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Evolution of left-sided thoracoscopic approach for long gap esophageal atresia repair

Hester F. Shieh [a,*], Thomas E. Hamilton [b], Michael A. Manfredi [c], Peter D. Ngo [c], Michael J. Wilsey [d], Jessica L. Yasuda [c], Benjamin Zendejas [b], C. Jason Smithers [a]

[a] Department of Surgery, Johns Hopkins All Children's Hospital, 501 6th Ave S, St. Petersburg, FL 33701, United States
[b] Department of Surgery, Boston Children's Hospital, 300 Longwood Ave, Boston, MA 02115, United States
[c] Department of Gastroenterology, Boston Children's Hospital, 300 Longwood Ave, Boston, MA 02115, United States
[d] Department of Gastroenterology, Johns Hopkins All Children's Hospital, 501 6th Ave S, St. Petersburg, FL 33701, United States

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Background: Left-sided repair for long gap esophageal atresia (LGEA) has been described for patients with a large leftward upper pouch, no thoracic tracheoesophageal fistula (TEF) nor tracheobronchomalacia (TBM), or as salvage plan after prior failed right-sided repair. We describe our experience with left-sided MIS traction induced growth process.
Methods: We retrospectively reviewed patients who underwent Foker process for LGEA at two institutions between December 2016 and November 2021. Patient characteristics, surgical techniques, and outcomes were reviewed.
Results: 71 patients underwent Foker process. Of 34 MIS cases, 28 patients (82%) underwent left-sided repair (median gap length 5 cm) at median age 4 months with median 3 (range 2–8) operations and median 13.5 (IQR 11–21) days on traction until esophageal anastomosis. 9 patients (32%) underwent completely MIS approach, whereas 5 patients (18%) converted to open at first operation and 14 patients (50%) converted to open later in the traction process. Traction was internal in 68%, external in 11%, and combination in 21%. Median follow-up was 15.4 (IQR 7.5–31.7) months after anastomosis. 14% had anastomotic leak managed with antibiotics and/or esophageal vacuum therapy. Median number of esophageal dilations was 3.5 (range 0–13). 18% required stricture resection. 39% underwent Nissen fundoplication. None have needed esophageal replacement.
Conclusions: For multiple reasons including the tendency of both esophageal pouches to have a leftward bias, less tracheal compression by upper pouch, and clean field of surgery for reoperative cases, we now more commonly use left-sided approach for MIS LGEA repair compared to right side, regardless of left aortic arch.
Level of evidence: Level IV Treatment Study.

1. Introduction

Long gap esophageal atresia (LGEA) describes a technically challenging subset of esophageal atresia (EA) cases, in which a primary anastomosis of the two ends of the esophagus cannot be performed under acceptable tension by the operating surgeon at the initial operation. There is limited consensus on its optimal treatment strategy, and surgical approach is often based on institutional experience [1,2]. However, most agree that the native esophagus is the preferred conduit for esophageal reconstruction. The Foker process utilizes staged tension induced growth for esophageal lengthening to preserve the native esophagus and achieve a primary repair. These techniques have evolved from open thoracotomy and external traction to include options for minimally invasive (MIS) thoracoscopy, internal traction, and left or right sided repair [3,4].

Historically, EA repair is approached from the right side in the majority of patients with a left sided aortic arch, with left-sided repair reserved for those with a right sided aortic arch. For LGEA, left-sided repair, regardless of aortic arch position, has been described by our group for patients with a large leftward upper

Abbreviations: LGEA, long gap esophageal atresia; EA, esophageal atresia; TEF, tracheoesophageal fistula; TBM, tracheobronchomalacia.

* Corresponding author at: Department of Surgery, Johns Hopkins All Children's Hospital, 501 6th Ave S, St. Petersburg, FL 33701, United States. Fax: +1 727 767-3295.

E-mail address: hshieh1@jhmi.edu (H.F. Shieh).

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pouch, no thoracic tracheoesophageal fistula (TEF) nor tracheobronchomalacia (TBM), or as a salvage plan after prior failed right-sided repair, with similar outcomes to right-sided repair [5]. The left-sided approach has evolved into our most commonly used MIS strategy, as has the use of internal traction. We describe our experience with left-sided MIS traction induced growth process for LGEA repair.

2. Methods

We retrospectively reviewed all patients who underwent Foker process traction induced growth and repair for LGEA at Boston Children's Hospital and Johns Hopkins All Children's Hospital between December 2016 and November 2021. This time period reflects when our operative approach transitioned to include the option of left sided repairs. These two institutions have multidisciplinary Esophageal and Airway Treatment (EAT) centers with similar approaches to management and operative strategies. Patient characteristics, surgical techniques, complications, and post-operative outcomes were reviewed.

Preoperative assessment included an echocardiogram and chest computed tomography (CT) to assess for structural heart disease, the side of the aortic arch, great vessel anomalies, and anatomic relationship of the mediastinal structures. Endoscopic airway and esophageal evaluation were performed by the multidisciplinary team and operating surgeons. Diagnostic laryngoscopy and bronchoscopy (DLB) were done under general anesthesia in spontaneously breathing patients. After assessment of supraglottic structures, vocal cord function, and the larynx for presence of a laryngeal cleft, a rigid ventilating bronchoscope was inserted through the cords to assess for TEF, tracheal diverticulum, cartilage shape, and degree and location of tracheobronchomalacia with anterior compression or posterior intrusion. Dynamic bronchoscopy was done in three phases: normal spontaneous breathing, lightened sedation to allow for agitation and coughing, and deepened sedation to allow for positive pressure airway distension. The upper and lower esophageal pouches were then assessed by contrast studies through an orogastric tube and a tube passed retrograde through the gastrostomy site. The gap between the two esophageal segments was measured off tension with contrast injection and on tension with Bakes dilators or an endoscope placing pressure on the esophageal pouches. The operating surgeons determined the operative plan and approach based on preoperative imaging and endoscopic evaluation.

Left-sided MIS repairs were considered for patients with a large leftward upper esophageal pouch, no thoracic TBM, and no thoracic TEF or a thoracic TEF that had previously been divided and repaired. History of any prior failed right-sided repair was noted, as a left-sided approach in this setting would be in a clean operative field. Isolated cervical TBM from the upper pouch compressing the trachea, without significant thoracic TBM, was considered for a left-sided approach with the addition of cervical posterior tracheopexy through a left neck dissection.

Our left-sided MIS approach has evolved during this time period and we describe our current approach. Recurrent laryngeal nerve monitoring is done with modified Dragonfly electrodes on the endotracheal tube and an APS nerve monitor placed around the main vagus nerve when a neck dissection is done. The patient is placed in a right lateral decubitus position, with the left chest, neck, and arm prepped into the field. A left neck dissection is usually done to fully mobilize the proximal esophageal pouch and divide its common wall attachments to the trachea, identify and preserve the recurrent laryngeal nerves, divide and repair a cervical TEF if present, and perform a cervical posterior tracheopexy if there is a cervical TEF or severe cervical TBM. The esophageal traction sutures are placed in the upper esophageal pouch in an open fashion. We typically use one double-armed 3–0 Ethibond with bovine pericardial pledgets to place a horizontal mattress suture on each side of the tip, such that it creates a double loop “bucket handle”. The traction suture is placed under endoscopic guidance to ensure bites of the muscle and submucosa that are not full thickness. Small clips are used to mark the two pledgets and a medium clip is sutured on the esophageal wall behind the pledgets as a trailing clip. A 2-0 silk suture is passed through the double loop bucket handle to help transfer it later into the chest and act as the pulley suture. We place a silastic sleeve around the upper esophageal pouch and secure it at the superior portion of the pouch dissection with interrupted 6–0 Prolene sutures to prevent scarring at the thoracic inlet. A window is started in front of the spine going into the thoracic inlet to deliver the upper esophageal pouch into the chest.

Next, we move to the chest and typically use four 4 mm ports, one at the tip of the scapula, one in the axilla, and two more inferior. The inferior pulmonary ligament is divided to expose the posterior mediastinum. The distal esophageal pouch and vagus nerves are identified. If present, the fibrous cord coming off the superior tip of the distal esophageal pouch can be used as a handle during dissection to fully mobilize the pouch before dividing it. The pledgeted traction sutures are placed in the lower pouch thoracoscopically with the same method as for the upper pouch, again with endoscopic guidance retrograde through the gastrostomy site, typically with 4–0 Ethibond for the lower pouch.

The posterior mediastinal pleura in front of the spine is opened at the apex of the left chest posterior to the left subclavian artery and superior to the aortic arch, to carry our dissection to and connect to the window started from the neck to deliver the proximal esophageal pouch and sleeve with the silk suture pulley suture into the chest. Traction can be set up with a 0 Ethibond suture tied with a one-way slip knot to connect the double loop bucket handles of the upper and lower pouches for pouch to pouch traction. Each pouch can also be set around a rib with #2 FiberWire going through the bucket handle and wrapped around a rib with the suture tied in the subcutaneous space. Seprafilm slurry is placed around the lower esophageal pouch to minimize adhesions on subsequent returns to the operating room for traction adjustments. A chest tube is placed through the most inferior port site, and the neck and chest incisions are closed.

Postoperatively, the clips marking the pledgets as well as trailing clips on each esophageal pouch are monitored with chest radiographs. For internal traction adjustments and suture tightening, we typically return to the operating room every 7–10 days, with the potential to extubate in between traction adjustments and no need for prolonged paralysis. When the esophageal pouches are found to be touching or overlapping, the anastomosis is performed either MIS or via thoracotomy. At any point in the traction process prior to and including the time of anastomosis, the operating surgeon may convert to an open approach or external traction for a variety of reasons, primarily suture pull outs or leaks during traction, or to improve technical quality during anastomosis.

3. Results

71 patients underwent Foker process from December 2016 to November 2021, 37 patients (52%) via open thoracotomy and 34 patients (48%) via MIS technique. Of 37 open cases, 31 patients (84%) underwent right thoracotomy (all with left aortic arch) and 6 patients (16%) left thoracotomy (4 with left aortic arch, 2 with right aortic arch). Of 34 MIS cases, 28 patients (82%) were approached from the left side, all with left aortic arch. These 28 LGEA patients underwent left-sided MIS repair and were included in the study.

Of included patients, 18 patients (64%) were male. 17 patients (61%) were premature and the median birth weight was 2.3 kg (IQR 1.8–2.6 kg). 12 patients (43%) had associated congenital heart disease. 15 patients (54%) had VACTERL syndrome and 4 patients (14%) had a genetic anomaly. The majority of patients (20, 71%) had type A EA, followed by type B EA (5, 18%) and type C EA (3, 11%). 7 patients (25%) had a prior failed right-sided repair and 2 patients (7%) had a prior cervical esophagostomy.

These patients underwent left-sided MIS repair at median age 4 months (IQR 2.75–5 months) with median weight 5.4 kg (IQR 4.3–6.5 kg) at the time of surgery. Median gap length was 5 cm (range 2–8 cm) off tension and 2.5 cm (range 1–5.5 cm) on tension. Patients underwent median 3 (range 2–8) operations and were on traction median 13.5 days (IQR 11–21 days) until esophageal anastomosis. 21 patients (75%) also underwent left neck dissection in addition to a left chest approach.

9 patients (32%) underwent a completely MIS approach, whereas 5 patients (18%) converted to open at the first operation and 14 patients (50%) converted to open later in the traction process. Of those converted to open at the first operation, the reasons for conversion were 3 for concern for tissue quality, 1 for poor tolerance of thoracoscopy, and 1 with too much tension on attempted primary anastomosis. Of those converted to open later in the traction process, the reasons for conversion were 6 to improve anastomosis quality, 3 for leaks on traction, 2 with takedown of the cervical esophagostomy, 1 for dense adhesions, 1 for an esophageal muscle tear, and 1 for failure to produce effective growth.

Traction was internal in 19 patients (68%), external in 3 patients (11%), and combination in 6 patients (21%). All external traction in these cases was performed by open thoracotomy. For the 23 patients who successfully underwent initial left thoracoscopic traction, traction was set up around ribs in 15 patients (65%), pouch to pouch in 4 patients (17%), and both pouch to pouch plus additional upper pouch traction around a rib in 4 patients (17%). Of the 33 thoracoscopic traction adjustments, 25 (78%) were traction adjustments around ribs, 7 (21%) were pouch to pouch, and 1 (3%) was pouch to pouch plus additional upper pouch traction around a rib.

10 patients (36%) underwent concomitant airway work for TBM during the traction process, mostly cervical posterior tracheopexy by open left neck approach (8, 29%). One patient underwent thoracoscopic descending aortopexy and one patient underwent open posterior tracheopexy to reinforce the tracheal diverticulum repair site at the common wall with the upper esophageal pouch. Two patients had the primary problem of severe TBM with inability to extubate, undergoing right sided posterior airway work before addressing the esophagus from the left side. Both of these patients also needed anterior airway work, one pre Foker process and one post Foker process, with one of these patients ultimately requiring a tracheostomy. One patient underwent stricture resection for a recalcitrant esophageal stricture and underwent concomitant descending aortopexy, anterior pericardiopexy, and thymectomy at that time through a left thoracotomy. One patient had a missed proximal TEF and later underwent repair and cervical posterior tracheopexy through a left neck approach.

Median length of paralysis was 5.5 (IQR 1–12) days, mechanical ventilation 14 (IQR 7.75–19.5) days, intensive care unit (ICU) length of stay 34 (IQR 30–62) days and hospital stay 66.5 (IQR 50–102.5) days. Median follow up was 15.4 months (IQR 7.5–31.7 months) after anastomosis. 4 patients (14%) had anastomotic leak managed with antibiotics and/or esophageal vacuum therapy. Median number of esophageal dilations was 3.5 (range 0–13). 5 patients (18%) required stricture resection. 1 patient (4%) had a chyle leak that resolved with formula changes and 1 patient (4%) had a deep vein thrombosis treated with anticoagulation. 6 patients (21%) had unilateral vocal cord injury, 5 on the left side and 1 on the right side. 4 were asymptomatic and incidentally found. 2 were symptomatic, 1 with a weak cry and voice that resolved and 1 with stridor and aspiration for which we do not have follow up. 11 patients (39%) underwent Nissen fundoplication. None have needed esophageal replacement. At latest follow-up, 12 patients (43%) were on full oral feeds, whereas 13 patients (46%) were receiving gastric feeds and 3 patients (11%) jejunal feeds.

4. Discussion

The management and approach to surgical treatment of LGEA continue to evolve. Tension-based Foker process esophageal growth induction techniques have been used to preserve the native esophagus. External traction through open thoracotomy has been well described. However, this technique has evolved in our centers to include open or MIS approach, external or internal traction, and right or left sided repair [6]. Certainly, a MIS internal traction approach has benefits compared to traditional open external traction, including no need for paralysis while on traction, ability to extubate between traction adjustments, shorter time in the ICU, less need for narcotic and sedative drips, and subsequently less withdrawal issues from prolonged sedation weans.

We have previously shown that left-sided repair for LGEA in select patients had similar outcomes to right-sided repair, including anastomotic leak rate, need for stricture resection, and feeding status [2]. The first patient in our series who underwent left-sided thoracoscopic repair was noted to have a very large upper esophageal pouch left of the trachea with significant tracheal compression, in which there was concern that mobilization and placement to the right of the trachea for a right sided repair would worsen posterior tracheal compression and TBM. We had seen this complication in several prior patients with the typical right sided approach. It could be that right neck dissection to get better esophageal mobilization would have addressed this issue as well. Our transition to the more common use of neck dissection and left sided approach occurred roughly simultaneously. Regardless of aortic arch position, a left-sided approach can be considered for those with minimal thoracic TBM and no thoracic TEF or one that has already been divided and repaired. A left-sided approach also allows for a clean operative field in patients with prior attempted right-sided repairs complicated by leak or infection, or those with a left-sided cervical esophagostomy following a failed repair. Over recent years, left-sided MIS internal traction has evolved into our most commonly used strategy for LGEA, primarily because of the natural leftward bias of both upper and lower esophageal pouches in the majority of patients.

We have found that the addition of a neck dissection on the same side as the chest approach has several advantages. It can be used to deal with the issue of a leftward upper esophageal pouch to fully mobilize it and more successfully pull it to the left side to facilitate less tracheal compression. The neck approach also allows for more straightforward recurrent laryngeal nerve identification and preservation. We additionally now routinely use recurrent laryngeal nerve monitoring in these cases. The major driver for consideration of neck dissection in our experience is scarring within the thoracic inlet that can be a tough obstacle for successful esophageal growth of the upper pouch. When the esophagus is scarred or otherwise attached (common wall with trachea) to surrounding structures, it won't grow well. Furthermore, repeated dissections in the thoracic inlet to deal with this scarring create significant additional risk of injury to the trachea, upper esophageal pouch, and especially the recurrent laryngeal nerves. We have found that using the silastic sleeve around the upper esophagus as it passes through the thoracic inlet is the best strategy to prevent this scarring problem, and therefore allow one good dissection of this area with no need to repeat, while facilitating the best chance for optimal esophageal growth.

Endoscopy during initial traction suture placement for the esophageal pouches was added to ensure no mucosal violation and lessen pouch leaks if the sutures pull out. The double loop bucket handle traction set up allows for traction adjustment without redoing the traction sutures each time. Traction can be placed pouch to pouch or around ribs, sometimes both for the upper pouch to create lateral pull away from the aorta and allow stronger traction on the upper pouch which typically has more robust wall thickness and tissue than the lower pouch. After initial traction suture placement, patients are on parenteral nutrition with no enteral feeds to allow for initial healing of the esophageal pouches. Based on how things look at the first traction adjustment, we often convert the gastrostomy to a gastrojejunostomy to feed distally and keep the stomach decompressed. If there is an esophageal pouch suture pull out or leak on traction, we would typically convert to an open approach. One option is to perform an anastomosis if the pouches are close enough, or if an anastomosis is not possible, we would generally fix the leak and convert to external traction, keeping the child paralyzed to prevent swallowing.

LGEA is more than a gap in the esophagus, as a significant percentage of patients have associated tracheobronchomalacia, great vessel anomalies, or other issues that can impact their long-term outcomes beyond achieving an esophageal anastomosis. Preoperative cross-sectional imaging and endoscopic evaluation are used to evaluate all of the esophageal and airway issues, as well as understand the anatomic relationship of the mediastinal structures, to design a customized approach. In cases with severe thoracic TBM, a right-sided approach can be used to correct TBM as a posterior tracheopexy cannot be performed well from the left side with a left aortic arch. We have generally preferred an open thoracotomy for these airway procedures, although MIS techniques may be considered as well. Interestingly, another advantage of the left approach for esophageal anastomosis regarding impact on the airway is the repositioning of the esophagus away from the membranous trachea in general. We have seen numerous cases of tracheomalacia in esophageal atresia patients that worsened over time based on posterior intrusion by the esophagus itself, especially when the esophagus becomes dilated, with or without a stricture. When the esophagus is moved to the left of the left aortic arch, it no longer occupies the anatomic space posterior to the trachea, and that source of posterior intrusion is thereby eliminated. In some cases, a staged approach can be considered, with a right-sided approach for airway work to stabilize the respiratory status, followed by a left-sided approach later for esophageal work in a clean field. In cases with primarily cervical TBM with a large leftward upper esophageal pouch, a left-sided neck and chest approach can keep the esophagus on the left side to lessen tracheal compression, as well as allow for cervical posterior tracheopexy. Certainly, there is selection bias at play, but we do think it is important to think about the airway in the evaluation of these patients. It is not that we always choose a particular side or approach for repair, but each particular case is individualized and over time, we have tended to favor a left-sided MIS approach with internal traction.

For multiple reasons including the tendency of both esophageal pouches to have a leftward bias, less tracheal compression by the upper pouch, and a clean field of surgery for reoperative cases, we now more commonly use the left-sided approach for MIS LGEA repair compared to the right side, even with a left aortic arch. We will continue to study and refine our algorithms in all respects for LGEA. Given the complexity of this patient population, we advocate for a comprehensive multidisciplinary evaluation of the esophagus, airway, and great vessel anomalies to design a customized approach to each individual patient to optimize esophageal and airway outcomes.

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