

## Esophageal Atresia; Short-gap and Long-Gap Management and Outcome

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### Abstract

The diagnosis and treatment of patients along the esophageal atresia (EA)/tracheoesophageal fistula (TEF) spectrum continues to evolve. Despite refinements in fetal imaging, prenatal diagnosis remains difficult, all though a dilated proximal esophageal pouch helps increase the predictive value for EA. Prematurity, cardiac and chromosomal defects are the most important predictors of overall outcome. Short-gap EA is usually associated with a distal TEF and less risk for prematurity. For this reason, thoracoscopic repair is becoming more frequent in these patients, with the promise of lower surgical morbidity when performed by advanced minimally invasive surgeons. Large comparative studies suggest equivalent outcomes when compared with open thoracotomy.

To date there is no consensus on the preferred method of esophageal reconstruction in infants with long-gap EA. Tension-induced growth of the esophagus now serves as a viable alternative to conduit reconstruction, although it remains an ongoing debate because it is a relatively new technique, still it is difficult to argue against the flexibility of former procedure and the inherent advantages of preserving the native esophagus with a primary esophageal repair.

**Keywords:** *Esophageal Atresia; Tracheoesophageal Fistula; Long Gap; Thoracoscopic Repair; Management*

### Introduction

Esophageal atresia (EA) and tracheoesophageal fistula (TEF) has an incidence of 1:3500 live births, with close to half of the patients presenting concomitant congenital anomalies requiring surgery. The surgical management of infants born with EA with or without TEF is one of the major triumphs of pediatric surgery in the twentieth century [1], beginning with the first successful primary repair of a neonate with EA/TEF, by Dr Cameron Heigh in 1941 [2]. Since this initial report, surgical and neonatal care advances have steadily improved survival rates in babies within the EA/TEF spectrum. Children with EA/TEF, birth weight greater than 1500g and no major cardiac anomalies have a survival rate of more than 98% [3].

### Embryology

During the fourth week of gestation the ventral portion of the foregut gives rise to the trachea and lungs, whereas the esophagus develops from its dorsal aspect [4]. Environmental factors that have been implicated in EA/TEF include exposure to methimazole, diethylstilbestrol, exogenous sex hormones, infectious diseases, maternal alcohol and tobacco use, maternal employment in agriculture or horticulture.

ture fields, first-trimester maternal diabetes, and advanced maternal age [5]. Hereditary factors seem to play a minimal role in the origin of EA/TEF. The risk that a second child would be born with EA in the same family is of 1%. Between 10% and 25% of infants with EA have three or more additional nonrandom anomalies, including vertebral, anorectal, cardiac, tracheal, esophageal, renal and limb abnormalities, which form the VACTERL spectrum [6]. An additional 10% are associated with genetic syndromes, including trisomy 18, 21 and 13.

### Prenatal diagnosis

Despite continuing advances in ultrasound imaging, the diagnosis of EA/TEF in the fetus remains difficult, allowing for 10% to 40% of patients being identified antenatally [7-9]. The most common finding suggestive of fetal EA is an absent or small stomach bubble in conjunction with polyhydramnios, with a positive predictive value for EA between 44% and 56% [8,10]. The upper pouch sign, which consists on identifying a dilated proximal esophageal pouch helps increase the predictive value for EA, unfortunately it is still difficult to detect and not visible until the third trimester [11].

The degree of prematurity and the presence of associated anomalies, particularly cardiac and chromosomal defects are the most important predictors of overall outcome. Prematurity has an increased incidence among infants with AE/TEF when compared with the general population, most likely in relation to polyhydramnios from fetal esophageal obstruction [12].

### Short-gap atresia

Any neonate with excessive oral secretions, feeding intolerance, and/or respiratory difficulties at birth should raise concern for EA. Failure to pass an orogastric tube into the stomach with the presence of abdominal bowel gas on plain film radiography suggests and EA with distal TEF. This is the most common form of EA/TEF and is found in about 85% of cases. Because the lower esophagus reaches the trachea, the gap between the esophageal segments is usually short.

### Initial management

Patients with EA and distal TEF have a high risk of aspiration pneumonia, which increases the urgency for surgical repair. To minimize this as risk as much as possible aspiration precautions must be initiated before surgery, including a Repogle-type orogastric tube to suction within the esophageal pouch, elevation of the head of the bed at 30 - 45° and acid suppressive therapy.

Positive pressure ventilation should be avoided whenever possible, because gastric distension secondary to air passing through the TEF can increase respiratory embarrassment, with no way of allowing gastric drainage.

All neonates with EA/TEF should undergo a preoperative diagnostic workup looking for VACTERL anomalies. The most important component in this evaluation is the echocardiogram, because 35% of these patients will have cardiac abnormalities. It is also very important to determine de side of the aortic arch during the echocardiogram, found on the left side in 95% of the cases. For this reason, the most common operative approach for the repair of short-gap EA is a right thoracotomy [1,13].

The goals of operative therapy are: 1) to close the fistula between the trachea and the lower esophagus and 2) to establish continuity of the esophagus. In most cases, the two ends of the esophagus are in proximity to each other, and a single-layer primary esophageal anastomosis can be performed. Feedings can be initiated through a transanastomotic tube 2 to 3 days after the operation in most cases, once bowel movement is present. Seven days after the procedure the orogastric tube can be removed and an esophagogram is performed to check the integrity of the anastomosis. A leak can be seen in 15% of cases [1,14]. If there is no leak oral feedings can be started. In the presence of a leak parenteral nutrition must continue, broad-spectrum antibiotics started, and a chest tube placed (in case it wasn't left during surgery) allowing drainage and the leak to spontaneously close in most cases.

Although many EA/TEF infants will have gastroesophageal reflux (GERD) after repair, in short-gap cases only 15% to 20% will be severe and persistent, requiring an antireflux procedure, such as a Nissen fundoplication. Significant postoperative strictures are also not uncommon but, if they occur, can be managed with serial endoscopic dilatation.

### Thoracoscopic repair for short-gap atresia

Pediatric thoracotomy incisions, even when muscle-sparing approaches are used, have been associated with long-term complications, including shoulder weakness, winged scapula, and thoracic scoliosis [15,16]. Thoracoscopic repair was first described in the late 1990s [17-19]. It also allows for superior visualization compared to open surgery. It is technically demanding and requires an experienced surgeon [20]. To minimize risk, candidates include those who weigh more than 2.5kg, and have minimal cardiopulmonary disease.

Clinical outcomes and postoperative complication rates have been reported to be comparable to those after traditional thoracotomy [21-23]. Strictures are slightly more common after thoracoscopic surgery, and 30% of patients will require one or more endoscopic dilations of the anastomotic site. Operative times during thoracoscopic repair tend to be longer, particularly during the learning phase of the procedure, and conversion rates to open thoracotomy have been reported in up to 32% of cases [24]. For this reason, minimal invasive surgery in smaller or more premature babies should only be performed by experienced surgeons.

### Long-gap atresia

When an orogastric tube cannot be passed into the stomach of a neonate found to have a gasless abdomen on the initial plain film, a long-gap atresia (defined as more than three vertebral bodies) should be suspected [12]. The most common form of long-gap atresia is an isolated EA without a TEF. It is the second most common form of EA, although only 8% will present this variant.

### Initial management

Neonates with an isolated EA are at lower risk of aspiration pneumonitis, nevertheless isolated EA infants are more challenging to repair because of the distance between the proximal and distal ends of the esophagus. Early primary repair of the esophagus in these cases is usually not possible [25].

The initial management includes screening for VACTERL anomalies, placement of a Repogle-type orogastric tube and elevation of the head of the bed at 30 - 45°. Traditionally, a cervical esophagostomy was advocated for long-gap EA and placement of a gastrostomy tube to allow feeding and evaluation of the lower esophagus. This is no longer recommended because the esophagostomy will only make subsequent attempts at a primary esophageal repair more difficult, and that gap measurement with contrast media is not reliable as the lower pouch might be underestimated in the absence of reflux [26] and new techniques allow for faster enteral feeding which advocates for parenteral nutrition during the first days of life.

The next routine procedure is an unstressed gapogram. In case placement of a gastrostomy is decided, the study would involve injecting radio-opaque contrast into the lower segment while a catheter is placed into the upper pouch. To allow for a more reliable measurement of the gap, a neonatal endoscope is recommended to help define the length of the lower esophageal segment on a gapogram study, but gentle pressure must be applied on the delicate newborn tissues to avoid distorting the true gap length.

There is no consensus on the preferred method of esophageal reconstruction to date [27-33]. According to the length of the lower esophagus, the appropriate method of repair will be defined. Fundamentally, the preservation of the native esophagus should be aimed before considering any replacement technique [34-36]. Repair techniques may range from a primary anastomosis under tension for shorter gaps to esophageal growth induction (Foker process) or conduit reconstruction for longer gaps. In the former situation, a delayed

primary repair without any specific intervention in 1 to 3 months' time is possible because of spontaneous growth of the esophagus from swallowing attempts (proximal) and gastric reflux (distal) [37].

### Conduit reconstruction

In ultra-long-gap EA (five or more vertebral bodies), a delayed primary repair is usually not possible. One option for definitive reconstruction in these children is esophageal replacement with an autologous interposition graft derived from stomach, colon, or jejunum [3,14,38]. These operations, which continue to remain in vogue at many referral centers worldwide, have been used successfully by pediatric surgeons for decades [25,39-41].

### Esophageal growth induction (Foker process)

Introduced by Dr. John E. Foker in 2005 [42] it is an inherently appealing approach to esophageal reconstruction because it preserves the native esophagus enabling a true primary repair [43]. Although the application of tension to the esophagus is not an entirely new concept, many of the previously described strategies for lengthening the esophagus, including hydrostatic pressure, serial bougienage, magnets, internal traction, among others [44-46] have never gained widespread popularity because of unreliable esophageal growth or high complication rates. The recent ERNICA Consensus on the Management of Long-Gap EA (2020) considered Esophageal Growth Induction as a novel technique that shows promise [47].

For the Foker Process a limited posterolateral thoracotomy or thoracoscopy are performed [42,48], horizontal sutures are placed through the esophageal wall and marked with clips adjacent to the atretic ends of the esophagus to allow subsequent assessment of the gap length on daily radiographs. The sutures are then externalized through the posterior chest wall in a crossed fashion and tied under tension. As esophageal growth occurs, tension is reestablished daily. Once the gap is less than two vertebral bodies in length the patient undergoes another thoracotomy with primary repair of the esophagus. Serial radiographs show the gap closing at an average rate of 0.5±0.2 cm per day, allowing for primary repair within 3 to 31 days, depending on the length of the gap [12].

The overall functional results in patients with this procedure include anastomotic strictures, which are common but usually amenable to endoscopic dilatation. Severe GERD and esophagitis are present in practically all patients despite the fact that the gastroesophageal junction remains below the diaphragm, more than 95% will require a Nissen fundoplication [49].

### Conclusion

The EA/TEF spectrum is a relatively frequent anomaly. Even with refinements in fetal imaging prenatal diagnosis is still not frequent. Prematurity, cardiac and chromosomal defects continue to be the most important predictors of outcome. Treatment has also evolved, allowing for a growth in minimal invasive techniques both for short-gap and long-gap EA, with similar results when compared to open surgery when performed by experienced surgeons. Tension-induced growth of the esophagus seems like a viable alternative in long-gap EA with the advantages of preserving the native esophagus with a primary esophageal repair, although long term evidence is still pending.

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