Review Article

Management of long-gap esophageal atresia

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Abstract: A uniquely challenging subset of infants diagnosed with esophageal atresia (EA) are those born with long-gap EA (LGEA). The common unifying feature in infants with LGEA is that the proximal and distal segments of the esophagus are too far apart to enable primary anastomosis via a single operation in the newborn period. Although any type of EA can technically result in a long gap, the Gross type A variant occurs in 8% of all EA cases and is most commonly associated with LGEA. In this review, we provide an evidence-based approach to the current challenges and management strategies employed in LGEA. There are fortunately a range of available surgical techniques for LGEA repair, including delayed primary repair, staged repair based on longitudinal traction strategies to lengthen both ends (e.g., Foker procedure, internal traction), and esophageal replacement using other portions of the gastrointestinal tract. The literature on the management of LGEA has long been dominated by single-center retrospective reviews, but the field has recently witnessed increased multi-center collaboration that has helped to increase our understanding on how to best manage this challenging patient population. Delayed primary repair is strongly preferred as the initial approach in management of LGEA in the United States as well as several European countries and is supported by the American Pediatric Surgery Association recommendations. Should esophageal replacement be required in cases where salvaging the native thoracic esophagus is not possible, gastric conduits are the preferred approach, based on the relative simplicity of the operation, low postoperative morbidity, and long-term durability. Long-term followup for monitoring of swallowing function, nutritional status, aspiration/respiratory illnesses, gastroesophageal reflux, and associated comorbidities is essential in the comprehensive care of these complex patients.

Keywords: Esophageal atresia (EA); long gap; Foker procedure; esophageal replacement

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Introduction

Esophageal atresia (EA) with or without tracheoesophageal fistula represents a set of relatively rare congenital anomalies that result in a disruption of esophageal continuity. EA is seen globally in approximately 3 in 10,000 live births. Infants with EA require surgical intervention or will eventually succumb to their disease secondary to chronic aspiration (1).

Approximately 10% of EA patients have a complex variant referred to as long-gap esophageal atresia (LGEA) (2), most commonly associated with Gross type A and B defects (*Figure 1*). Although there is no universally accepted definition of LGEA, one accepted functional definition is any EA anatomy where an anastomosis between the

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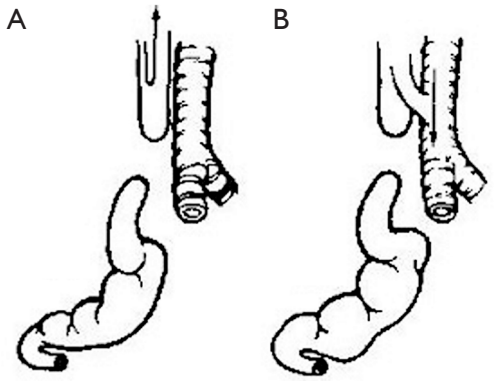

Figure 1 Schematic illustrations of common long-gap esophageal atresia variants based on the Gross classification. (A) Type A: isolated esophageal atresia without tracheoesophageal fistula. (B) Type B: esophageal atresia with proximal tracheoesophageal fistula. In both variants, the stomach is hypoplastic secondary to lack of amniotic fluid transit in utero.

upper and lower esophageal segments (pouches) cannot be successfully performed at birth without creation of a hiatal hernia (3). Displacement of the gastroesophageal junction into the chest is generally not viewed as an acceptable long-term outcome for LGEA since this would lead to overwhelming gastroesophageal reflux disease (GERD), chronic aspiration, and end-stage lung disease. Under this definition, some cases with a very short upper esophageal segment and distal tracheoesophageal fistula or those where the distal fistula is connected to the left or right bronchus (as opposed to the trachea) would be included in this surgical definition of LGEA. Moreover, many extremely premature EA patients may also be difficult to repair at birth, regardless of the specific anatomy, due to the fragility of the esophagus in very low birthweight infants (4).

Diagnostic workup

Prenatal diagnosis

While most patients with EA are diagnosed within 24–48 hours after birth, the anomaly is suspected prenatally in approximately 10–20% of patients (5). In such cases, fetal ultrasonography after 25 weeks gestation reveals an absent or small stomach bubble in combination with polyhydramnios. Skilled sonographers can sometimes identify a dilated upper esophageal pouch which increases specificity for the diagnosis of EA. Because studies have shown an increased risk for long-gap disease when EA is suspected in utero (6), prenatal counseling discussions with families should raise the possibility of LGEA, which has a different postnatal management course and is associated with prolonged hospitalization when compared to the majority of EA cases. Some pediatric surgeons and maternal-fetal medicine physicians have advocated for the use of fetal magnetic resonance imaging (MRI) to further clarify anatomy in prenatally suspected cases (7); however MRI has yet to become part of the standard work up at many institutions as these studies are often inconclusive and rarely alter prenatal management. Regardless, physicians should ensure that expectant parents receive comprehensive counseling regarding potential associated anomalies (e.g., cardiac) and the increased risk for premature birth.

Postnatal diagnosis

Shortly after birth, EA should be suspected in any newborn who presents with excessive drooling, a barking cough, and inability to tolerate feeds. Placement of an orogastric (OG) tube by a skilled provider should be attempted. Classically, infants with EA will demonstrate a failure to pass the OG tube beyond 10 cm with confirmation that the tip of the OG catheter does not pass beyond the upper thoracic region (T4) on plain radiographs. The concomitant presence of a gasless abdomen is a highly suspicious for LGEA, and more specifically the type A (or more rarely type B) variant (*Figure 2*). Some surgeons have advocated for the administration of 1–2 milliliters of contrast into the upper pouch to confirm the diagnosis, but this study should be reserved for equivocal cases given the aspiration risk associated with contrast administration in this setting (5).

Associated anomalies

Because EA is known to be associated with other birth defects, it is critical that pediatric surgeons have a complete understanding of all co-morbidities prior to proceeding to the operation room. All EA patients, including those with LGEA, should be assessed for VACTERL anomalies, namely *v*ertebral, *a*norectal, *c*ardiac, *t*rachea-*e*sophageal, *r*enal, and radial *l*imb defects. Physical exam should include auscultation of the heart, examination of all distal extremities, and assessment for a normally placed anus. Although spinal and renal ultrasounds should be obtained, the most important preoperative test is an echocardiogram to rule out cardiac defects (over 50% of cases) and to assess the laterality of the aortic arch.

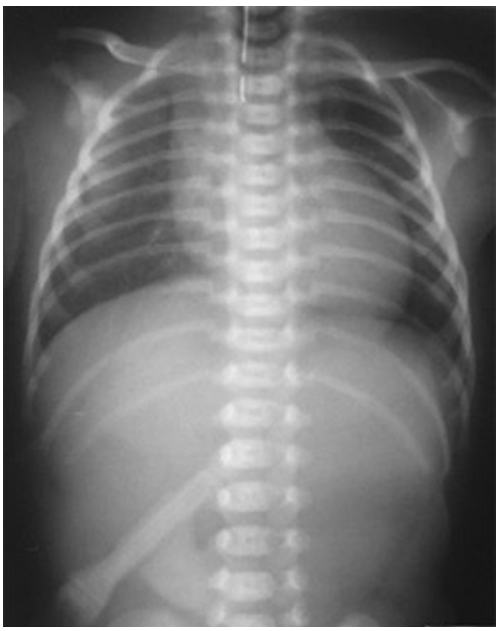

Figure 2 Anteroposterior plain film radiograph of a representative newborn with long-gap esophageal atresia. The orogastric catheter terminates in the upper thorax, and the abdomen is gasless.

VACTERL syndrome, which is formally defined as the presence of 3 or more VACTERL anomalies, may be seen in up to 25% of LGEA infants (8). If a right-sided aortic arch is detected by echocardiogram, patients should be further evaluated with cross-sectional imaging (either a computed tomography or magnetic resonance imaging scan of the chest), to rule out the presence of a vascular ring (9). Outside of the VACTERL disorders, it is important to consider chromosomal anomalies, particularly trisomy 21, and genetic syndromes such as CHARGE syndrome (coloboma, heart defects, atresia choanae, retardation of growth and development, genital/urinary abnormalities, and ear anomalies).

Preoperative management and repair timing

Once the diagnosis of LGEA is established based on failure to pass an OG tube and the presence of a gasless abdomen, surgeons should consider whether repair in the early neonatal period is appropriate. Although most LGEA patients do not undergo esophageal reconstruction until at least 30 days of age, a handful of experienced, large-volume centers have advocated for staged, traction-based thoracoscopic approaches to LGEA repair in the early neonatal period in selected patients (e.g., full-term and 3 kg without significant comorbidities) (10). The rationale for very early repair includes avoidance of a gastrostomy tube for nutritional support and potentially reduced rates of oral aversion.

In the vast majority of newborns with LGEA, placement of a gastrostomy tube (GT) remains the standard of care. The GT serves two purposes: (I) to provide stable enteral feeding access, and (II) to allow for subsequent assessment of the esophageal gap. Although the procedure may be performed laparoscopically, it is a technically challenging operation since the stomach is usually hypoplastic. Most surgeons therefore elect to carefully construct a GT in LGEA neonates in an open fashion through a small left upper quadrant laparotomy. Surgeons should take care to place the GT away form the greater curvature, in order to preserve the gastroepiploic blood vessels in the instance that a gastric conduit be required at a later date, and fill the balloon with minimal fluid to prevent iatrogenic gastric outlet obstruction. Placement of a Malecot style tube (10 or 12 Fr) is another option to help prevent pyloric obstruction during enteral feeding.

Once the GT site has healed, gap studies to evaluate the distance between the upper and lower esophageal segments can be performed approximately every 2 to 4 weeks. The utility and optimal technique for gap measurement is controversial and not well standardized across institutions. In one approach, contrast is simply injected through the GT in the radiology suite while observing GERD of the contrast into the distal esophageal pouch. Alternatively, the study can be done in the operating room under fluoroscopy where a flexible neonatal (6 mm) endoscope is placed into the GT site and advanced under visualization into the distal esophageal pouch (*Figure 3*) (11,12). This method of retrograde esophagogastroduodenoscopy (EGD) typically offers the most accurate evaluation of gap length, as contrast can be injected directly into the distal pouch for optimal visualization of gap length at varying degrees of tension. It is important to consider that assessments of gap length may differ substantially, depending on the level of tension applied to the esophageal pouches. Although evaluating the elasticity of the esophageal tissue under tension provides valuable information regarding tissue approximation, standard off-tension assessments are the gold-standard for documenting gap length and tissue growth. Placing a radiopaque ruler under the patient during imaging may further assist with standardization of measurements for comparison to earlier timepoints. Written informed consent for publication of *Figures 2*,*3*,*5* was not obtained from the patient or the

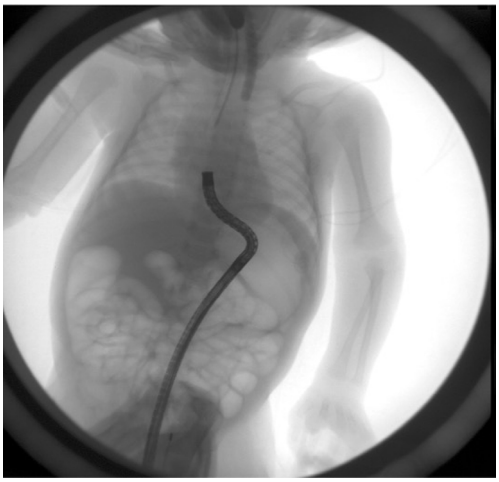

Figure 3 Gap study under general endotracheal anesthesia in a 1-month-old infant with long-gap esophageal atresia. In this anteroposterior fluoroscopic spot image, there is a transoral bougie dilator in the upper esophageal pouch. A small, flexible endoscope was placed through the gastrostomy site and advanced into the lower esophageal pouch. The esophageal gap was over 4 vertebral bodies.

relatives after all possible attempts were made.

The diagnosis of LGEA is synonymous with a prolonged hospitalization and worse long-term outcomes when compared to those without EA (4,13,14). There have been several expert opinion consensus guidelines established by the American Pediatric Surgical Association (APSA), as well as the European Reference Network for Rare Inherited Congenital Anomalies (ERNICA) and the International Network of Esophageal Atresia (INoEA) (2,3,15). Common principles include general avoidance of cervical esophagostomies and promotion of sham feeding to prevent oral aversion (2,15). To prevent recurrent oropharyngeal aspiration, a well-functioning Replogle catheter attached to suction is required and the head of the isolette should be elevated at all times.

Beyond management of LGEA as an isolated condition, these neonates require comprehensive care and collaboration with the neonatal intensivist team for optimization of respiratory and nutritional status, as well as consideration and management of potential comorbid conditions. Additional assessments including pre-operative flexible nasolaryngoscopy may be considered to establish a baseline and evaluate for vocal fold movement impairment, given the risk of recurrent laryngeal nerve damage during surgical repair.

Surgical repair

The ultimate goal in the management of infants with LGEA is to enable esophageal continuity to allow for oral feeding. There are multiple described operations for achieving this objective, which reflects some inherent limitations of each procedure as well as the heterogeneity of long-gap disease itself (16). Surgical repair involves either a primary anastomosis, attained either through spontaneous native growth with time or assisted by traction, or replacing the esophagus itself with another autologous gastrointestinal conduit, namely stomach, jejunum, or colon. Although evidence supporting best practices are generally of low quality, the two major principles of contemporary LGEA management are (I) the use of the esophagus for reconstruction whenever feasible and (II) continuity establishment without displacement of the gastroesophageal junction into the chest (e.g., partial gastric pullup).

Prior to esophageal repair, rigid tracheobronchoscopy by an expert operator is essential to confirm the anatomy of the aerodigestive tract. Key anatomy to completely rule out are the presence of a proximal tracheoesophageal fistula (Gross type B), severe tracheomalacia, and a laryngeal cleft (17). Flexible bronchoscopy through an existing endotracheal tube usually provides a suboptimal evaluation of proximal airway anomalies and is therefore discouraged as an alternative.

Delayed primary repair

Delayed primary repair (DPR) without traction remains the most commonly performed procedure to repair LGEA in contemporary studies (18-20). As the name suggests, these patients undergo a period of waiting after birth to allow for spontaneous growth of the upper and lower esophageal pouches, ultimately enabling a primary anastomosis of the native esophagus similar to those without long-gap disease (21). Spontaneous growth of the distal segment is thought to occur due to increased intraluminal pressure from reflux, while growth of the proximal segment is thought to occur through the swallowing reflex (22). Although most surgeons advocate for bolus GT feeds, preoperative serial bougieneage to increase upper esophageal segment length is has fallen out of favor in the United States, given the potential for perforation and subsequent intrathoracic infections. However, the practice of bougienage remains common in other countries, with recent studies from China finding it safe and effective in

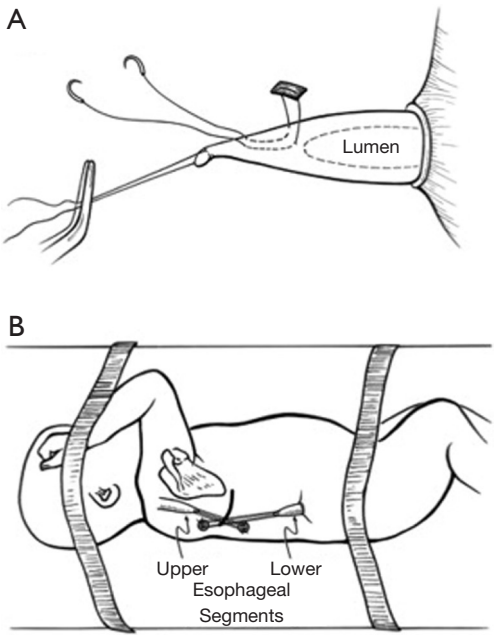

Figure 4 Schematic illustrations showing esophageal growth induction in long-gap esophageal atresia using external traction (Foker process). (A) Placement of pledgeted traction sutures. Deep bites are taken into the tip of each esophageal segment with care not to enter the lumen. (B) The sutures are brought out posteriorly on the chest wall. The upper segment sutures exit above the thoracotomy incision while the lower segment sutures are brought out below. Traction is placed on the segment by threading the sutures through silastic buttons on the skin surface. Small pieces of silastic tubing are placed under the sutures to increase the tension. © [2005] Elsevier Inc. Reprinted, with permission, from (37).

promoting esophageal elongation in LGEA patients (23,24).

The timing of DPR depends on the amount of esophageal growth that has been attained, as dictated by gap studies (18). Based on recent data, most LGEA patients are repaired using this approach at approximately three months of life (19,25). However, timing of repair should be dependent on patient- and resource-related factors, with some successful DPRs occurring later at 4–5 months (26,27), or even over one year of age (28). As a general rule, if the gap between the esophageal ends off-tension measures 2 or fewer vertebral bodies on imaging without prior surgical mobilization, then a successful primary repair can likely be achieved. Unfortunately, some patients do not experience robust spontaneous growth based on serial gap studies, and others have such "ultra-long" gaps (e.g., more than 5 vertebral bodies due to very short upper and/or lower pouches). In these scenarios, it is unlikely that they will be able to achieve a primary repair, even under high tension; therefore, traction-based surgical modalities or some form of esophageal replacement should be entertained (20,29,30).

The 2019 APSA consensus guidelines state that delayed primary repair should be considered the best initial option for LGEA (3). Open repair with complete mobilization of the upper and lower esophageal segments is usually required. A primary anastomosis under a considerable amount of tension is the norm. For those with advanced minimally invasive skills, thoracoscopy may be helpful for mobilizing of upper esophageal pouch up to the thoracic inlet (31). Some groups have advocated for additional cervical esophageal mobilization through a neck incision in selected cases to achieve a primary anastomosis. However, this maneuver does have its drawbacks, including devascularization of the proximal segment and increased risk for recurrent laryngeal nerve injury. Anastomotic leaks and strictures are common postoperative complications due to high tension and relative ischemia at the anastomosis (19). Other potential issues in these infants include GERD which may require a subsequent Nissen fundoplication and the need for long-term surveillance given the increased risk for Barrett's esophagus and malignant transformation (32-34). However, overall long-term outcomes, as defined by oral feeding, growth parameters, and respiratory function, are quite favorable (35).

Traction staged repair

Whereas spontaneous growth of the proximal and distal esophagus is sufficient to enable surgical repair of an esophageal gap in the majority of LGEA infants, staged procedures that impose longitudinal traction at the ends of the esophagus can lengthen the esophagus, thereby enabling a primary anastomosis in a shorter amount of time. Based on a recent multicenter cohort study, approximately 10% of LGEA infants undergo traction-based methods of repair (20).

Historically, the most widely known traction repair strategy was an external traction procedure known as the Foker Process. In the original description by Foker at the University of Minnesota, both ends of the esophagus are extensively mobilized, often through two separate thoracotomy incisions (36,37) (*Figure 4*). Once the inability to perform a primary anastomosis is confirmed, a series of partial thickness, horizontal mattress sutures are carefully placed through each esophageal pouch. Bovine pericardial pledgets are commonly used, and simultaneous intraluminal

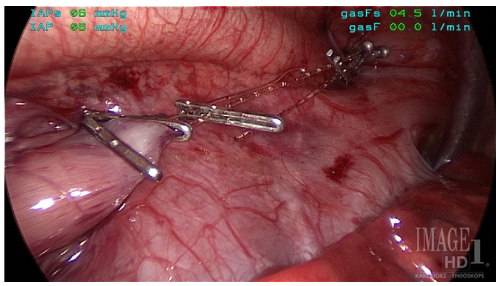

Figure 5 Intraoperative photograph demonstrating right thoracoscopic internal traction for long-gap esophageal atresia. Radio-opaque clips are placed to mark the ends of each esophageal segment. The suture is tightened twice weekly to induce esophageal growth until a primary anastomosis can be performed.

visualization with a flexible endoscope can be helpful to confirm partial thickness placement (38). The sutures, which are then marked with radio-opaque clips, and externalized in crossed fashion onto the posterior chest wall superior and inferior to the incision site. Anti-adhesion agents (e.g., sodium hyaluronate/carboxymethylcellulose) are placed into the thoracic cavity prior to rib closure to minimize the postoperative formation of pleural adhesions. Over a period of 1 to 3 weeks, increasing amounts of traction are placed on each esophageal segment by placing small pieces of silastic tubing under the externalized sutures. The location of the radio-opaque clips on serial radiographic studies helps to determine the feasibility and timing of performing a primary repair. Chemical paralysis is recommended for all patients during the external traction phase to prevent inadvertent tension being placed on the sutures.

The group at Boston Children's Hospital helped to popularized the Foker process, demonstrating its potential in many LGEA cases where primary anastomosis was previously not thought to be possible due to ultra-long-gap disease (39,40). Nevertheless, enthusiasm for the Foker technique has waned by some surgeons in recent years, in part due to the morbidity of multiple thoracotomies and chemical paralysis as well as issues related to tearing of the esophagus when placed under too much tension, leading to potentially lethal mediastinitis and sepsis (41,42). The distal esophageal segment tends to be more hypoplastic and is therefore more susceptible to tearing, leading to spillage of gastric contents into the mediastinum. Due to the many technical nuances specific to performing the operation, the associated postoperative care, and the high volume of cases required for surgical mastery of this approach, traction-based repair has also been difficult to replicate with the same success at other children's hospitals (43). Long-term issues after traction repair include recalcitrant anastomotic strictures that may require stricture resection. The vast majority of patients eventually require a Nissen fundoplication to address severe GERD (44). More recently, different approaches based on the Foker traction concept, including thoracoscopic internal traction, have been demonstrated in both European and North American institutions with substantial experience with complex esophageal procedures (*Figure 5*) (15,45). Due to the complexity and high risk for complications, including anastomotic leak or failure (43), traction procedures remain limited to relatively few specialist centers. Traction procedures should be viewed with a high degree of caution by centers without significant prior experience and should not be attempted by an inexperienced surgeons, with a recent study questioning the role of any lengthening procedures in LGEA (46).

In addition to surgical traction procedures, there are also emerging endoluminal technologies that have been devised as minimally invasive alternatives to induce esophageal growth. One notable approach is known as magnamosis, which promotes stretching using magnets placed endoscopically at the end of each esophageal pouch. The magnetic forces stretch the two blind ends towards each other, resulting in tissue approximation and necrosis between the two magnetic points, thereby creating an esophageal anastomosis without sutures (47). While a novel and minimally-invasive procedure, which also eliminates several operative complications including injury to the recurrent laryngeal nerve, magnamosis is most effective for gaps that are relatively short (e.g., 2–3 vertebral bodies). Magnamosis outcomes have also been mixed, with successful cases being hampered by recalcitrant strictures, many of which require serial endoscopic esophageal dilations and eventual surgical resection (48).

Esophageal replacement

The indications for esophageal replacement procedures as a treatment for LGEA remains somewhat controversial. Because replacement procedures are generally associated with higher operative risk and relatively poor long-term function compared to esophageal preservation approaches (30), esophageal replacement has increasingly been considered as a last line LGEA reconstruction option in the United States and elsewhere (2). While it is obvious

that the patient's own esophagus is the best esophagus, persisting with attempts to retain a native esophagus with no function and at all costs is futile and usually detrimental to the child's well-being. Accordingly, complete replacement may be required in patients with unusually long gaps after several months of observation (e.g., more than 4 vertebral bodies) or in cases where there is substantial loss of esophageal tissue following prior attempts at primary repair (27,49).

Given the high rate of anastomotic leaks associated with esophageal replacement procedures, many surgeons avoid intra-thoracic anastomoses because of the higher morbidity and mortality associated with acute mediastinitis. To achieve an anastomosis in the neck, there are three major routes: posterior mediastinal, transpleural, and retrosternal. The posterior mediastinum is normally favored because it the shortest distance for esophageal replacement (50), but other routes may be preferred in the setting of a hostile posterior mediastinum. It is also important to emphasize that the conduit should replace the esophagus as opposed to bypassing it. Complications in the retained esophagus after bypass procedures are well described and include chronic inflammation, Barrett's esophagus, mucocele, and empyema (30). Lastly, there are multiple conduit choices and approaches for esophageal replacement, each with their own advantages and disadvantages (30,51,52). In most cases, it is generally advisable to wait at least three months of age and a weight of 5 kg or more before committing to a replacement procedure (50). Those candidates in need of esophageal replacement who have recurrent aspiration episodes or have extensive co-morbidities may benefit from having a cervical esophagostomy prior to the replacement procedure.

Gastric conduits

There are two described approaches using a gastric conduit for esophageal replacement in LGEA. In the first approach, known as a gastric transposition or complete gastric pull up, the entire stomach is dissected from most of its attachments, relying on the right gastroepiploic vessels as its major blood supply (53). The second part of the duodenum is Kocherized to obtain maximum mobility of the pylorus. After the native thoracic esophagus is removed, the entire stomach (often hypoplastic and linear in shape) is then tunneled through the posterior mediastinum to enable creation of an anastomosis between the cervical esophageal remnant and the gastric fundus (*Figure 6*). The procedure can be performed without a thoracotomy or sternotomy incision and therefore has many similarities to the transhiatal esophagectomy without thoracotomy as performed in adults (50). Laparoscopic-assisted approaches have also been described (54,55). Regardless of the approach, a pyloromyotomy or pyloroplasty is performed to facilitate gastric emptying (56).

Gastric transposition has been a popular first choice for esophageal replacement in several countries, in part because it is a relatively straightforward operation in infants and requires only one anastomosis (3,27,57,58). Nevertheless, conduit emptying and GERD can still be problematic in many patients. Because the upper portion of the conduit tends to be relatively ischemic, there is a nontrivial leak rate and a stricture rate of over 40% (56). Since there is complete loss of the stomach as a reservoir to handle bolus feeds, infants generally require a feeding jejunostomy for stable enteral access with reflux (*Figure 7*).

The second major gastric conduit approach, referred to as a reversed gastric tube or gastric tube esophagoplasty, uses part of the native stomach (59,60). This technique involves the creation of a tubularized conduit along the greater curvature of the stomach based on the left gastroepiploic vessels (*Figure 8*). Several applications of a linear gastrointestinal stapler device are employed to create this tube. The right gastroepiploic and short gastrics need to be divided to create enough length. This approach has been criticized as a complete replacement procedure because conduit length can be inadequate given that the EA stomach is often severely hypoplastic in early infancy (30). The leak rate can be substantial due to the long suture line. Poor conduit peristalsis and chronic GERD can also be long-term issues.

Jejunal conduits

Jejunal conduits may be employed as interposition grafts for complete esophageal replacement, or more commonly, jejunum may be used in a Roux-en-Y approach, in which an esophagojejunal anastomosis is constructed in the neck, and a jejuno-jejunostomy in the abdomen. The use of a substernal jejunal interposition as a viable approach for complete esophageal replacement for LGEA has enjoyed a renaissance in the past decade largely due to refinements in operative approach and microvascular surgical techniques (2,49). Children with jejunal grafts tend to have excellent long-term functional outcomes when compared to other esophageal replacement patients due to strong conduit peristalsis as well as better size match between the small intestine and the native

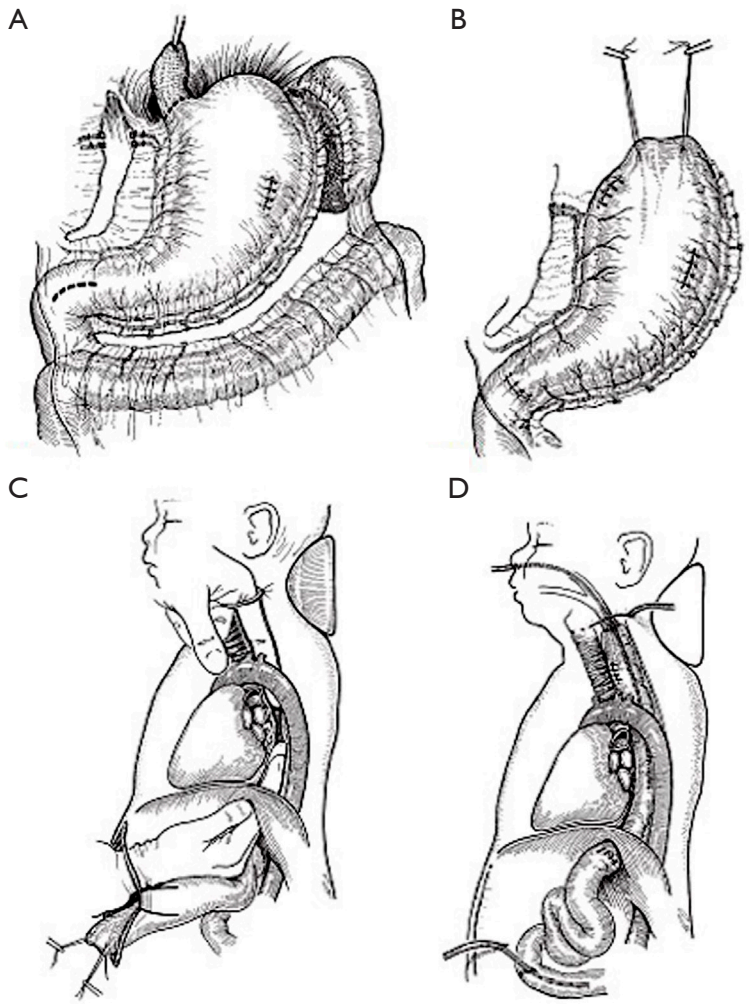

Figure 6 Technique of gastric transposition. (A) Mobilization of the stomach along the greater and lesser curvature, preserving the right gastroepiploic and right gastric vessels. The short gastric vessels are carefully divided, with care being taken to avoid trauma to the spleen. The left gastric vessels have been ligated and divided. The duodenum (to which the Kocher technique has been applied) and the site of pyloroplasty are indicated. The short stump of the esophagus in a case of isolated atresia is shown being mobilized from with the esophageal hiatus of the diaphragm. The gastrostomy site has been sutured. (B) Pyloroplasty has been completed, the distal esophageal stump has been resected, and the two sutures on the fundus of the stomach indicate the highest point of the stomach at the proposed site for the esophagogastric anastomosis. (C) Fashioning of the posterior mediastinal tunnel by blunt dissection from above by means of a cervical incision to mobilize the esophagostomy or expose the esophagus in the case of caustic injury and from below through the esophageal hiatus in the diaphragm. The dissection is done strictly in the midline in the prevertebral plan. (D) The final position of the stomach in the posterior mediastinum with the esophagogastric anastomosis in the lower neck and a pyloroplasty situated within the peritoneal cavity just below the esophageal hiatus. A jejunostomy tube has been placed for postoperative feeding. © [2017] Elsevier Inc. Reprinted, with permission, from (30).

cervical esophagus (61,62). However, jejunal interposition grafts are very technically challenging to perform in infants and young children due to the more tenuous blood supply (*Figure 9*). Complete esophageal replacement with jejunum requires "supercharging" the conduit via a microvascular anastomosis from the internal mammary vessels to small bowel mesenteric vessels; additionally, in these cases, three anastomoses of the alimentary tract are required. As with all esophageal replacement techniques, complications are common, and complete graft necrosis remains a possibility

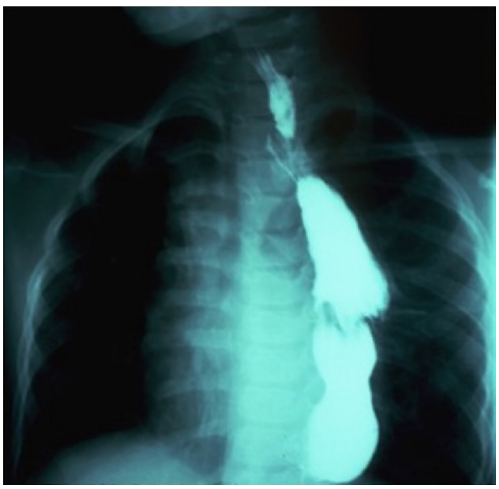

Figure 7 Radiograph demonstrating the postoperative appearance of a gastric transposition conduit in the posterior mediastinum after injection of oral contrast. © [2017] Elsevier Inc. Reprinted, with permission, from (30).

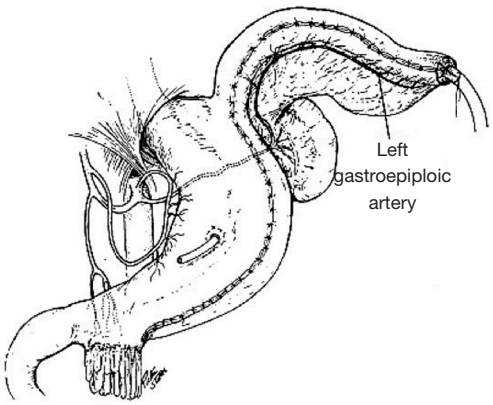

Figure 8 Reversed gastric tube conduit based on the left gastroepiploic artery. Oversewing of the staple line and gastrostomy tube placement are shown. © [2017] Elsevier Inc. Reprinted, with permission, from (30).

even in the hands of the most experienced surgical teams (30).

Colon conduits

The final conduit option in LGEA is large intestine, usually as an interposition graft based off the right colon (retrosternal) or left colon (retrohilar or posterior mediastinal) (*Figure 10*) (63). Historically, many surgeons preferred

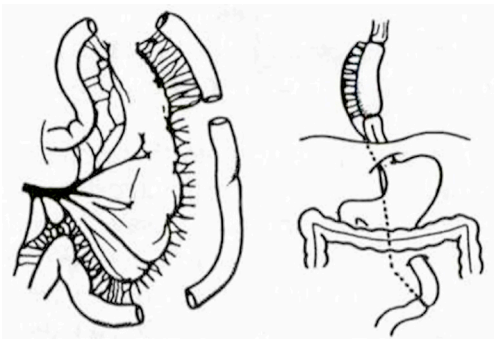

Figure 9 Pedicled jejunal interposition graft. In many cases, small bowel needs to be resected in order to obtain adequate length of the vascular pedicle to reach proximal esophageal segment. © [2017] Elsevier Inc. Reprinted, with permission, from (30).

colon conduits since a pedicled, isoperistaltic colon graft could be used for complete esophageal replacement (64). However, complete colon grafts for infants and young children have fallen out of favor at many institutions because graft function tends to diminish by adolescence or early adulthood. Affected patients can develop dysphagia, delayed emptying, and stasis, which increase the risk for regurgitation and aspiration (51). This loss of function is thought to be related to substantial longitudinal redundancy of the conduit over time, and based on some long-term series, up to one third of these patients will require conduit revision or another replacement later in life.

Tissue engineering

Tissue engineering represents an exciting new frontier in esophageal replacement; however, at present it remains highly experimental and in preclinical developmental stages in animal models. Numerous materials have been described for esophageal scaffolds, including collagen, decellularized extracellular matrices, and more recently patient-specific 3D printed models (65). Preliminary animal studies suggest tissue engineering may one day be a feasible approach to esophageal replacement, but human studies are early and require further optimization of materials and technique to become a functional conduit. Tissue engineered esophageal tissue currently lacks a vascular supply and innervation required for peristalsis (66). Despite this, advances in tissue engineering are encouraging and may one day become a practical option in LGEA repair.

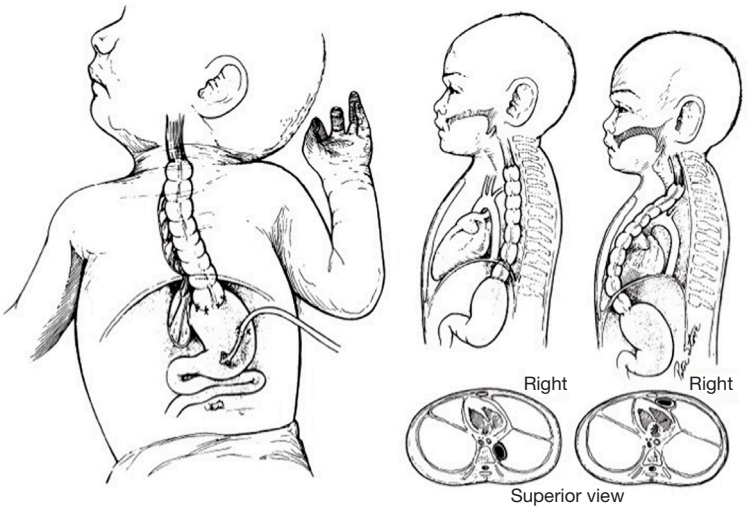

Figure 10 Colon esophagoplasty. The colon interposition may be placed in the retrohilar or substernal position. © [2017] Elsevier Inc. Reprinted, with permission, from (30).

Choosing a surgical approach

Surgical management of LGEA is highly complicated, as it involves decision-making based on patient-specific factors at multiple time points in a rare condition with which the majority of pediatric surgeons have relatively little operative experience. Continuous assessments of dynamic factors, including the patient's birth weight, gap length, respiratory status, and medical comorbidities including presence of other EA-associated anomalies, should all be carefully evaluated in the selection of a technique and timing of surgical repair. Therefore, we recommend that less experienced surgeons strongly consider early transfer of LGEA infants to a high-volume esophageal center to optimize patient outcomes. In addition to surgical expertise, such centers are equipped with comprehensive clinical care teams (including experienced pediatric anesthesia staff, pulmonologists, neonatal intensivists, respiratory therapists, and speech-language pathologists).

Postoperative management and long-term follow-up

Although mortality rates within the first postoperative year are relatively low (approximately 5%) (67), the post-operative morbidity associated with LGEA repair is high. Immediate post-operative complications related to surgical technique, including vocal cord paralysis and tracheomalacia, are uncommon but have high morbidity when they do occur. Anastomotic leaks occur in approximately 25% of cases and are typically managed nonoperatively, with reoperation required in only 2–6% of patients (67,68). Infants more commonly endure anastomotic stricture after DPR, which is less common after colonic interposition. Respiratory (20%) and gastrointestinal complications (56%) are common morbidities. Anti-reflux surgery within the first year postoperatively has been documented in 30% of those after LGEA repair (51,69).

Due to the high incidence of clinical and subclinical gastroesophageal reflux, it is recommended that all repaired LGEA patients should be maintained on medications for gastric acid suppression. Anti-reflux surgery, particularly in those undergoing esophageal lengthening by traction, may be helpful for alleviating symptoms (69). Although the actual incidence of esophageal cancer after LGEA repair is unknown, screening esophagoscopy for dysplasia and cancer should begin in adolescence and be repeated at minimum every 10 years due to high rates of subclinical reflux. Esophageal motility remains abnormal despite repair, and patients often experience long-term dysphagia with an increased risk of aspiration and long-term respiratory complications. Weight gain and nutrition may require close monitoring in LGEA patients, as delayed growth is often exacerbated by oral aversion induced by prolonged NPO status at birth (3). Despite these potential long-term consequences, a 2019 systematic review of the literature summarizing quality of life outcomes found no difference in quality of life for LGEA patients, compared to non-long-gap EA patients (3).

Conclusions

In summary, the management of infants with LGEA is a complex and technically challenging endeavor. Delayed primary repair is strongly preferred as the initial approach in management of LGEA in the United States as well as several European countries (19,28,67), and is supported by the American Pediatric Surgery Association recommendations (3). Should esophageal replacement be required in cases where salvaging the native thoracic esophagus is not possible, gastric conduits are the preferred approach, based on the relative simplicity of the operation, low postoperative morbidity, and long-term durability. Long-term follow-up for monitoring of swallowing function, nutritional status, aspiration/respiratory illnesses, gastroesophageal reflux, and associated comorbidities is essential in the comprehensive care of these complex patients.

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Footnote

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