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Long-gap esophageal atresia

Hester F. Shieh, MD, Russell W. Jennings, MD*

Department of Surgery, Boston Children's Hospital, Harvard Medical School, 300 Longwood Ave, Fegan 3, Boston, MA 02115

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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.

* Corresponding author.
E-mail address: russell.jennings@childrens.harvard.edu (R.W. Jennings).

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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