Difficult esophageal atresia: Trick and treat

Andrea Conforti, MD, Francesco Morini, MD, Pietro Bagolan, MD*
Neonatal Surgery Unit, Department of Medical and Surgical Neonatology, Bambino Gesù Children Hospital – Research Institute, Piazza S. Onofrio 4, 00165 Rome, Italy

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ABSTRACT

Although most patients with esophageal atresia (EA) and tracheo-esophageal fistula (TEF) may benefit from “standard” management, which is deferred emergency surgery, some may present unexpected elements that change this paradigm. Birth weight, associated anomalies, and long gap can influence the therapeutic schedule of the patients with EA/TEF and can make their treatment tricky. As a consequence, detailed information on these aspects gives the power to develop a decision-making process as correct as possible. In this article, we will review the most important factors influencing the treatment of patients with EA/TEF and will share our experience on the diagnostic and therapeutic tips that may provide pivotal help in the management of such patients.

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Introduction

In the last 50 years, the concept of esophageal atresia (EA) in pediatric surgeon’s mind has not changed much. Willis J. Potts, pioneer of pediatric surgery, once stated, “to anastomose the ends of an infant’s esophagus, the surgeon must be as delicate and precise as a skilled watchmaker. No other operation offers a greater opportunity for pure technical artistry.” Since then, the approach to EA patients has been poorly modified, with no consensus on preoperative assessment, perioperative treatment, and postoperative care.

EA, with or without tracheo-esophageal fistula (TEF), remains the most common congenital anomaly of the esophagus. The current mortality is minimal1,2 in spite of the frequent association with other malformations and the reported decline in trainee experience.3 The survival improvement is probably dependent on several factors such as advances in neonatal intensive care, anesthesia and intraoperative monitoring, refined surgical approach and techniques, management of complications, parenteral nutrition, and antibiotics. Despite the outcome being generally good, EA may still represent a challenge to pediatric surgeons.4 Nowadays, the centralization of these patients and their rigorous and homogeneous preoperative assessment are not yet a common standard.

The focus of surgeons with EA subspecialty interest shifted recently to the “more difficult” cases. Two main aspects can be included in the definition of “more difficult”: (1) general patients’ characteristics such as prematurity and associated life-threatening cardiac and bowel anomalies and (2) specific surgery-related issues including laryngotraheal anatomy, esophageal anatomy, and redo surgery. For these babies, the optimal timing of procedure and preoperative assessment, the definition of the so-called “long gap,” and the possible approaches to difficult or impossible anastomosis (including technical refinements) are of paramount importance but still source of controversies.

In this article, we will endeavor to browse all these topics to provide the most recent data and knowledge regarding perioperative diagnostic assessment and therapeutic approaches, to highlight helpful refinements in difficult cases, and to suggest a possible personal flowchart as a guide.

General patients’ characteristics

In the vast majority of patients with EA/TEF, operative repair is urgent but not an emergency, and overrides the treatment of other associated problems. However, in specific clinical conditions, it may be necessary to step out from this paradigm. The most important patients’ characteristics that can change the treatment schedule include birth weight (and prematurity) and some associated anomalies, which are also the most important factors in determining survival in historic risk classifications1,5

Very low-birth-weight (VLBW) neonates (< 1500 g) with EA/TEF represent a distinct subgroup of patients that need particular therapeutic attention. Prematurity and low birth weight are relatively frequent in association with EA/TEF, especially in patients without distal TEF, due to reduced amniotic fluid absorption and subsequent polyhydramnios. VLBW infants are prone to develop respiratory distress syndrome and require mechanical
ventilation. In case of decreased lung compliance, increased airway resistance, or need for mechanical ventilation, the fistula may act as a low-resistance vent through which air may preferentially pass into the esophagus. As a consequence, the loss of ventilation and the progressive gastric distension will worsen the respiratory distress syndrome, also facilitating gastroesophageal reflux and inhalation, increasing the risk of gastric perforation with high mortality rate. For VLBW neonates with EA/TEF and respiratory distress, several maneuvers have been suggested to prevent or to stop the air leak through the fistula, including gastric division, silastic banding of the distal esophagus, and distal positioning of the endotracheal tube with the bevel pointing anteriorly, antegrade or retrograde occlusion of the fistula with a Fogarty balloon, water-seal gastrostomy, and high-frequency ventilation. Most recent series suggest early thoracotomy with ligation of the fistula. In the emergency setting, the approach should be transthoracic to expedite the operation. If the patient is stable, the extrapleural approach is preferable due to the possibility of more severe consequences of an anastomotic leak after a transthoracic approach. The fistula should be ligated in continuity, ideally with non-absorbable sutures. After the fistula is closed, if the patient is stable and the anatomy is favorable (see section Preoperative assessment of EA), the fistula may be divided and a primary anastomosis can be attempted. Otherwise, a delayed primary anastomosis should be programmed. In VLBW infants, the esophageal tissue is extremely fragile. Therefore, lengthening maneuvers should be performed very cautiously as, with the words of Dr. Rickham, “the tissue paper thick walls of the 2 segments will disintegrate.” Minimal handling is recommended on the ends of the esophagus completing the anastomosis with no more than 8 stitches. When the fistula is ligated in continuity and a delayed anastomosis is programmed, this should be attempted within 1–2 weeks, as failure to do so may result in recanalization of the TEF. Other authors wait until the patient is clinically stable and reaches 2 kg. In such cases, to avoid the risk of refistulization, the fistula is ligated, divided, and anchored to the spine with moderate traction if needed. In addition, a cervical esophagostomy (CE) should be considered to make the nursing of these patients easier and to reduce the risk of saliva aspiration (see section Difficult/Impossible anastomosis).

VLBW infants with EA/TEF and respiratory distress are at high risk of gastric rupture. In a large series of 623 patients, 6 developed a gastric rupture, 5 of which were preterm. If such a complication occurs, tension pneumoperitoneum may lead to diaphragmatic splitting, further reducing the lung compliance and deteriorating cardiorespiratory status. Emergency abdominal decompression should be performed, quickly followed by laparotomy, and emergency gastrostomy for air leak control with a Foley catheter in the lower esophagus. In the series from Maae et al., this had an immediate beneficial effect on ventilation allowing the authors to proceed to the division of the fistula through an extrapleural approach and primary esophageal anastomosis.

Associated anomalies are very frequent in patients with EA/TEF. The cardiovascular system is the most commonly affected with a prevalence reaching 50%. Major cardiac anomalies may cause a hemodynamic instability so severe that it does not allow prolonged thoracic surgery. In these cases, emergency treatment of the cardiac anomaly may represent a priority over esophageal surgery. Healey et al. report that out of 118 patients with EA/TEF, 23 required delayed primary repair. The presence of a life-threatening associated cardiac anomaly was the second most frequent cause of primary anastomosis delay after long-gap EA. In a small series of 6 patients with both EA/TEF and congenital cardiac anomalies, Hayashi et al. report delayed primary anastomosis in 4 of them. They had a gastrostomy performed the first day of life, and the anastomosis was delayed between 3 days and 3 months of life. All above data indicate that early echocardiography is strongly recommended to tailor the treatment specifically to each patient.

Right aortic arch (RAA) and right descending aorta may cause operative difficulties in patients with EA/TEF. Ideally, preoperative cardiac ultrasound should be also performed to define the situation of the aortic arch. However, preoperative recognition of a RAA may be difficult with a low detection rate even in experienced hands. As a consequence, the surgeon should be prepared to face a RAA even with a negative preoperative ultrasound scan. With preoperative diagnosis of RAA, a left thoracotomy is suggested. Other authors also achieved satisfactory results via a right thoracotomy in all patients with a RAA, and they support this approach in all EA patients.

Duodenal obstruction (DO) in the form of duodenal atresia or stenosis may be associated with EA/TEF in up to 6% of patients. Several treatment options have been proposed for this association. Spitz et al. recommend that when a TEF is present, the primary approach should be to divide the fistula and repair the EA, performing a wide gastrostomy to decompress the stomach. For patients with pure EA, they recommend duodenoduodenostomy and gastrostomy leaving a transanastomotic feeding tube. Others suggest to repair the DO first followed by the delayed repair of EA/TEF, ideally after 1 week. The advantages of staging the repair with DO first would be to give the opportunity for improvement in growth and pulmonary function and to allow some resolution of the delayed gastric emptying of DO, thus stressing less the esophageal anastomosis. Finally, in patients with pure EA, it offers the possibility for the distal esophageal segment to elongate. As a general principle, when a distal TEF is present, the priority is to avoid the risk of inhalation. Thus, emergency decompressive gastrostomy, immediately followed by the closure of the fistula, is recommended. Then, if clinical conditions are stable, esophageal anastomosis and duodenoduodenostomy should be considered in the same operation. If the clinical conditions remain critical, the esophageal and/or duodenal anastomoses should be delayed after stabilization. In patients with no distal TEF, duodenoduodenostomy and gastrostomy are the first surgical steps. The esophageal gap can be measured (see section Preoperative assessment of EA) and a delayed esophageal anastomosis planned.

Congenital diaphragmatic hernia (CDH) is very rarely associated with EA, with an estimated incidence of 1:200,000. The coexistence of these anomalies carries a very poor prognosis, with a survival rate below 30%. The presence of a distal TEF poses serious management problems when associated to CDH as it generally limits the possibility of positive pressure ventilation. On the one side, the escape of gasses through the fistula reduces the ventilation to the lungs, and on the other side, it leads to progressive inflation of the bowel loops that further compress the hypoplastic lungs. For that reason, urgent fistula closure is necessary. A delayed esophageal anastomosis can be done when stabilization is obtained, at the time of CDH repair.

Chromosomal anomalies are found in up to 10% of patients with EA. The most common is Edwards syndrome (trisomy 18), followed by Down syndrome (trisomy 21), and more rarely Patau syndrome (trisomy 13). The occurrence of a chromosomal anomaly in a patient with EA may represent an ethical quandary. This is well illustrated by the case of Baby Doe who was born in April 1982 with Down syndrome and associated EA. Currently, that case would not pose the same ethical problems, as the presence of Down syndrome is not regarded anymore as a motivation for treatment withhold. Nowadays, Edwards syndrome and Patau syndrome are facing the same debate. In such syndromes, with poor long-term survival, surgical treatment of associated EA may be seen as cause of unjustified, prolonged suffering to the infant and a waste of the scarce medical resources. A recent report...
describes the results of surgical treatment of EA in 24 patients with Edwards syndrome. The authors performed palliative surgery in 9 patients (gastrostomy or jejunostomy with/without TEF ligation) and complete repair (primary or delayed) in 15 patients. Of the 24 patients, 17 achieved enteral feeding (3 by mouth) and 3 could be discharged home. The median survival was 44 days, and the principal factor associated with death was the association of congenital heart defects. These data pose the question of the quality of life that surgical treatment can allow to patients with chromosomal syndromes considered with no prospectus of long-term survival, and to their families and if this is a price that the “scarce medical resources” can pay. It seems reasonable that surgical treatment should be tailored to each single patient's conditions to warrant the best quality of life possible, independent of its length. Aims of surgery (palliative or not) should be to give the possibility to reach enteral feeding and, if possible discharge the child home, in the best interest of the child and the family.

**Preoperative assessment of EA**

In the last few years, preoperative tracheobronchoscopy (TBS) gained attention to evaluate the presence of proximal TEF. Nonetheless, beyond confirming the presence and number of esophageal fistula, TBS offers the possibility to evaluate vocal cord motility, to assess the presence of other foregut specific associated anomalies (tracheomalacia, tracheal clefts, etc.), and to preoperatively define the esophageal gap. Thus, endoscopic assessment is essential to define the surgical plan.

Preoperative evaluation of esophageal gap length is a critical part of assessment in patients affected by EA even though no consensus has been gained yet.

Preoperative gap measurements promote comparability between centers, and limit intraoperative findings of unsuspected difficult cases. Only a few authors have tried to find a reproducible way to assess preoperatively esophageal gap length, and in most of the cases, this evaluation is limited to those cases of EA without distal fistula.

The common opinion persists that only EA without distal TEF may present with a gap “long.” However, the evidence demonstrates that type C is the most frequent variant of long-gap EA (LGEA). Firstly proposed to measure esophageal gap was the infusion of water-soluble contrast via the gastrostomy tube to evaluate the lower esophageal pouch. Neither special equipment nor anesthesia is needed, but the gastroesophageal junction must be incompetent to allow the reflux. The insertion of a Hegar dilator through a gastrostomy into the lower pouch was also proposed as simple and safe.

However, some authors consider to blind pass a rigid dilator through the gastroesophageal junction into the distal pouch difficult. Therefore, others proposed to measure esophageal gap using a flexible endoscope to intubate the lower pouch, but with this technique, it is not possible to quantify and standardize the boost pressure applied, losing information on elasticity of the lower pouch. Common limit of above-mentioned techniques is that they are not suitable for cases with distal TEF.

Preoperative CT evaluation of EA patients was also proposed, but it involves significant exposure to ionizing radiation and possible risk of radiation-induced cancer. Additionally, in up to 20% of cases, the fistula could not be located limiting the indication to evaluate complex associated malformations (cardiac, tracheal, etc.).

Since 2004, we developed a preoperative standardized approach to measure the gap, reducing intraoperative “surprises” such as laryngotracheal anomalies and vocal cord dysfunction in all spectrum of EA.

TBS is performed in all patients to define the presence of proximal TEF (defining/excluding Gross type B/D), vocal cord motility, and foregut anomalies. Subsequently to anatomical definition, TBS is used to immediately measure the gap in types C/D: a 10 Ch radiopaque Nelaton probe is inserted under pressure into the upper pouch. At the same time, the tracheoscope's tip is placed at the level of the tracheal opening of the distal fistula. A chest fluoroscopy shows the distance, thus the gap, between them (Figure 1A and B).

A chest fluoroscopy shows the distance, thus the gap, between them (Figure 1A and B). When a cernal fistula is present, the option of its occlusion (by a 3.5-Fr Fogarty catheter; the balloon inflated with 0.2 mL of saline solution) is discussed with anesthesiologist to make the mechanical ventilation easier, by avoiding gastric overdistension and gastroesophageal reflux.

Instead, gap measurement is delayed 15 days after gastrostomy fashioning in types A/B EA. A number 4 or 5 Hagar dilator is inserted into the lower esophagus through the gastrostomy and pushed upward to evaluate esophageal elasticity. The thrust applied is measured with an electronic device (dynamometer) connected to the Hegar (Figure 1C and D). The force applied to the dynamometer ranges between 250 and 300 g. The gap is then measured, under fluoroscopy, both without and under pressure. A comparable method is used for patients with cervical esophagogastrostomy, marking the stoma with a radiopaque device or with a small quantity of contrast medium (Figure 1E).

**Difficult/impossible anastomosis**

In children with LGEA, opinions differ significantly about the “best” solution for the child. “Efforts should be made to conserve the native esophagus since it is the best conduit for esophageal reconstruction” remains, also in difficult cases. Maintaining the own esophagus has been reported as a challenging surgical goal to achieve, and a primary anastomosis is often described as “impossible” only on the basis of surgeon's skills. As previously described, no consensus has been gained on esophageal gap measurement, even though a standardized and comparable method has been recently reported. Thus, the surgical choice currently remains subjective depending on individual training and experience. Therefore, there is a clear need for high-quality, comparative studies. Besides, 2 further main points must be highlighted to fully understand the challenge of LGEA. Firstly, Type C EA must be included in the long-gap/tricky group of patients indeed, since a meta-analysis found that the most common variant of LGEA is EA with distal fistula (56%).

Secondly, patient referred after a failed attempt of primary anastomosis are a further source of “secondary long-gap,” and more “difficult” anastomosis. Referral centers should nowadays expect 20–30 new LGEA/100 consecutive EA observed: 8–10 as patients referred after failed attempt; 8–10 as type A/B EA (reported prevalence of “pure” EA), and 8–10 as type C/D LGEA. Thus, the expected number of long-gap/difficult anastomosis cases makes this problem not so rare for experienced EA subspecialty interested surgeons and for referring centers.

Various ingenious techniques have been developed to bridge LGEA and maintaining the native esophagus. Most “esophagus-sparing” techniques consist of gradual traction of esophageal segments, inducing growth and reducing the gap. Some other refinements have been developed to reduce the anastomotic tension (esophageal flaps and myotomy). These different technical options are considered and shortly discussed, to propose a decisional algorithm for neonates affected by EA and LGEA either primary or after a failed attempt.
Simple delay

Time is commonly employed to allow the segments to grow, and we believe that this is true in EA without distal TEF in which the lower esophagus and stomach are hypoplastic such as any post-atretic intestinal loop. Following this rationale, bolus gastrostomy feedings are progressively given to promote spontaneous growth by meal and gastric air reflux into the lower esophagus. Spontaneous growth of the esophageal segments may occur during the first 3 months, but there are almost no experimental or scientific data about this phenomenon. At intervals of 3–6 weeks or until the infant is approximately 3.5–4 kg is considered an appropriate period of time for “wait-and-see” only program. Performing serial gap measurements, every 15 days, allows to decide on delayed anastomosis, possible as one-step procedure when the remaining gap is ≤ 2 vertebral bodies in our personal experience.

Esophageal dissection

The esophageal pouches should be extensively mobilized. Early in the thoracic dissection, several 5–6/0 silk stay sutures are placed on both pouches to aid mobilization with minimal handling of the tissues. When, despite extensive mobilization, the remaining gap is deemed too long to bridge, further dissection is possible through a cervical incision. Particular attention is given to avoid trauma to the blind end of the pouches (possible further traction and/or flap). Once the esophagus has been fully mobilized, the gap is measured again.

Traction and growth

When the residual gap is still not able to be bridged, the technique exploited by most authors is traction and growth procedure. Lengthening of the neck by traction is an ancient practice for cosmetic reasons; just think of the giraffe women of the Kenyan tribes who wear rings to lengthen it. Other examples of stretching and growth procedures are surgical techniques of limb traction, such as the Ilizarov procedure, or the use of skin expanders, causing the skin and soft tissue growth, as well as autologous bowel-lengthening procedures. Therefore, traction has been considered a good system to induce esophageal growth and elongation. In a rat animal model of EA, continuous traction on the esophagus has been shown to increase esophageal mass preserving histopathological morphology of the esophagus without major tissue damage. Many esophageal-lengthening procedures, such as hydrostatic pressure, serial bougienage, and magnets, among others, have been reported even though they have never gained widespread popularity.

External traction (ET. Foker’s technique)

In 1994, Boyle et al. reported preliminary results on systematic primary repair of ultra-long-gap EA (3.5 cm or greater) without lengthening procedures in 8 consecutive patients, concluding that although tension may contribute to strictures and gastroesophageal reflux, primary repair results in a clinically functional native esophagus. A few years later, the same group reported their personal experience on 70 consecutive EA (not only “long gap”) concluding that the esophageal anastomosis can withstand considerable tension, and allows a reliable true primary repair for the full EA spectrum. In 2003, the group from Montreal published 3 consecutive LGEA treated with definitive esophageal anastomosis 10–14 days after birth, adopting the same technique of ET and Skarsgard reported 2 cases of LGEA successfully treated with ET technique as well. Currently, the Foker procedure, or its

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Fig. 1. Gap measurement in different types of esophageal atresia. A: Diagram showing gap measurement for type C EA: the tube in the proximal pouch, flexible bronchoscope tip at the level of the distal fistula. B: Chest X-ray (or fluoroscopy) to measure the gap in type C EA: 3 vb gap. C: Diagram showing the dynamometer connected to the Hegar dilator to exactly measure the boost pressure. D: Chest X-ray (or fluoroscopy) is performed and the gap expressed in vertebral bodies (vb): 1 vb gap. E: Gap measurement in a referred patient with cervical esophagostomy.

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modifications, has been successfully used in more than 100 infants with LGEA worldwide.54,67–72

In the largest single author experience on 63 consecutive patients with a gap greater than 2.5 cm, the ET allowed primary repair of the esophagus in all cases within 3–31 days (mean 14 ± 7 days), with a rate of gap shortening of 0.53 ± 0.2 cm/day.73 In 16% of patients, a further thoracotomy was required to either reconfigure or replace traction sutures. Failures and complications have been reported after ET but also accounted for the need of a learning curve.73,74 Recently a large case series based on traction and growth approach, without external traction, arrived at the same conclusions.39 This report includes 57 cases with LGEA (≥ 3 vertebral bodies), 27 of which were referred cases after a failed attempt. Only 1 referred patient, with CE/no lower esophagus and cardia stretch, ultimately required esophageal substitution. Referred LGEA patients, showed a higher rate of CE (44% vs 3%).39 In 2013, Sroka et al.38 reported their comparable experience on 15 LGEA cases (5–14 cm), 9 of which with a previous CE fashioned. Only 2 referred patients with CE required esophageal substitution due to postoperative complications.

Early complications after traction procedure and anastomosis are anastomotic leaks in up to 50% of patients, which were mostly minor leaks and subsided spontaneously. Major disruption and failure of conservative management with need for drainage or reoperation was reported in up to 15% of dated series and in 1–13% of the most recent reports.39,58,73,75,76 Lastly, esophageal replacement for unsatisfactory results after delayed anastomosis was required in 14%.

To critically evaluate the existing literature on this topic and to compare the ET Foker technique to the more conventional delayed primary anastomosis, Nasr and Langer performed a cumulative meta-analysis.77 Overall, 71 infants undergoing the ET procedure were compared with 451 children utilizing delayed primary anastomosis. The initial gap length was 5.4 cm (range: 3–12.5 cm). No study reported data about the preoperative gap length. The Foker procedure, despite the risk of bias due to the retrospective cohort, was associated with a significantly lower risk of complications (primary outcome of the study) and with a significantly shorter time to definitive anastomosis.77

Since delayed primary anastomosis, with either “wait-and-see” delay or with “traction to induce growth” to bridge the more difficult gap, provides good immediate- and long-term functional results, a concerted and rigorous effort to achieve an end-to-end esophageal anastomosis should be made before considering esophageal substitution.

Extrathoracic esophageal elongation (ETEE, Kimura’s technique)

Ken Kimura firstly reported ETEE in a child with long-gap EA whose parents’ requested reconstructing the esophagus using the native conduit.78 Since then, ETEE has been widely used as a “traction” procedure, either as a technique of choice in patients who had been electively treated with CE for a variety of reasons (primary esophagostomy) or as an esophagus “rescue” procedure for those babies in which an esophagostomy had been performed because of a previous failed attempt (secondary esophagostomy).39,58 Advantages of the ETEE are to maintain the native esophagus, to allow early oral alimentation, and to shorten hospital stay while waiting for the final esophago-esophagostomy. Preferably, the esophagostomy is created on the right side of the neck. The upper pouch is dissected as proximal as possible and brought to the skin. To make the possible subsequent dissections of the esophageal stump easier, the proximal esophagus can also be wrapped with Goretex81 (W.L. Gore and Associates, Inc., Flagstaff, AZ, USA).79 At each elongation step, the neck is flexed and the esophagus gently stretched caudally and possibly anchored to the pectoralis major fascia with 2 or 3 absorbable sutures. Neither thoracotomy nor prolonged sedation and muscle paralysis is needed. Only few studies report comparison of esophageal-lengthening techniques. Long-term outcomes are limited, and patient selection is frequently different. Results of studies on ETEE are reported in Table. Recently, Sroka et al.38 pooled experiences of 2 European centers to compare ET and ETEE techniques. They conclude that ET of both pouches results in a high rate of primary repairs in children with LGEA and no previous esophageal operations. However, the combination of ETEE and ET, applied to patients with a CE already fashioned at a previous failed attempt of esophageal anastomosis, results in an equivalent rate of primary repair, but the number of complications increase significantly. Therefore, the ETEE has been considered the first choice for those selected patients with a previous cervical esophagostomy.78

Upper esophageal flap (UEF, Gough and Bianchi)

MH Gough initially reported the anterior full-thickness flap of the upper pouch to bridge a long gap, in 5 consecutive patients with a difficult anastomosis.81 None required reoperation, but 2 or more dilatations were necessary in all infants.81 After 15 years, the same group reported the outcome (mean = 2 years) of 15 consecutive LGEA in which an UEF had been performed to preserve the native esophagus. Complications included leaks (27%); strictures (87%), 2 of which requiring a segmental resection; gastro-esophageal reflux requiring fundoplication (20%); recurrent TEF (13%); and esophageal motility incomodation (60%). Overall, 70% of patients achieved a normal growth, leading the authors to conclude that despite the considerable morbidity, flap procedure reduces the need for esophageal substitution with a satisfactory outcome.82 The UEF was routinely performed, irrespective of gap length, in 25 patients by Davenport and Bianchi.77–79 Despite complications, 28% leakage (all treated conservatively) and 8% recurrent TEF, 92% of infants retained their esophagus and had no long-term swallowing problems, leading the authors to conclude that UEF is able to provide sufficient tissue to bridge long-gap defects (up to 4 vertebral bodies). In 2006, Castanon et al.80 reported UEF in 3 cases with LGEA, concluding that it represents their first choice for treatment of LGEA since it allowed esophageal preservation, despite re-intervention in 2 patients for persistent stenosis. Morabito et al. arrived at the same conclusion, reporting their experience on 67 cases (6 type A LGEA) treated with UEF and lower pouch augmentation. Minor leaks (spontaneous resolution) was found in 11 (16%); anastomotic stricture in 2 (3%), which eventually required redo-anastomosis; and recurrent TEF in 5 (7.5%), all requiring a second operation.83 Lastly, our group has recently reported the use of esophageal flap in selected patients with LGEA. Comparison between patients with LGEA requiring (6 patients) or not (13 patients) UEF to bridge a type C LGEA showed that only stricture length was significantly higher in neonates with an esophageal flap (2.2 vs 1.0 cm).84 At subsequent long-term follow-up, only 1 patient, out of 9 treated with UEF, required segmental resection of a persistent stenosis, unresponsive to dilatation, 13 months after surgery.89

Minimal invasive surgery (MIS) and difficult anastomosis

In 2005, Holcomb et al.85 reported thoracoscopic repair of EA, with results comparable to those of babies undergoing an open thoracotomy repair, from a multicentre study including 104 children, leading to the conclusion that MIS represents a natural operative evolution and can be safely performed in babies with EA, by experienced surgeons. However, 8 years later, a survey92 noted that open thoracotomy is still the preferred surgical approach by 94% of surgeons. Increasing experience in the thoracoscopic repair
of EA has led some surgeons to attempt MIS repair for difficult cases. A total of 4 patients with LGEA approached with thorascoposcopic elongation were reported by van der Zee.\textsuperscript{86} 2 of them needed jejunal interposition due to postoperative complications. Martinez-Ferro\textsuperscript{87} in 2010 reported his flexible MIS approach to LGEA with no short- or long-term results. Rothenberg recently published his own 10-year experience with thorascoposcopic repair of 62 cases of EA, including 9 patients with LGEA.\textsuperscript{88} Overall, 61 procedures were completed thorascopically. One of 9 patients with LGEA needed open conversion. In the immediate postoperative course, 3 leaks were observed, 2 (22%) in babies with LGEA, all resolved spontaneously. However, neither surgical technique to acquire approximation of the pouches nor short- or long-term results are specifically reported on the LGEA group of patients. The reported advantages of the thorascoposcopic approach are less pain, less/no damage of shoulder musculature and consequent shoulder asymmetry with scoliosis in later years, and better cosmesis. When complications occur, a broad armamentarium of alternative techniques should be available to address the problems. On the other hand, nowadays the same advantages can be obtained with a minimally invasive open surgery including minimal axillary or thoracic incision, muscle-sparing, avoidance of costal synostosis. Thus, conclusions of these 2 approaches should be drawn only comparing the most recent series. Moreover, thoracotomy presents the specific advantage of using an extrapleural approach, with its several benefits in these difficult cases, with a higher risk of leaks due to the high anastomotic tension.

The limited number of physiologic studies concerning neonatal thorascoscopy seems to demonstrate that brain oxygenation could be impaired for some time after the operation as a consequence of anomalous gas exchange with CO\textsubscript{2} reabsorption and possibly thoracoscopy seems to demonstrate that brain oxygenation could be impaired for some time after the operation as a consequence of anomalous gas exchange with CO\textsubscript{2} reabsorption and possibly prolonged increased superior vena cava pressure. In a pilot randomized controlled trial\textsuperscript{89} including 20 patients with EA (10 patients) and congenital diaphragmatic hernia (10 patients), the intraoperative PaCO\textsubscript{2} and pH in the thorascoposcopic group were significantly worse compared with that in open approach group (83 vs 61 mmHg and 7.13 vs 7.24, respectively). Moreover, the duration of hypercapnia and acidosis was longer in thorascoscopy compared with the open approach.\textsuperscript{89} Considering only patients affected by EA (10 cases), hypercapnia and acidosis showed more extreme values in the thorascoposcopic group. However, the difference was not significant. Considering these aspects, some authors concluded that thorascoscopy should not be performed for the correction of LGEA, as this defect requires more invasive dissection, and there may be difficulty with the anastomosis, leading to an increased duration of surgery.\textsuperscript{86}

In conclusion, delayed esophageal anastomosis, either primary or after traction, avoids esophageal replacement in almost 100% of cases in most recent series,\textsuperscript{39,58,73} and the possible failed attempt to save the native esophagus does not invalidate a later esophageal replacement. The proposed LGEA algorithm we are now adopting is a practical sketch to assess the patient, classify the anomaly and the gap preoperatively, and follow a strict surgical course, according to the patients’ clinical status, aimed to safely save his own esophagus (Figure 2).

Present and future

Use of biodegradable and biocompatible scaffolds for tissue engineering the esophagus promises to be an effective regenerative strategy. Tissue-engineered esophagus may eventually be a promising replacement in the future, and esophagus repairs in animal models, and even clinical tests, are continually being attempted. However, some key problems like stricture requiring dilation, little or no muscle regeneration, vascularization, and innervation in the implants need to be addressed before
engineered esophagus can be a viable esophageal conduit for surgical replacement in the human being. Totonelli et al. recently summarized that acellular scaffolds can be either transplanted with the aim of being repopulated by host cells or seeded prior to transplantation. When acellular scaffolds are used as a patch or to replace a short tubular defect, they allow epithelial and partial muscular migration, whereas when employed for long tubular defects, the results are poor, leading to an increased rate of stenosis and mortality. Results in animal models, which have used seeded scaffolds, strongly suggest that seeding of both muscle and epithelial cells on scaffolds prior to implantation is a prerequisite for complete esophageal replacement. Although esophageal tissue engineering potentially offers a real alternative to conventional treatments for severe esophageal disease, important barriers remain that need to be addressed and solved.

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