Review Article

Postoperative complications and long-term outcomes of tracheoesophageal fistula repair

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Abstract: Congenital tracheoesophageal fistula (TEF) is part of the esophageal atresia complex and can be associated with multiple congenital anomalies. With advancements in anesthesiology and neonatal intensive care support, the survival rate of infants with TEF has surpassed 90% and at present, the rare cases of postoperative mortality are typically due to extremely low birth weight and/or to the presence of associated comorbidities, such as congenital heart defects. TEF morbidity is partly secondary to the surgical repair and partly intrinsic to the anatomical anomaly. Postoperative complications include anastomotic leak, stricture, and recurrent fistula. Moreover, some patients develop long-term outcomes that include gastroesophageal reflux (GER) and esophagitis, dysphagia, tracheomalacia, vocal cord disorders, and risk of esophageal malignancy. Many aspects of the management of these short- and long-term sequelae remain controversial and lack a standardized approach. Nonetheless, it is widely accepted that patients with TEF require a multidisciplinary treatment and long-term follow-up. The purpose of this review was to evaluate the recent literature on TEF postoperative complications and long-term outcomes, with focus on the incidence, medical and surgical treatment, and controversial aspect of TEF postoperative management. Understanding the long-term functional outcomes helps to re-evaluate the impact of the old classical approaches of treatment and to optimize the modern medical and surgical techniques and follow-up management.

Keywords: Tracheoesophageal fistula (EA/TEF); tracheoesophageal fistula repair; anastomotic leak; anastomotic stricture; recurrent fistula

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Introduction

Tracheoesophageal fistula (TEF) is an abnormal communication in one or more places between the esophagus and the trachea. TEF can present as a congenital anomaly, which is usually associated with esophageal atresia (EA), or as an acquired condition, which is typically iatrogenic or secondary to trauma, malignancy, or infectious processes. Herein, we report the most recent literature on postoperative complications and long-term outcomes of infants with the congenital form of TEF (*Table 1*).

Postoperative mortality

With advancements in anesthesiology and neonatal intensive care support, the survival rate of infants with TEF has surpassed 90% (1,2). Nowadays, the rare cases of postoperative mortality are usually due to extremely low birth weight (<1,000 g) and/or to the presence of associated comorbidities (3,4). This is in line with a risk classification model proposed by Lewis Spitz in 1994 and based on birth weight and the coexistence of cardiac malformations (5):

- Group 1: Birth weight >1,500 g without presence

Table 1 The most common postoperative and long-term complications in TEF patients

Postoperative and long-term complications	Incidence (%)
Mortality	5.4–7.5
Postoperative complications	
• Anastomotic leak (overall)	6–8
• Major leak	3.5
• Anastomotic leak in long gap EA	58
• Anastomotic stricture (overall)	10–59
• Anastomotic stricture in long gap EA	44
• Associated with trans-anastomotic feeding tube	56
• Recurrent TEF	5–10
Long-term outcomes	
• Dysphagia	75–100
• Oral dysphagia	36
• Pharyngeal dysphagia	75
• GER	40–75
• Eosinophilic esophagitis	21
• Metaplasia	21–43
• Tracheomalacia (overall)	90
• Tracheomalacia requiring surgical intervention	10
• Vocal cord dysfunction in H-type EA	50
• Vocal cord dysfunction in other types of EA	4.6
• Malignancy	Lower than general population

TEF, tracheoesophageal fistula; EA, esophageal atresia; GER, gastroesophageal reflux.

of major cardiac anomaly;

- ❖ Group 2: Birth weight <1,500 g or presence of major cardiac anomaly;
- ❖ Group 3: Birth weight <1,500 g and presence of major cardiac anomaly.

In 2006, Spitz *et al*. retested the validity of their classification and reported an increase in survival mainly in group 2 and 3, which they interpreted as due to the improvements in intensive care rather than in surgical techniques (6). More recently, the Midwest Pediatric Surgery Consortium has published the outcomes of their series of infants with EA/TEF and reported that the mortality (7.5%) was significantly associated with the presence of congenital heart disease (3).

Postoperative morbidities

Anastomotic leak

Definition and presentation

One of the most common postoperative complications associated with EA/TEF repair is anastomotic leak. Leaks are divided into minor and major according to the degree of anastomotic disruption (7). The distinction can be made based on radiological findings and/or clinical presentation (7-9). Major leaks typically have most of the contrast medium dribbling from the anastomosis into the mediastinum, and present rapid deterioration with life-threatening pneumothorax or fluid collection. These leaks are likely due to a disruption of >25% of the anastomosis.

Conversely, minor leaks have most of the contrast drained through the distal esophagus, with only a little amount dribbling out of the anastomosis, and patients are usually asymptomatic.

Incidence

The reported incidence of anastomotic leak ranges from 6% to 58% (3,8,10-14), with a higher incidence reported in the group of pure EA. However, it is well established that the leak rate depends on whether an esophagogram is routinely performed postoperatively, as in this case even minor leaks are detected in asymptomatic patients. Similarly, the higher rates reported in patients with pure EA could be attributed to the fact that all these patients routinely receive a post-operative esophagram, so that even clinically insignificant leaks are recorded (10).

Etiology

Numerous risk factors have been considered in the literature. As leaks are more common in patients with long-gap EA compared to those without long gap, tension between the esophageal leading to blood supply compromise is considered the most significant factor (3,15). Some surgical techniques, such as the two-layer anastomosis and the Livaditis circular myotomy, have also been associated with higher rates of anastomotic leak than other procedures (8,9,15). Evaluation of the suture materials showed that braided silk sutures were associated with a significantly increased risk of anastomotic leak compared with polyglycolic acid or polypropylene sutures (16). Conversely, the approach employed for surgical repair does not seem to affect the leak rate, as shown in a meta-analysis of retrospective comparative studies, where no differences were reported between the thoracoscopic and open approaches (17). Another risk factor described in literature as associated with high incidence of leak rate is the use of postoperative non-invasive ventilation, especially if longer than 48 h (18).

Management

To prevent anastomotic leaks, some surgeons opt for postoperative muscle paralysis, endotracheal intubation with mechanical ventilation, and neck flexion (PVF) in patients with an esophageal anastomosis under tension. The PVF protocol, whose duration varies between 2 and 5 days (19), has been shown to reduce the anastomotic leak rate (20). Classically, minor leaks discovered with a routine esophagogram are successfully managed conservatively by parenteral nutrition, broad-spectrum antibiotics, observation, and chest tube drainage if required (11,13,14,16,21). The use of anticholinergic agents has also been described to reduce excessive salivary secretion, control the leak, and promote spontaneous closure of the leak (21). Conversely, major anastomotic leaks should instead be operated on while the anastomotic edge is still fresh for re-suturing before inflammation and necrosis can cause tissue damage. The rationale for the intervention is to prevent serious and potentially fatal complications, such as mediastinitis, mediastinal abscess, sepsis, and tension pneumothorax (1,11,22).

Anastomotic stricture

Definition and incidence

Anastomotic stricture is a common morbidity following EA/TEF repair with an incidence ranging between 10% and 59% (3,14,23,24). The anastomotic narrowing seen on early postoperative contrast esophagograms is not to be considered as a stricture, rather as postoperative changes due to tissue edema, and it has been proven not to be associated with the development of a stricture in the long-term (15,25,26).

Etiology

Common risk factors implicated in stricture formation include anastomotic tension, anastomotic leak, and gastroesophageal reflux (GER) (1). Moreover, any vascular compromise during the mobilization of the lower esophagus which has a segmental blood supply from the aorta and the intercostal vessels, may yield to ischemia at the esophageal anastomosis, which may lead to stricture formation (7). Patients with long gap EA undergoing a staged repair have been reported to be at higher risk of stricture formation over those who undergo primary repair (22,27). Nonetheless, the Midwest Pediatric Surgery Consortium reported no differences in stricture rate between patients with and without a long gap EA (3). Several studies have shown that the risk of esophageal stricture is significantly higher in infants that develop an anastomotic leak compared to those without leak, possibly due to local inflammation and scar formation (11,13,22). GER has always been considered a significant predisposing factor for stricture formation (15,22,28,29) and recent evidence shows that a third of EA/TEF patients with clinico-pathological evidence of eosinophilic esophagitis had a history of recurrent stricture (30). Classically anastomotic strictures

have also been associated with the choice of suture material, with braided silk sutures being considered a significant predisposing factor to stricture formation compared to polyglycolic acid and polypropylene sutures (28). However, a more recent study demonstrated no difference in the incidence of anastomotic stricture between monofilament and braided sutures as well as between absorbable and non-absorbable sutures (31). The use of magnets to achieve esophageal continuity, also known as magnamosis, has also been reported as a cause of stricture, given the higher need for esophageal dilatations than in patients treated with conventional anastomosis techniques (28). The use of a trans-anastomotic feeding tube has also been suggested as a cause for anastomotic stricture, with a higher stricture rate in patients managed with such a tube compared to those managed without (32). It remains controversial whether anastomotic strictures have similar incidence after open or thoracoscopic TEF repair. One study indicated that the thoracoscopic surgery increased the risk of stricture in 40% versus 16.3% stricture rate after open thoracotomy (27). With the development and improvement of thoracoscopic technique recent reviews of the literature and meta-analysis shows no difference in stricture rates in patients between the thoracoscopic and open approaches (3,17,33).

Management

Anastomotic esophageal strictures are commonly treated with dilatation (26). Bougies were popular in the past and have largely been replaced by balloon dilatations, which can be done under fluoroscopic or endoscopic guidance (26). Timing of the dilatation has also changed over time: in the past, some centers were advocating for esophageal dilations to be performed routinely, in order to prevent the development of a stricture. There is now good evidence showing that postoperative dilatations are necessary only if the patient is symptomatic (34). This approach has been corroborated by a recently published international survey that reported the lack of consensus on the management of anastomotic strictures post-EA/TEF repair and formulated some advisory statements, including performing dilatation procedures in symptomatic patients (35).

In cases of recurrent strictures, esophageal dilatations can be associated with topical application of mitomycin C, intralesional steroids injection, endoscopic treatment with stents or using an endoscopic knife (25,36,37). The use of Mitomycin C, an alkylating agent used in cancer chemotherapy, is based on its anti-fibrotic properties, whereby it inhibits wound healing by downregulating the expression of extracellular matrix proteins, and prevents postoperative scar formation (38-40). Similarly, the use of triamcinolone acetonide, a synthetic corticosteroid with an unclear mechanism of action, is based on its known effects in the treatment of hypertrophic scars of the skin and keloid. However, the success of intralesional triamcinolone acetonide injections has been reported as inconsistent in improving the stricture, and its use accompanied by potential complications including intramural infection, esophageal perforation, mediastinitis, pleural effusion, and adrenal suppression from exogenous systemic steroid administration (41).

Stenting has been reported as an alternative method for refractory esophageal stricture treatment (26,41-43). In the literature, different types of stents have been described based on their mechanisms of action: self-expanding plastic stent; retrievable, fully covered, self-expanding metal stents; and self-expandable, biodegradable stents that allow passage of food inside the stent (41,42). However, stenting has little success in the pediatric population and is often associated with complications and with the disadvantage of the lack of specific sizes for children. A recent systematic review showed that esophageal stents may have a role as a bridge to definitive surgery, rather than achieving complete resolution (44).

When conservative management of recurrent anastomotic strictures fails, surgery in the form of stricturoplasty, stricture resection and reanastomosis, or substitutive interventions (gastric pull-up, colon or jejunal interposition) should be considered (45). The timing of surgery remains controversial. A survey of the International Pediatric Endosurgery Group members reported that some surgeons would opt for surgical intervention after three dilations (46). Conversely, a single-center study of 103 consecutive patients showed that surgery is best predicted if >10 dilations are required (47).

Recurrent TEF

This is a severe complication occurring in approximately 5–10% of cases, most often between 2 and 18 months following initial repair (12,13,45,48-51).

Etiology

Recently, Smithers *et al*. proposed a classification of recurrent TEF based on etiology and anatomy (45): (I) congenital TEF, which persist after TEF repair as they were missed or the repair was incomplete; (II) recurrent TEF that occurs in the same location after primary repair of TEF;

(III) acquired TEF which occurs in a new location on either the airway side (from the trachea to bronchi to pulmonary parenchyma) or the digestive side (esophageal anastomosis or colon or gastric conduit), or both. In case of a congenital TEF, one should consider the possibility that during the first surgery a rare variant of TEF, such as proximal TEF or double TEF, might have been missed (22,45,52). In case of a recurrent TEF, the surgical approach does not seem to play a role, as no differences have been reported between open and thoracoscopic TEF repair (53). Other causes may include an anastomotic leak, tissue erosion by the sutures or clips applied, trauma secondary to esophageal dilations, and infection (45,49).

Diagnosis

Diagnostic investigations for recurrent TEF include esophagram, bronchoscopy, esophagoscopy, contrast swallowing, and CT scan. Combining multiple modalities, such as esophagogram, esophagoscopy and bronchoscopy, is recommended by most of the surgeons, as when only one is performed the recurrent TEF is often missed (45,54,55). The diagnosis and localization of recurrent TEF can be difficult due to postoperative of the inflammation. To maximize the chances of localizing the TEF, different maneuvers have been described. These include applying methylene blue into the esophagus under endoscopic guidance (54), introducing a catheter through the fistula from the trachea under bronchoscopy (22), backwards contrasting esophagram (51), CT scan with 3D reconstruction to have a full anatomic picture of the organo-vascular relationship (45).

Management

Correction of a recurrent TEF can be challenging and requires experienced skills for a surgeon. Endoscopic techniques have recently been advocated as the primary approach prior to surgical repair. These include stents placement, administration of tissue adhesive substances, combination of electrocautery with tissue glue (cyanoacrylate or fibrin glue) or laser (54,56). For the latter, the authors advocated the use of Bugbee electrocautery at low power on a narrow fistula followed by the application of Tisseel with additional aprotinin to gain better coagulation of the tract (54). However, in some cases, endoscopic attempts are not effective, and can be associated with complications, such as near fatal airway occlusion (45).

Surgery is the classical approach to treat recurrent TEF. According to Coran, it is critical to have the TEF identified and catheterized before opening the chest, the trachea should be completely separated from the esophagus before dividing the fistula, and viable tissue, such as a flap of pericardium or pleura or a lymph node with its blood supply, should be interposed between the suture lines (49). Some surgeons advocate for the rotation of the esophagus away from the trachea in order to separate the suture lines and prevent further recurrence (45).

Long-term morbidities

Dysphagia

This is one of the most common symptoms and complaints among children and adult who had EA/TEF repair, with rates as high as 75–100% (13,57). In a recent study, Coppens *et al*. showed that the prevalence of dysphagia decreases over time: 55% in patients <1 year of age, 51% in those 1–4 years old, 17% in those 5–11 years old, and 21% in those 12–18 years old (58). Manometry studies have demonstrated weak or absent esophageal peristalsis with impaired or absent contraction pattern (11). Experiments in rats have shown an abnormal intrinsic innervation of the distal esophagus that affects both excitatory and inhibitory intramural nerves (1).

Diagnosis and management

The video fluoroscopic swallow study is helpful to objectively assess the oral and pharyngeal phases of swallowing, which are affected 36% and 75% patients after EA/TEF repair, respectively (58). The severity of dysphagia post-TEF repair can be non-invasively evaluated using the Functional Oral Intake Scale, which includes seven levels ranging from nothing by mouth (level 1) to total oral diet with no restrictions (level 7) (59,60).

Children with TEF/EA and long-term dysphagia often also have structural airway abnormalities such as laryngomalacia, vocal cord paralysis, and tracheomalacia, oropharyngeal abnormalities, laryngeal clefts. Therefore, dysphagia and respiratory dysfunction may present with similar clinical signs, such as choking spells, aspiration, chest discomfort, or food impaction.

Dysphagia is also strongly associated with GER in children with repaired EA, regardless whether they had anti-reflux surgery in the form of fundoplication (58).

GER and esophagitis

Definition and etiology

GER is very common following repair of EA/TEF. It can be defined as reflux of gastric contents causing symptoms such as recurrent regurgitation with or without vomiting, poor weight gain, irritability, heartburn, or coughing (11). GER is caused by congenital dysmotility of the esophagus, incompetence of the lower esophageal sphincter mechanism, possibly due to shortened intra-abdominal esophagus, changes to the angle of His, injury to the vagus nerve during surgery, and widened esophageal hiatus (1,61-64). Anastomotic tension certainly plays a role in the development of GER, as evidenced by the high prevalence of GER in children with long-gap EA (65). A meta-analysis of complications and long-term outcome of children with long-gap EA managed with delayed primary anastomosis, a third of patients required a fundoplication within the first year after surgical repair due to either symptomatic GER (65). Nonetheless, GER can still occur following uncomplicated, tension-free anastomosis where the distal esophagus has remained untouched (1).

The sequelae of GER are substantial and range from recurrent anastomotic strictures to peptic esophagitis, eosinophilic esophagitis, and metaplasia (66). In particular, chronic GER may lead to Barrett's esophagus, a premalignant condition characterized by metaplastic changes of the epithelium of the lower portion of the esophagus. In Barrett's esophagus, the normal stratified squamous epithelium changes to simple columnar epithelium with interspersed goblet cells that are normally present only in the small intestine and large intestine. These histological changes can lead to esophagogastric junctional adenocarcinoma, a devastating malignant tumour with a mortality rate of over 85%. In the last decade, several studies showed that a substantial proportion of patients with repaired EA/TEF developed Barrett's esophagus, predominantly with gastric metaplasia (67,68).

Diagnosis

The ESPGHAN-NASPGHAN guidelines for the evaluation of GER in children with EA recommend the use of pH-impedance to assess and correlate non-acid GER with symptoms in selected patients, i.e., those symptomatic on antireflux medical therapy (proton pump inhibitors - PPI), those on continuous feeding, those with extra-digestive symptoms, and those with a normal pH-probe and endoscopy. In fact, non-acid reflux events are usually missed on pH monitoring without impedance tracing, a modality that has the additional benefit of correlating extra-esophageal symptoms with GER events. Interestingly, many children with repaired EA/TEF that are not on medications may have a normal reflux index yet experience a significant number of non-acid retrograde bolus movements (69).

Another modality that the ESPGHAN-NASPGHAN guidelines consider is the upper gastrointestinal endoscopy with biopsies, which allows to evaluate the status of the esophagus and to rule out the presence of eosinophilic esophagitis (EoE). EoE is a recognized chronic allergic/immune condition of the esophagus, characterized by an infiltration of eosinophils in the esophageal mucosa. EoE, whose pathogenesis is unclear, has been reported in several patients following EA/TEF repair (30,70,71), with a prevalence that seems >100-fold higher than in the general population (72). The ESPGHAN-NASPGHAN guidelines recommend ruling out EoE in patients with GER symptoms and refractory to PPI, before proceeding to anti-reflux (72). For all of those who instead are asymptomatic, the guidelines recommended routine endoscopy throughout childhood, i.e., after stopping PPI therapy, before the age of 10 years, at transition to adulthood (25).

Treatment

The management of GER after TEF/EA repair varies widely from conservative anti-reflux medications to several anti-reflux surgical approaches. For medical management, there is no consensus in the literature on the type and the duration of agent of anti-reflux therapy. Surveys of members of the Canadian Association of Pediatric Surgeons and the Midwest Pediatric Surgery Consortium showed that infants with repaired EA/TEF are prophylactically started on anti-reflux medications (PPI and/or H2-receptor antagonists), which are discontinued at different time points up to a year of age (2,73). This approach is reinforced in the ESPGHAN-NASPGHAN guidelines which recommend the prophylactic use of PPI up to the first year of life or longer, in case of persistence of GER symptoms (25). Nonetheless, studies have shown that histological complications, anastomotic stricture and leak rates, and pneumonia were found in patients with repaired EA/TEF regardless of the use of anti-reflux medications (2,66).

The proportion of patients requiring fundoplication widely ranges in various series from 15% to 100% (74-77). There are various techniques to perform anti-reflux surgery and they include the Nissen fundoplication, and the partial anterior (Thal, Ashcraft, Boix-Ochoa) and the posterior

(Toupet) hemi-fundoplication. According to a survey of EA/TEF on-line communities, which included EA/TEF Family Support Connection and Facebook TEF communities, 73% of patients underwent a Nissen fundoplication and 14% had a partial wrap (77).

Tracheomalacia

Definition and etiology

Tracheomalacia, which is defined as any degree of tracheal collapse during exhalation, is associated with congenital EA/TEF in almost 90% of children with EA/TEF (60,78). However, tracheomalacia is clinically significant only in 10% of patient with EA/TEF (1). When >70% of the tracheal wall collapses, usually patients present with symptoms (60,79). Interestingly, the incidence of tracheomalacia in the general population is only 0.05% (80). The high incidence in EA/TEF patients is partially explained by the fact that both the esophagus and trachea develop from the primitive foregut, and the same developmental stress factor affecting one organ likely affects also the other (81). Moreover, patients with repaired EA/TEF might have a weakness of the tracheal wall due to postoperative changes, prolonged intubation, and esophageal strictures (63).

Diagnosis

Flexible and rigid laryngotracheobronchoscopy is the method of choice, which is important to perform with spontaneous breathing as tracheomalacia is underestimated in a deeply anesthetized patient (60). To study the airways, a dynamic CT scan performed in two phases, end-inhalation and end-exhalation, can provide useful information about the location and extent of the wall collapse, as well as about the surrounding intra-thoracic structures and vascular anatomy (45,79).

Treatment

Usually, tracheomalacia improves with age. Nonetheless, symptomatic patients with blue spells, apnea, and recurrent chest infections may require treatment. Medical management consists in optimizing airway clearance using ipratropium bromide or saline nebulizing (79). It is important to remember that administration of beta-agonist bronchodilators to infants and children with tracheomalacia may worsen the airflow as it relaxes airway smooth muscle tone (79,82). The classical surgical treatment for tracheomalacia is aortopexy, that is the anchoring of the aorta to the posterior aspect of the sternum, thus drawing the anterior tracheal wall to the front and opening up the airway. More recently, the posterior tracheopexy technique has been described as an alternative, whereby horizontal mattress sutures are passed between the posterior tracheal and mainstem bronchial membrane to the anterior longitudinal spinal ligament under bronchoscopic guidance (83,84).

Vocal cord dysfunction (VCD)

Etiology

VCD in patients with repaired EA/TEF is characterized by hoarseness and dysphagia complicated with aspiration (85-88). The rate of postoperative VCD is highest (50%) in infants with TEF without EA (H-type) and lowest (4.6%) in those with EA and a distal TEF (3,88). VCD has also been reported to be more common often in patients with long gap EA (11%) than in patients with non-long gap EA (6%) (3). The etiology is typically attributed to an iatrogenic injury to the vagus nerve or to one or both recurrent laryngeal nerves as secondary to intraoperative dissection. This explains the high rates in patients with H-type TEF and long-gap EA that require more extensive dissection of the neck, thoracic inlet, and tracheoesophageal groove, and around the upper pouch (86,87). Similarly, thoracoscopic repair of EA appears to have higher rates of VCD probable due to high dissection of the esophagus into the thoracic inlet (89).

Prevention and treatment

Preoperative laryngo-trachea-bronchoscopy is highly recommended to investigate the airways and evaluate vocal cord motility at baseline (90). Postoperatively, vocal fold movement is best observed with the patient awake using flexible nasopharyngoscopy (85,90). Intraoperative nerve monitoring with electrodes on the vocal cords has been recommended in order to avoid recurrent laryngeal nerve injury during H-type TEF repair (91). Nonetheless, nerve monitoring does not seem to reduce nerve injury over visual identification of the nerve (92,93).

Most patients after TEF repair with VCD improve spontaneously, although the clinical improvement does not always correlate with vocal cord functional recovery (85). In fact, it has been reported that two-thirds of the children with VCD do not regain vocal cord function (85,88). Otolaryngology consultation and follow-up is recommended in symptomatic patients (88,90). Bilateral VCD may present as life-threatening airway obstruction, requiring tracheostomy or open airway surgery (86).

Patients with oropharyngeal aspiration may require feeding tube insertion and avoiding of oral feeds. VCD may result in delayed phonation and speech development (90).

Esophageal malignancy

Esophageal malignancies have been reported in patients with repaired EA/TEF repair, typically in the form of adenocarcinoma or squamous cell carcinoma, but also colorectal carcinoma in patients who underwent colonic interposition (48,94). However, two population-based studies showed that adults with repaired EA/TEF are at no higher risk for malignancy than the general population. One study presenting >50-year follow-up for 502 patients reported no cases of esophageal cancer (95), whereas the other described only three patients who had brain cancer, lymphoma and cervical cancer (96).

Conclusions

Overall, infants and children born with EA/TEF have high survival rates, but they are often affected by a number of postoperative complications. Some of these are intrinsic to the anatomical anomaly and some are secondary to the surgical repair. Many aspects of the management of these short- and long-term sequelae remain controversial and lack a standardized approach. Patients with recurrent strictures, severe dysphagia and GER are still challenging to treat. Long-term multidisciplinary clinic follow-up are common nowadays and help patients to be monitored longitudinally and surgeons to increase their understanding regarding the prevention and treatment of this complex congenital anomaly.

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