is located too far anteriorly on cross-sectional imaging such that the midportion of the left mainstem bronchus is trapped between the descending aorta and the pulmonary artery, resulting in narrowing of the bronchus.[41,42] Identification of the Artery of Adamkiewicz during the MDCT guides the surgeon in avoiding injury to this artery during the case.

Long-term results of posterior tracheopexy (**Fig. 6**) for severe TBM were evaluated for 98 consecutive patients at our institution; with a 5-month follow-up period, we reported improvements in clinical symptoms, including chronic cough, noisy breathing, prolonged and recurrent respiratory infections, transient respiratory distress requiring

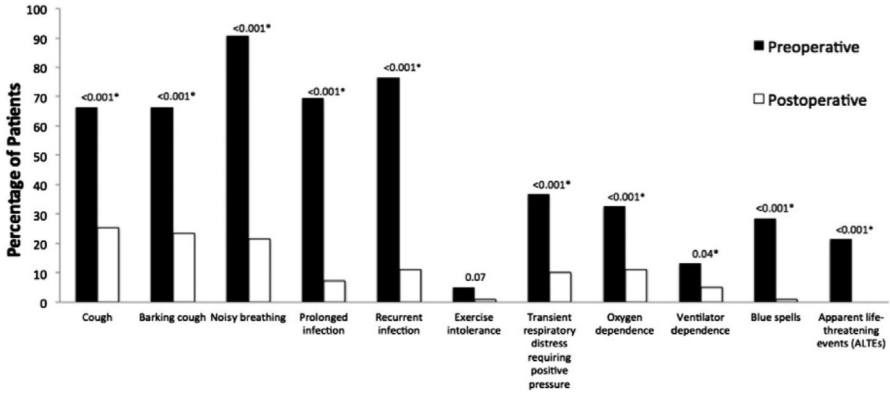
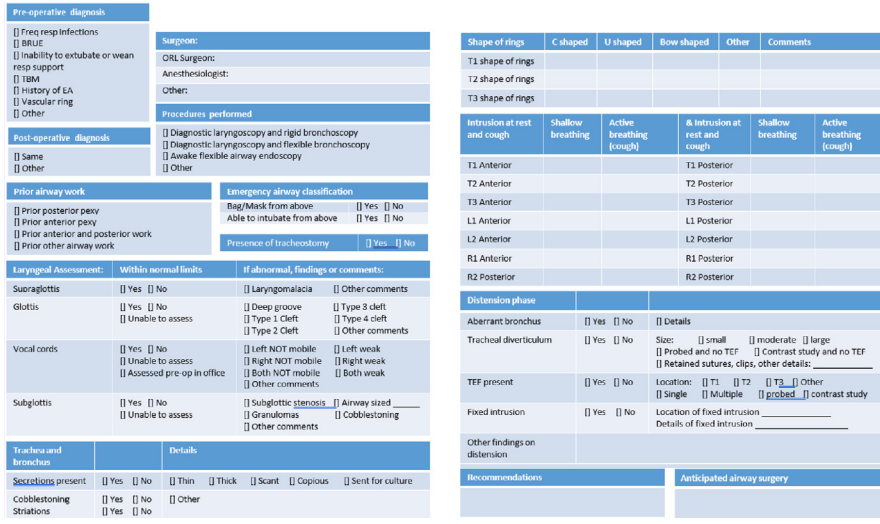


Fig. 6. Preoperative and postoperative clinical symptoms with relation to posterior tracheopexy. There were statistically significant improvements in clinical symptoms after posterior tracheopexy, including report of barking cough, noisy breathing, prolonged and recurrent respiratory infections, need for positive pressure, oxygen dependence, blue spells, and ALTEs. *statistical significance. (*From S*hieh HF, Smithers CJ, Hamilton TE, Zurakowski D, Rhein LM, Manfredi MA, Baird CW, Jennings RW. Posterior tracheopexy for severe tracheomalacia. J Pediatr Surg. 2017 Jun;52(6):951-955.)

positive pressure, oxygen dependence, blue spells, ALTEs, and ventilator dependence (**Fig. 7**).[37] Only 9% of patients had persistent symptomatic tracheomalacia requiring reoperation (in the form of anterior aortopexy).[37]

Most patients with EA/TEF will likely not need surgical management for tracheomalacia but if such interventions are being considered, patients should be referred to institutions with experience performing the procedures described above.



Pre-operative diagnosis

[] Freq resp infections
[] BRUE
[] Inability to extubate or wean resp support
[] TBM
[] History of EA
[] Vascular ring
[] Other

Surgeon:

ORL Surgeon:
Anesthesiologist:
Other:

Post-operative diagnosis

[] Same
[] Other

Procedures performed

[] Diagnostic laryngoscopy and rigid bronchoscopy
[] Diagnostic laryngoscopy and flexible bronchoscopy
[] Awake flexible airway endoscopy
[] Other

Prior airway work

[] Prior posterior pexy
[] Prior anterior pexy
[] Prior anterior and posterior work
[] Prior other airway work

Emergency airway classification

Bag/Mask from above	[] Yes [] No
Able to intubate from above	[] Yes [] No

Presence of tracheostomy [] Yes [] No

Laryngeal Assessment:	Within normal limits	If abnormal, findings or comments:	
Supraglottis	[] Yes [] No	[] Laryngomalacia	[] Other comments
Glottis	[] Yes [] No [] Unable to assess	[] Deep groove [] Type 1 Cleft [] Type 2 Cleft	[] Type 3 cleft [] Type 4 cleft [] Other comments
Vocal cords	[] Yes [] No [] Unable to assess [] Assessed pre-op in office	[] Left NOT mobile [] Right NOT mobile [] Both NOT mobile [] Other comments	[] Left weak [] Right weak [] Both weak
Subglottis	[] Yes [] No [] Unable to assess	[] Subglottic stenosis [] Granulomas [] Other comments	[] Airway sized ____ [] Cobblestoning

Trachea and bronchus		Details				
Secretions present	[] Yes [] No	[] Thin	[] Thick	[] Scant	[] Copious	[] Sent for culture
Cobblestoning	[] Yes [] No	[] Other				
Striations	[] Yes [] No					

Shape of rings	C shaped	U shaped	Bow shaped	Other	Comments
T1 shape of rings					
T2 shape of rings					
T3 shape of rings					

Intrusion at rest and cough	Shallow breathing	Active breathing (cough)	& Intrusion at rest and cough	Shallow breathing	Active breathing (cough)
T1 Anterior			T1 Posterior		
T2 Anterior			T2 Posterior		
T3 Anterior			T3 Posterior		
L1 Anterior			L1 Posterior		
L2 Anterior			L2 Posterior		
R1 Anterior			R1 Posterior		
R2 Posterior			R2 Posterior		

Distension phase		
Aberrant bronchus	[] Yes [] No	[] Details
Tracheal diverticulum	[] Yes [] No	Size: [] small [] moderate [] large [] Probed and no TEF [] Contrast study and no TEF [] Retained sutures, clips, other details: _____
TEF present	[] Yes [] No	Location: [] T1 [] T2 [] T3 [] Other [] Single [] Multiple [] probed [] contrast study
Fixed intrusion	[] Yes [] No	Location of fixed intrusion _____ Details of fixed intrusion _____
Other findings on distension		

Recommendations	Anticipated airway surgery

Fig. 7. Standardized airway evaluation. This form is used in a systematic fashion for airway evaluation in our patients.

LONG-TERM MULTIDISCIPLINARY CARE AND MANAGEMENT

Of utmost importance in the care of patients with complex esophageal and airway disorders is a comprehensive and multidisciplinary team-based mentality. Patients treated by our institution are followed longitudinally across numerous disciplines, including but not limited to general surgery, gastroenterology, pulmonology, otolaryngology, speech language pathology, nutrition, social work, anesthesia, cardiac surgery, plastic surgery, and orthopedic surgery. It is the multidisciplinary team's tireless efforts that has yielded durable success in the surgical and nonsurgical management of this highly complicated subset of neonatal and pediatric patients.

REFERENCES

1. Sfeir R, Bonnard A, Khen-Dunlop N, et al. Esophageal atresia: data from a national cohort. J Pediatr Surg 2013;48(8):1664–9.
2. Pinheiro PF, Simoes e Silva AC, Pereira RM. Current knowledge on esophageal atresia. World J Gastroenterol 2012;18(28):3662–72.
3. Goyal A, Jones MO, Couriel JM, et al. Oesophageal atresia and tracheo-oesophageal fistula. Arch Dis Child Fetal Neonatal 2006;91(5):F381–4.
4. Gupta DK, Sharma S. Esophageal atresia: the total care in a high-risk population. Semin Pediatr Surg 2008;17(4):236–43.
5. Bradshaw CJ, Thakkar H, Knutzen L, et al. Accuracy of prenatal detection of tracheoesophageal fistula and oesophageal atresia. J Pediatr Surg 2016;51(8): 1268–72.
6. Fallon SC, Ethun CG, Olutoye OO, et al. Comparing characteristics and outcomes in infants with prenatal and postnatal diagnosis of esophageal atresia. J Surg Res 2014;190(1):242–5.
7. Kunisaki SM, Bruch SW, Hirschl RB, et al. The diagnosis of fetal esophageal atresia and its implications on perinatal outcome. Pediatr Surg Int 2014;30(10): 971–7.
8. Garabedian CSR, Langlois C, Bonnard A, et al. Does prenatal diagnosis modify neonatal treatment and early outcome of children with esophageal atresia? Am J Obstet Gynecol 2015;212(3):340–7.
9. Ethun CG, Fallon SC, Cassady CI, et al. Fetal MRI improves diagnostic accuracy in patients referred to a fetal center for suspected esophageal atresia. J Pediatr Surg 2014;49(5):712–5.
10. Tracy S, Buchmiller TL, Ben-Ishay O, et al. The distended fetal hypopharynx: a Sensitive and Novel sign for the prenatal diagnosis of esophageal atresia. J Pediatr Surg 2018;53(6):1137–41.
11. van der Zee DCBP, Faure C. Position paper of INoEA working group on long-gap esophageal atresia: for better care. Front Pediatr 2017;5:1–3.
12. Svetanoff WJ, Zendejas B, Hernandez K, et al. Contemporary outcomes of the Foker process and evolution of treatment algorithms for long-gap esophageal atresia. J Pediatr Surg 2021;56(12):2180–91.
13. Foust AM, Zendejas B, Mohammed S, et al. Radiographic assessment of traction-induced esophageal growth and traction-related complications of the Foker process for treatment of long-gap esophageal atresia. Pediatr Radiol 2021. https://doi.org/10.1007/s00247-021-05228-z.
14. Yasuda JL, Taslitsky GN, Staffa SJ, et al. Utility of repeated therapeutic endoscopies for pediatric esophageal anastomotic strictures. Dis Esophagus 2020; 33(12). https://doi.org/10.1093/dote/doaa031.

15. Baghdadi O, Clark S, Ngo P, et al. Initial esophageal anastomosis diameter predicts treatment outcomes in esophageal atresia patients with a high risk for stricture development. Front Pediatr 2021;9:710363.
16. Liu J, Yang Y, Zheng C, et al. Surgical outcomes of different approaches to esophageal replacement in long-gap esophageal atresia: a systematic review. Medicine (Baltimore) 2017;96(21):e6942.
17. Lal DR, Gadepalli SK, Downard CD, et al. Perioperative management and outcomes of esophageal atresia and tracheoesophageal fistula. J Pediatr Surg 2017;52(8):1245–51.
18. Manfredi MA, Clark SJ, Staffa SJ, et al. Endoscopic esophageal vacuum therapy: a Novel therapy for esophageal Perforations in pediatric patients. J Pediatr Gastroenterol Nutr 2018;67(6):706–12.
19. Yasuda JL, Svetanoff WJ, Staffa SJ, et al. Prophylactic negative vacuum therapy of high-risk esophageal anastomoses in pediatric patients. J Pediatr Surg 2021;56(5):944–50.
20. Heits N, Stapel L, Reichert B, et al. Endoscopic endoluminal vacuum therapy in esophageal perforation. Ann Thorac Surg 2014;97(3):1029–35.
21. Loske G, Schorsch T, Muller C. Intraluminal and intracavitary vacuum therapy for esophageal leakage: a new endoscopic minimally invasive approach. Endoscopy 2011;43(6):540–4.
22. Schorsch T, Muller C, Loske G. Endoscopic vacuum therapy of anastomotic leakage and iatrogenic perforation in the esophagus. Surg Endosc 2013;27(6):2040–5.
23. Lal DR, Oldham KT. Recurrent tracheoesophageal fistula. Eur J Pediatr Surg 2013;23(3):214–8.
24. Coran AG. Redo esophageal surgery: the diagnosis and management of recurrent tracheoesophageal fistula. Pediatr Surg Int 2013;29(10):995–9.
25. Koivusalo AI, Pakarinen MP, Lindahl HG, et al. Revisional surgery for recurrent tracheoesophageal fistula and anastomotic complications after repair of esophageal atresia in 258 infants. J Pediatr Surg 2015;50(2):250–4.
26. Kovesi T, Rubin S. Long-term complications of congenital esophageal atresia and/or tracheoesophageal fistula. Chest 2004;126(3):915–25.
27. Ein SH, Stringer DA, Stephens CA, et al. Recurrent tracheoesophageal fistulas seventeen-year review. J Pediatr Surg 1983;18(4):436–41.
28. Aworanti O, Awadalla S. Management of recurrent tracheoesophageal fistulas: a systematic review. Eur J Pediatr Surg 2014;24(5):365–75.
29. Daniel SJ, Smith MM. Tracheoesophageal fistula: open versus endoscopic repair. Curr Opin Otolaryngol Head Neck Surg 2016;24(6):510–5.
30. Meier JD, Sulman CG, Almond PS, et al. Endoscopic management of recurrent congenital tracheoesophageal fistula: a review of techniques and results. Int J Pediatr Otorhinolaryngol 2007;71(5):691–7.
31. Richter GT, Ryckman F, Brown RL, et al. Endoscopic management of recurrent tracheoesophageal fistula. J Pediatr Surg 2008;43(1):238–45.
32. Smithers CJ, Hamilton TE, Manfredi MA, et al. Categorization and repair of recurrent and acquired tracheoesophageal fistulae occurring after esophageal atresia repair. J Pediatr Surg 2017;52(3):424–30.
33. Kamran A, Zendejas B, Meisner J, et al. Effect of posterior tracheopexy on risk of recurrence in children after recurrent tracheo-esophageal fistula repair. J Am Coll Surg 2021;232(5):690–8.

34. Boogaard R, Huijsmans SH, Pijnenburg MW, et al. Tracheomalacia and bronchomalacia in children: incidence and patient characteristics. Chest 2005;128(5):3391–7.
35. Spitz L, Kiely E, Brereton RJ. Esophageal atresia: five year experience with 148 cases. J Pediatr Surg 1987;22(2):103–8.
36. Filler RM, Messineo A, Vinograd I. Severe tracheomalacia associated with esophageal atresia: results of surgical treatment. J Pediatr Surg 1992;27(8):1136–40 [discussion: 40–1].
37. Shieh HF, Smithers CJ, Hamilton TE, et al. Posterior tracheopexy for severe tracheomalacia. J Pediatr Surg 2017;52(6):951–5.
38. Fraga JC, Jennings RW, Kim PC. Pediatric tracheomalacia. Semin Pediatr Surg 2016;25(3):156–64.
39. Kamran A, Jennings RW. Tracheomalacia and tracheobronchomalacia in pediatrics: an Overview of evaluation, medical management, and surgical treatment. Front Pediatr 2019;7:512.
40. Choi S, Lawlor C, Rahbar R, et al. Diagnosis, classification, and management of pediatric tracheobronchomalacia: a review. JAMA Otolaryngol Head Neck Surg 2019;145(3):265–75.
41. Shieh HF, Smithers CJ, Hamilton TE, et al. Descending aortopexy and posterior tracheopexy for severe tracheomalacia and left mainstem bronchomalacia. Semin Thorac Cardiovasc Surg 2019;31(3):479–85.
42. Svetanoff WJ, Zendejas B, Frain L, et al. When to consider a posterolateral descending aortopexy in addition to a posterior tracheopexy for the surgical treatment of symptomatic tracheobronchomalacia. J Pediatr Surg 2020;55(12):2682–9.