Long Gap Esophageal Atresia

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Introduction

Long gap esophageal atresia (LGEA) is defined, functionally, as a distance between the ends of an atretic esophagus too wide to allow for primary repair without undue tension. Unfortunately, there is no consensus on anatomical definition. Some authors define LGEA as an esophageal gap that is greater than 2 cm, while others use the definition of a distance greater than two vertebral bodies. Thus, it is difficult to find consistency in recommended management strategies.

It is generally believed that the native esophagus is the best conduit for repair of LGEA and that every effort should be made to preserve the native esophagus, including esophageal lengthening techniques. However, esophageal replacement may afford patients an effective repair with the shortest time to initiation of oral feeds. Presently, there is no prospectively collected data investigating the superiority of esophageal lengthening versus esophageal replacement in patients with LGEA. Despite maximal efforts, esophageal preservation may not be possible in some patients, thereby mandating esophageal replacement. In this chapter, we will review the indications for esophageal replacement in children with LGEA.

1. Long gap atresia without tracheoesophageal fistula (LGEA-W/O-TEF)
   (a) Wait and watch 6–12 weeks with or without stretching (NG tube suction and gastrostomy) – if fails
      (i) Extra-/intrathoracic elongation
      (ii) Esophageal replacement

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   (b) Esophagostomy with gastrostomy
      (i) Extrathoracic elongation
      (ii) Esophageal replacement
   (c) Primary esophageal replacement
2. Long gap atresia with tracheoesophageal fistula (LGEA-W-TEF)
   (a) Ligate fistula only and delayed repair (sick baby)
   (b) Ligate fistula with intrathoracic elongation techniques
   (c) Ligate fistula with primary replacement
3. Failed previous primary repair, with or without a preceding lengthening procedure

1. *Long gap esophageal atresia without tracheoesophageal fistula*
   While strategies that preserve the native esophagus have been strongly favored over the years, these approaches are not always feasible. Delayed primary repair allows for potential growth of the atretic ends of the esophagus, allowing the esophagus to naturally lengthen. When this is not possible but use of the native esophagus is still desired, techniques have been employed to mechanically increase the length at one or both ends of the esophagus. Esophageal replacement remains an option for patients in whom primary repair is not feasible.
   (a) *Wait and watch 6–12 weeks with or without stretching (NG tube suction and gastrostomy)*
      Delayed repair of LGEA will be possible if the atretic ends of the esophagus grow, leading the gap to shrink. This approach is of particular benefit for very premature or critically ill infants who might not otherwise tolerate an extended procedure. Data has shown that the atretic esophagus may grow over time, particularly the distal esophagus in response to bolus gastric feedings. For that reason, an infant with LGEA without a fistula typically can undergo placement of a replogle tube in the proximal esophageal pouch plus gastrostomy creation to allow for intragastric feeds. The patient will then receive bolus gastric feeding for a period of time until the gap is narrowed enough to achieve a primary repair. Friedmacher and colleagues performed a meta-analysis of 451 newborns with LGEA managed by delayed primary anastomosis (DPA). They found that the initial gap length ranged from 1.9 to 7.0 cm. The mean time of DPA was 11.9 weeks (range 0.5–54), at which time the gap lengths had decreased to 0.5–3.0 cm [1]. However, some authors believe that DPA is a passive process and is typically futile [2]. Data are inconstant at best, with many patients experiencing delays lasting many months. Seguier-Lipszyc et al. reported their case series of ten patients with LGEA who were managed with DPA; the average time to perform the definitive repair procedure was 102 days (range 41–147 days) [3]. Despite a prolonged delay, only six of their patients successfully underwent esophagoplasty; the other four patients required colonic interposition.

(b) *Esophagostomy with gastrostomy*
An alternative method of managing a patient with LGEA without TEF is to create an esophagostomy of the proximal esophageal pouch and a gastrostomy. Efforts to accelerate esophageal lengthening have led to the creation of a multistaged extrathoracic esophageal elongation (ETEE) technique known as the Kimura technique (discussed in Chap. 6). In this approach, the proximal esophagostomy is sequentially translocated and stretched 2–3 cm at a time along the anterior chest wall, every 2–3 months until the gap is narrow enough to achieve a primary esophago-esophagostomy. Kimura and his colleagues published one of their case series of 12 patients in 2001; they were able to successfully accomplish the definitive esophageal reconstruction in all 12 patients with a mean number of 2.1 elongations (range 1–5 elongations) [4].

One other major advantage in creating an esophagostomy and gastrostomy is that the infant potentially can be discharged home and allowed to grow before the definitive procedure. In doing so, the patient can be "sham feedings" in a limited amount to avoid oral aversion from a prolonged NPO. Additionally, the patient will undoubtably be much bigger and physiologically stronger by the time he/she has esophageal continuity operation. The disadvantage to this approach is that, without forceful mechanical stretching, the proximal pouch will not lengthen which will most likely lead to an esophageal replacement.

(c) *Primary early esophageal replacement*
Primary early esophageal replacement may provide LGEA patients with an effective repair and the earliest time to initiation of oral feeds. According to some authors, the time it takes for a long gap defect to narrow sufficiently to allow for a primary anastomosis could be weeks to months. Furthermore, this primary anastomosis often requires significant tension, thus strengthening the benefit of early esophageal replacement.

In 2007, Gupta and colleagues published a large series of 27 neonates who underwent esophageal replacement for LGEA, with a mean birth weight of 2.32 kg (range: 1.86–3.0 kg) at a mean age 6.08 days at the time of repair [5]. The procedures were successful in all 27 patients. However, six neonates developed "ongoing serious chest infections" and three experienced lung collapse. The average length of time requiring ventilator support was 10.6 days (range: 2–40 days). Nine patients had esophagogastric anastomotic leaks that all healed spontaneously. The average hospital length of stay was 32.6 days (range: 9–87 days). Additionally, four patients died from sepsis and 11 of 23 (47.8%) patients exhibited duodeno-gastric reflux. Zeng et al. published similar outcomes, although it was a relatively smaller series [6]. Of 14 neonates who underwent repair at an average of 32 hours of life and with an average weight at 2550 gm, there were two deaths due to respiratory failure, representing a mortality rate of 14.3%. Seven of the patients in this cohort developed pneumonia, three patients developed early anastomotic leaks, and four developed anastomotic stricture requiring subsequent dilations. Additionally, 7 of the 12 remaining patients exhibited

GERD. Although neonatal esophageal replacement for LGEA is possible, results demonstrate a significant risk of complications.

2. *Long gap esophageal atresia with tracheoesophageal fistula*
It is generally believed by many that true LGEA only occurs in patients with pure esophageal atresia without TEF (type A). Saud Al-Shanafey et al. and Mariusz Sroka et al. reported that nearly 50% of LGEA in their respective series had a coinciding fistula as well [7, 8]. Similarly, a meta-analysis of 44 articles by Friedmacher revealed that 257 of 451 patients (57%) with LGEA had tracheoesophageal fistula (TEF) [1].

Both the presence of an associated TEF and its respective location are important features that may affect the decision regarding the management of patients with LGEA. The presence of a TEF mandates an early intervention for ligation of the fistula, which greatly impacts future surgical planning. When LGEA is determined at the time of fistula ligation and primary repair is not possible, there are three options to be considered.

(a) *Ligate fistula only and delayed repair (sick baby)*
As in the case of patients with LGEA without TEF, delayed primary repair in a patient with LGEA and fistula remains a viable treatment option. These patients must undergo an early ligation to avoid serious consequences such as aspiration pneumonia and severe gastric distension that may lead to respiratory failure. This approach can also be employed in critically ill infants or very premature infants as a temporizing measure. Petrosyan et al. retrospectively evaluated their cohort of patients with esophageal atresia and TEF at very low birth weight – less than 1500 gm [9]. When compared with their primarily repaired group, their delayed (staged) repair group resulted in much better outcomes with leak rate 0% vs 50%, stricture rate 33% vs 81%, and much lower incidence of postoperative pneumonia.

Like patients with pure esophageal atresia, these patients would most likely receive a gastrostomy for nutritional supports and promote distal esophageal growth. The time at which the definitive procedure is considered depends on how the patient is doing and the type of the procedure is entertained.

(b) *Ligate fistula with intrathoracic elongation techniques*
There have been number of techniques used over the years to lengthen the esophagus in an attempt to preserve the native esophagus. The extrathoracic approach like Kimura's procedure has been discussed elsewhere. One approach to immediately gain esophageal length is a circular or spiral myotomy. Vizas et al. at Hospital for Sick Children demonstrated that circular myotomy could produce at least 1 cm in length without interfering with perfusion [10]. However, in their 3-year follow-up, the esophagus showed ballooning at the myotomy sites [11]. In addition to this concern, cutting through the esophageal muscle may lead to denervation and dysmotility. Interestingly, Sumitomo and colleagues reported that, manometrically, the circular myotomy sites have normal contractions and propagations compared to those of non-myotomy esophagus [12].

The Foker technique is currently a popular approach that relies upon external tension to induce esophageal lengthening for primary esophageal reconstruction. Mochizuki et al. reported a case series comparing the outcomes of the Foker technique to their historical results. Although the number of cases captured was small, the results demonstrated a clear advantage in the cohort undergoing the Foker technique, based on a number of important parameters—average weight at surgery (2.0 kg vs 2.5 kg), mean day of operation (28 days vs 227 days), and time to full feeds (76 days vs 686 days) [13]. In 2015, Bairdain and colleagues published a larger series of patients undergoing the Foker procedure [14]. Of their cohort, 27 patients underwent a primary Foker repair and 25 patients underwent a secondary repair because they had an initial surgery elsewhere. Of the primary repair cases, the median time to anastomosis was 14 days, compared to 35 days for the secondary repair group, with excellent outcomes. Nasr and Langer performed a systematic review and cumulative meta-analysis in which they reviewed the outcomes of 71 patients who underwent Foker procedure of the 451 children with LGEA. They noted that the Foker procedure was associated with a significantly lower risk of complications including leak, stricture, and gastroesophageal reflux (GERD), plus a shorter time to definitive anastomosis [15]. While esophageal lengthening techniques are effective for LGEA, these techniques are associated with increased complication rates in the setting of larger defects. When elongation techniques fail, esophageal replacement is the next best option.

(c) *Ligate fistula with primary replacement*

Despite with all the efforts, saving the native esophagus may not be possible. In such a situation, the esophageal replacement is the only option. Work has been done in an attempt to establish criteria to better predict which patients with LGEA will eventually require an esophageal replacement. One such criterion is the presence of an "ultra-long gap" or a gap that is greater than 3.5 cm in length.

Some authors have attempted to subcategorize the location of the fistula as a predictor in needing esophageal replacement. Kolvusalo and his colleagues reported that patients with a fistula at the carina tended have long gap defect, resulted in poor outcomes and a higher rate of needing esophageal replacements [16]. Furthermore, patients with long gap type C atresia had outcomes similar to patients with type A or type B atresia. They concluded that the need for esophageal replacement was more common in patients with type C malformations with more distal fistulae (i.e., those with longer esophageal gaps), but was not as common as for patients with type A or type B atresia. These "predictors" have yielded mixed results and yet not been validated.

3. *Failed previous primary repair, with or without a preceding lengthening technique*

Although there have been reports of successful primary repair of these patients, the risks of complication significantly increase in this population [17]. With the

recent success and popularity of the Foker procedure, another common indication for an esophageal replacement is a failed previous primary repair, with or without a preceding lengthening technique. This might come in the form of a severe anastomotic leak or recalcitrant strictures [18]. According to Upadhyaya et al., the risks of stricture and/or leak following primary repair were directly proportional to the starting esophageal gap length, with the rates of complication significantly increasing for gaps greater than 2 cm [19]. Mild anastomotic leaks often resolve spontaneously with conservative management. However, severe anastomotic disruption can cause life-threatening infection and requires surgical intervention which includes re-exploration, either attempted repair of the leak or diverting esophagostomy, and eventual future esophageal replacement [20]. The majority of esophageal strictures can be managed with esophageal dilations. Recalcitrant strictures that are resistant to repeated dilations may require either segmental resection, if the stricture area is short, or esophageal replacement, if the stricture segment is long.

Conclusion

Long gap esophageal atresia (LGEA) in neonates is one of the most challenging surgical dilemmas a pediatric surgeon may face. Currently, there is no consensus regarding the best surgical approach for the condition. Giving the recently favorable outcomes of the intrathoracic lengthening techniques and their popularity, they should be considered as a first-line treatment to increase esophageal length to achieve primary anastomosis. Although esophageal replacement techniques have shown good results, they do have a high morbidity associated with them. Several attempts at elongation trying to save the native esophagus are also associated with increased morbidity. Decision of prolonged attempts at elongation techniques vs esophageal replacements should be based on individual needs and the resources at the location of care.

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