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Long-gap esophageal atresia

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ABSTRACT

The management of long-gap esophageal atresia remains challenging with limited consensus on the definition, evaluation, and surgical approach to treatment. Efforts to preserve the native esophagus have been successful with delayed primary anastomosis and tension-based esophageal growth induction processes. Esophageal replacement is necessary in a minority of cases, with the conduit of choice and patient outcomes largely dependent on institutional expertise. Given the complexity of this patient population with significant morbidity, treatment and long-term follow-up are best done in multidisciplinary esophageal and airway treatment centers.

Introduction

Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) is the most common congenital anomaly of the esophagus.[1] Pure or isolated EA (type A) accounts for 8% of all EA cases with an incidence of 1 in 40,000 live births.[2] Pure EA is often described as long-gap EA (LGEA), although there is evidence that type C with a distal TEF is the most common variant of LGEA.[3]

There is little consensus on the definition of LGEA; however, the term is used to describe cases in which a primary anastomosis of the proximal and distal ends of the esophagus cannot easily be performed under acceptable tension by the operating surgeon. The gap length used to define LGEA varies widely across the literature, with a cutoff ranging from 2 to over 3 cm.[4–7] Typically, when early primary repair is not possible, alternative techniques are necessary to bridge the gap and restore esophageal continuity. LGEA remains a technically challenging subset of EA cases, and there are currently no definitive standardized guidelines for the evaluation, management, and surgical approach to the treatment of LGEA.[1,8–12]

Preoperative assessment

Pure EA is often suspected on prenatal imaging due to an "absent stomach" at multiple fetal imaging time points, combined with the development of polyhydramnios after 24 weeks' gestation. EA is usually clinically diagnosed after delivery due to a distal TEF that allows gas accumulation in the stomach so that there is no "absent stomach." In such cases, a baby that chokes and sputters may have an attempted orogastric tube placement, but when the orogastric tube is unable to be passed into the stomach, the tube may be seen within a blind-ending proximal esophageal pouch on plain film. A radiographic "gasless abdomen" suggests pure EA without a distal TEF or EA with solely a proximal TEF.

Diagnostic evaluation includes screening for associated anomalies in the VACTERL syndrome. A preoperative echocardiogram is important for identifying congenital heart disease and vascular anomalies that may affect operative planning. These include the side of the aortic arch (right or left), and the presence of aberrant subclavian vessels, double aortic arch or other types of vascular rings. Initial management includes proximal esophageal pouch decompression, aspiration precautions such as elevated head of bed and frequent suctioning, and gastrostomy placement for enteral feeding and evaluation of the distal esophageal pouch, although this is somewhat controversial.[1,13]

Gapogram

Assessment of EA is incomplete without an intraluminal contrast study of both the proximal and distal esophageal segments. These studies help to identify other issues such as TEFs, congenital esophageal strictures, and esophageal duplications or cysts—all of which increase the complexity of esophageal reconstruction. These studies also help to define the luminal lengths and the distance between the 2 lumens—we call this a gapogram. The gap can be measured by fluoroscopy with water-soluble contrast injected into the distal esophageal pouch via the gastrostomy and a catheter placed in the proximal esophageal pouch. Water-soluble isotonic contrast is used to minimize the consequences of aspiration of contrast.

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Other methods to measure the gap have been described using dilators or a flexible endoscope to define the distal esophageal pouch; however, results can be inconsistent depending on variable forward pressure applied by the operator.[14] The gap can be expressed in centimeters or the number of vertebral bodies.

Endoscopic airway evaluation

An adjunct to the preoperative assessment of EA is preoperative endoscopic airway evaluation. Its routine use in this setting is controversial, with only 21.5–60% of surgeons performing preoperative tracheobronchoscopy.[15–17] We highly encourage all patients to undergo preoperative tracheobronchoscopy, both with very active breathing to assess airway dynamics, and with relatively deep anesthesia to assess static airway structure. Diagnostic laryngoscopy and video tracheobronchoscopy (DLB) are used to assess supraglottic structures and vocal cord function, the larynx for presence of a laryngeal cleft or laryngotracheoesophageal cleft, the presence of one or more TEFs or tracheal diverticula, and anomalies within the tracheobronchial tree such as complete rings or airway compression. Associated tracheobronchial anomalies are present in nearly half of EA patients, and DLB findings can impact clinical management in 21–45% of patients.[18–20] DLB can be particularly useful in the pure EA cohort in identifying a proximal TEF in 20–33% of patients.[19–21]

Tracheobronchoscopy is the gold standard to evaluate tracheomalacia, a common respiratory problem among EA patients. Older studies report a prevalence of 11–33% in this population, likely an underestimate given the wide spectrum of disease and common misdiagnosis, with a recent study reporting tracheomalacia in 87% of EA patients.[22–25] DLB is done in spontaneously breathing (with coughing or Valsalva) patients to fully evaluate dynamic motion in the tracheobronchial tree throughout the respiratory cycle. Severe tracheomalacia is characterized by coaptation of the airways with anterior and posterior collapse during expiration in spontaneously breathing patients. The early and accurate diagnosis of tracheomalacia is important because excessive airway collapse or obstruction leads to ineffective ventilation and poor clearance of secretions. This can result in frequent respiratory infections, possibly progressing to permanent lung damage in 27% of patients by 8 years of age, and in the most severe cases, blue spells and apparent life-threatening events.[25–28] If severe tracheomalacia is identified, surgical correction may be warranted.[28–31]

Management of LGEA

The management of LGEA depends largely on the gap length and the size and quality of the proximal and distal esophageal segments, as well as any associated anomalies such as TEFs, strictures, duplications or cysts, and vascular anomalies. The size of the distal esophageal pouch can range from a tiny primordium below the diaphragm to a sizable segment reaching well into the mediastinum.

All surgery is local, so the local surgical experience is critical for success if surgery is attempted. Shorter gaps may be brought together primarily depending on the above factors and the surgical experience, typically gaps between 0 and 3 cm, although some surgical teams may be able to get longer gaps together. We do not think that mobilizing the stomach up into the mediastinum is a good long-term solution, although it may be needed in some cases.[32]

Some surgical teams may choose delayed primary anastomosis, while others may choose a variety of intraoperative techniques that can be utilized to gain the adequate length to facilitate a primary anastomosis. For longer gaps, staged tension-based esophageal lengthening techniques are often used in attempts to preserve the native esophagus, but in some cases, esophageal replacement may be necessary. LGEA remains a surgical challenge, although most agree that the native esophagus is the preferred conduit for esophageal reconstruction. The ideal approach to restoring esophageal continuity in LGEA has been widely debated for decades with little consensus, and is often dependent on the training and experience of a particular center.[33–37]

Delayed primary anastomosis

In some cases, particularly in those with pure LGEA and no TEF, a primary repair may be achieved by giving time for the esophageal ends to grow.[2,38] Patients are managed with proximal esophageal pouch decompression and bolus gastrostomy feedings over a period of 1–3 months. The gap tends to increase as the spine grows longer, and decrease as the esophageal segments grow longer. The gap sometimes tends to narrow by spontaneous growth, thought to be in part related to swallowing attempts for the proximal esophageal pouch and gastric reflux into the distal esophageal pouch. Some use bougienage as a concurrent mechanical technique to stretch the esophageal pouches while waiting. Weighted bougies are passed into the proximal and distal esophageal pouches with forward pressure applied once or twice daily. Gapograms are serially performed during this waiting period, and the timing of operation for delayed anastomosis typically occurs when the esophageal ends are radiographically less than 2 vertebral bodies apart. Successful primary anastomosis with delays of up to 12 months and gaps of up to 7 cm or 8 vertebral bodies have been reported with good long-term functional results.[3,39–41] However, failure using this technique is not uncommon, and the delays result in prolonged hospitalization and oral aversion that may take years to overcome.

Esophageal myotomy

Historically, mobilization of the distal esophageal pouch was thought to be limited by a tenuous vascular supply; however, it is now common for both proximal and distal esophageal pouches to be fully mobilized with low risk for ischemia. If the ends of the esophagus still do not come together and the remaining gap is short, an esophageal myotomy can be performed to gain approximately 0.5 cm of length per myotomy. Circular and spiral myotomies of the proximal and distal esophageal pouches have been described to gain adequate length to facilitate a primary anastomosis.[42–44] Myotomies tend to divide much of the muscular blood supply to the esophagus distal to the myotomy, causing relatively worse ischemia. Esophageal myotomies can be associated with impaction of food particles in the myotomy site and formation of pseudodiverticula in up to 20% of patients, as well as increased risks of anastomotic leakage and stricture formation.[7,45]

Extrathoracic esophageal elongation: Kimura technique

Staged esophageal elongation procedures are based on tension-induced lengthening by application of external or internal traction to the ends of the esophageal pouches. It remains controversial whether esophageal lengthening occurs by stretch or true growth, although growth seems to be occurring in animal models.[46] Kimura described an external traction technique in which a cutaneous cervical esophagostomy of the proximal esophagus is serially translocated down the anterior chest wall until sufficient length is obtained for primary anastomosis.[47] Oral sham feedings can be continued during the staged repair, preventing oral aversion. Small case series have reported the ability to achieve primary

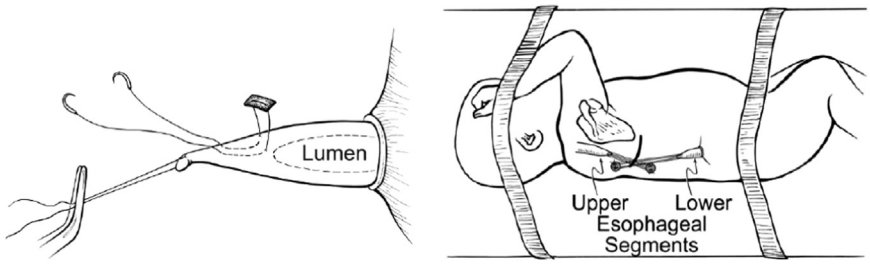

Fig. 1. Tension-induced esophageal growth induction—Foker process. Pledgeted traction sutures are placed on both proximal and distal esophageal pouches with care not to enter the lumen. The sutures are externalized through the chest wall, with traction placed on externalized sutures to lengthen the pouches and narrow the gap. (Adapted with permission from Foker et al.[52]).

anastomosis with this technique; however, it is not widely used due to technical difficulties and the need for externalization of the esophagus.[48–51]

Internal traction-induced esophageal growth: Foker process

The Foker process uses intrathoracic axial tension-induced growth for esophageal lengthening to allow for primary esophageal reconstruction[7,52] (Figure 1). Initial growth induction occurs in stage 1 with placement of traction sutures on both proximal and distal esophageal pouches, and externalization of these sutures through the chest wall. Tension on externalized sutures is applied over days to weeks to lengthen the pouches and narrow the gap until primary anastomosis in stage 2. During this period, patients are mechanically ventilated and paralyzed to minimize disruption of sutures. Nevertheless, traction sutures can become dislodged, requiring suture reconfiguration via repeat thoracotomy.

Many institutions, including our own, use the Foker process in LGEA, and have demonstrated the ability to preserve the native esophagus and achieve a primary repair.[50,52–55] In Foker's personal case series of 63 patients with a gap greater than 2.5 cm, a primary repair was achieved in all cases with an average of 14 $\pm$ 7 days (range 3–31 days).[11] Our institution reported our experience in 52 patients, comparing Foker primary repair ($n = 27$) and secondary repair in patients with previous attempts at EA repair ($n = 25$).[56] Esophageal continuity was restored in 96% of primary cases and 68% of secondary cases, with median times to anastomosis 14 days for primary cases and 35 days for secondary cases. Secondary cases and number of thoracotomies were significant predictors for the inability to achieve an anastomosis and development of a leak. In total, 63% of primary cases and 9% of secondary cases were able to eat by mouth, with primary cases and patients with longer follow-up as predictors of progression to full oral feeding. The Foker process proved to be possible, but more challenging in secondary cases. A recent systematic review and meta-analysis concluded that the Foker process is at least as effective as delayed primary anastomosis in LGEA.[57] The Foker process was associated with lower risk of complications, including leak, stricture, and gastroesophageal reflux, as well as a shorter time to anastomosis.

Thoracoscopic repair

The first thoracoscopic repair of pure EA was reported in 1999. Since then, there has been interest in minimally invasive techniques to minimize pain, scarring, and long-term musculoskeletal morbidity associated with thoracotomy.[9] Although most surgeons still utilize an open approach, thoracoscopic repair of LGEA and the Foker process have been reported.[58,59] Technical benefits include improved visualization and better access to the esophageal pouches for mobilization; however, it is technically challenging with a steep learning curve.

Esophageal replacement

In some cases of LGEA, esophageal replacement may be necessary when repeated attempts to preserve the native esophagus and restore esophageal continuity fail. Esophageal replacement is also considered in cases where there have been complications from an anastomotic leak, refractory stricture formation, and recurrence of TEF or poor functional outcome affecting quality of life. There is little consensus on the preferred conduit for esophageal replacement; however, use of the stomach, colon, and small bowel has been described.

Gastric transposition

Gastric transposition has gained favor at many institutions and is the most commonly used method of esophageal replacement, likely due to its relative technical ease.[33,35] The procedure utilizes a well-vascularized conduit with adequate length and a single anastomosis. It can be performed via cervical and laparotomy incisions, avoiding a thoracotomy, and minimally invasive techniques are becoming more popular.[60–62] The good aspects of this procedure are that it is relatively easy to perform, and has reliable results and few immediate complications other than anastomotic leaks and strictures that are managed relatively easily. In some low resource areas, this may be the best that can be accomplished safely.

The problem is that the gastroesophageal junction is displaced into the chest or neck, resulting in gastroesophageal reflux with increased risk for metaplasia and Barrett's esophagus, as well as chronic aspiration.[8,63] Other complications include delayed gastric emptying, loss of gastric reservoir function resulting in poor weight gain and anemia, and pulmonary compromise from mass effect in the chest. In a large series of 236 patients, Spitz[64,65] reported a leak rate of 12%, stricture in 20%, and mortality in 2.5%. Other series report higher complication rates, leak in 14–34%, stricture in 30–40%, and gastroesophageal reflux in 40–70%.[35,66–68]

Gastric tubes

Among gastric conduits, gastric tube esophagoplasty is less commonly used than gastric transposition. The tube is created along the length of the greater curvature, and can be positioned in a peristaltic or antiperistaltic fashion. The caliber of the tube approximates the esophagus with adequate length and good blood supply; however, there is a long suture line with increased risk of gastroesophageal reflux, leak, and stricture.[69,70] In a study comparing gastric tubes with delayed primary anastomosis for LGEA, gastric tubes resulted in more long-term complications with reflux and stricture.[71]

Colon interposition

Based on the Waterston operation for replacing the esophagus in esophageal cancer cases, colon interposition is less

commonly used than gastric transposition for esophageal replacement in the pediatric population.[33] The colonic graft can be placed in either a peristaltic or antiperistaltic orientation in the posterior mediastinum or substernally. The operation is more challenging than gastric transposition, requiring 3 anastomoses, including proximal and distal interposition and a colo-colonic anastomosis to establish continuity. Besides leak, stricture, and reflux, the colon interposition can elongate and dilate in the chest over time, becoming redundant with poor emptying, leading to stasis, halitosis, and pulmonary compromise.[72–74] A minimally invasive technique with laparoscopically assisted esophagectomy and colon interposition has recently been described in a small series in children.[75]

Jejunal interposition

Jejunal interposition is the most technically challenging of the esophageal replacement methods; however, it arguably has the best long-term functional outcomes, working well after 30 or more years in the longest reported follow-up.[76–78] A jejunal conduit can be fashioned at any length with a caliber approximating the esophagus. Thoracic esophageal anastomoses are possible for shorter gaps; however, longer interpositions into the neck with additional vascular anastomoses may be necessary for longer gaps (Figure 2). Most importantly, the jejunal conduit maintains its intrinsic peristalsis, rendering it more resistant to gastroesophageal reflux, and it does not become dilated enough to affect pulmonary function as do gastric and colon conduits. The technical demands require multidisciplinary surgical expertise, often utilizing cardiothoracic, microvascular, and even otolaryngology skills to optimize outcomes. Because of the favorable long-term outcomes, it has been our institution's preferred esophageal replacement conduit since 2010. Our institution reported early outcomes of jejunal interposition in our first 10 patients, with 4 patients utilizing microvascular supercharging for a long jejunal graft. At median follow-up of 1.5 years, there was no long-term graft loss or deaths, with 6 patients eating completely by mouth.[79] In a study comparing gastric transposition and jejunal interposition, jejunal interposition had more early anastomotic complications, but less reflux and better oral feeding and growth.[63]

A recent meta-analysis of esophageal replacement in LGEA concluded that gastric transposition and colon interposition had comparable mortality, anastomotic complications, and graft loss.[80] In the long-term, gastric transposition was associated with higher respiratory morbidity but fewer gastrointestinal complications than colon interposition. Only 6% of cases used jejunal interposition, making it difficult to draw conclusions. We feel that as the skill sets that experienced teams build to successfully perform jejunal interpositions for esophageal replacement, the outcomes will justify the development of specialized centers.

Future directions in tissue engineering

Recent advances in regenerative medicine have made progress in tissue engineering an esophagus for esophageal replacement.[81,82] Scaffolds can be acellular or seeded with epithelial and muscle cells, but further work needs to be done to vascularize and innervate the graft to optimize long-term functional outcomes. These techniques remain of research interest only at this point.

Conclusions

The management of LGEA continues to evolve and remains one of the most challenging entities in pediatric surgery. In most cases of LGEA, the native esophagus can be preserved with delayed esophageal anastomosis and traction techniques, and failed attempts to save the esophagus do not preclude esophageal replacement.[13] True comparative studies are lacking in the literature, with variation in practice and limited consensus on the definition, evaluation, and optimal treatment

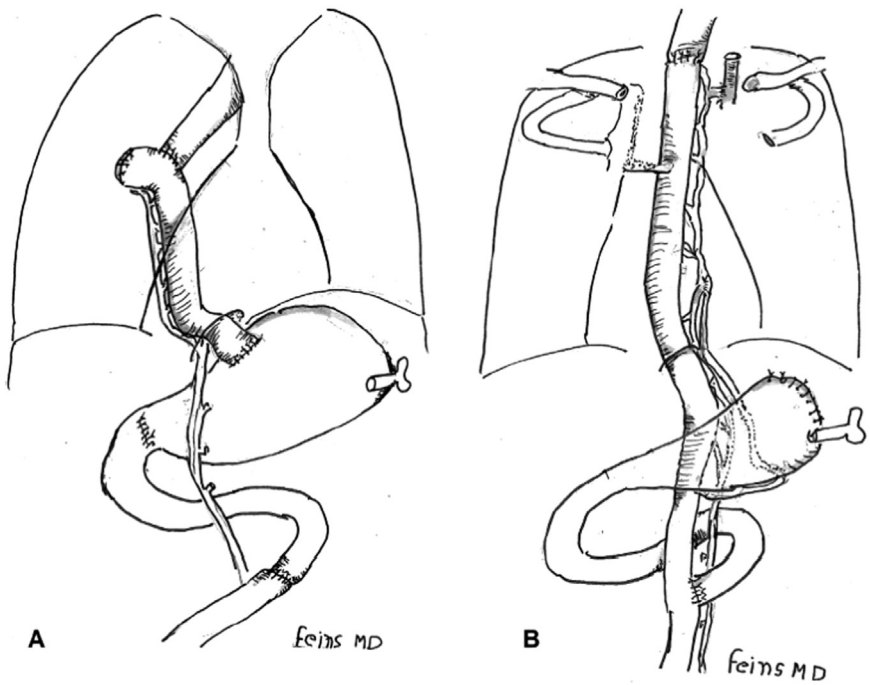

Fig. 2. (A) Jejunal interposition from thoracic esophagus to stomach without microvascular supercharge. (B) Jejunal interposition to cervical esophagus and Roux-en-Y reconstruction with supercharging using internal mammary and gastroepiploic vascular anastomoses. (Adapted with permission from Bairdain et al.[79]).

strategies to improve patient outcomes.[15,16] Surgical approach is often based on institution expertise. Early referral to a multidisciplinary center may be warranted in complex cases, including those with a "gasless abdomen" implying LGEA.[55,56] We also suggest that any patient who fails an initial operation for EA repair be referred to a center that specializes in the care of EA patients.

LGEA patients can have significant long-term respiratory and gastrointestinal morbidity.[25,39,83–85] Given the heterogeneity and complexity of this patient population, multidisciplinary esophageal and airway treatment centers are recommended for their care and long-term follow-up.[86]

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