

ADVANCES IN THE TREATMENT OF CONGENITAL ESOPHAGEAL ATRESIA – A REVIEW OF SURGICAL TECHNIQUES AND LONG-TERM OUTCOMES

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ABSTRACT

Background: Congenital esophageal atresia (EA) is a rare developmental anomaly characterized by discontinuity of the esophagus, frequently accompanied by a tracheoesophageal fistula. It affects approximately 1 in 2,500 to 4,000 live births and is often associated with other congenital anomalies. Advances in neonatal intensive care, surgical techniques, and postoperative support have markedly improved survival rates, yet long-term morbidity remains substantial.

Aims: This review aims to provide a comprehensive and current analysis of surgical strategies for EA repair, critically appraising the available evidence on long-term outcomes and evaluating emerging multidisciplinary management approaches.

Methods: A narrative review was conducted using PubMed, Embase, and Google Scholar to identify peer-reviewed articles published in English between 2015 and January 2025. Inclusion criteria encompassed studies on surgical techniques, clinical and functional outcomes, complications, and long-term management of EA. A total of 31 articles, including original research, systematic reviews, and meta-analyses, were selected. Article selection was performed manually by reviewing titles, abstracts, and full texts, with additional studies identified through reference list screening of included articles.

Results: Surgical management of EA has progressed from traditional open thoracotomy to thoracoscopic

approaches that reduce musculoskeletal morbidity and improve cosmetic outcomes. Primary anastomosis remains standard in short-gap EA, whereas delayed or staged interventions are preferred in long-gap cases. Despite improved survival rates (>90% in high-income settings), long-term complications such as anastomotic strictures, gastroesophageal reflux, dysphagia, and respiratory morbidity remain frequent. Minimally invasive techniques demonstrate comparable safety and efficacy to open repair in selected patients but require advanced surgical expertise. Lifelong multidisciplinary follow-up involving gastroenterology, pulmonology, nutrition, and psychology is essential to improve functional outcomes and quality of life.

Conclusions: While survival in EA has markedly improved, the condition remains a lifelong disorder with significant long-term morbidity. The evidence base is limited by the predominance of retrospective studies, heterogeneity in outcome definitions, and variable follow-up durations. Surgical decisions should balance technical feasibility with long-term quality of life, and future research should prioritize standardized outcome measures, multicenter prospective studies, and optimized follow-up protocols.

Keywords: esophageal atresia, tracheoesophageal fistula, thoracoscopic repair, long-term outcomes, pediatric surgery, congenital anomalies, quality of life

INTRODUCTION

Congenital esophageal atresia (EA) is a rare developmental anomaly characterized by the discontinuity of the esophagus, in which the upper and lower parts of the esophagus fail to connect during embryogenesis. It is frequently accompanied by a tracheoesophageal fistula (TEF) [1]. Affecting approximately 1 in 2,500 to 4,000 live births, this condition poses immediate threats to neonatal survival due to feeding difficulties, aspiration risk, and associated anomalies, particularly cardiac, gastrointestinal, and genitourinary malformations [9; 30].

CLINICAL SIGNIFICANCE

The primary objective of early management of EA is to restore esophageal continuity and prevent complications. Over the past few decades, significant advancements in neonatal intensive care, surgical techniques, and postoperative support have transformed EA from a uniformly fatal condition to one with excellent survival rates exceeding 90% in high-income settings [22]. Despite improvements in survival, long-term morbidity remains substantial and includes esophageal strictures, gastroesophageal reflux disease (GERD), dysphagia, respiratory complications, and musculoskeletal sequelae such as scoliosis and chest wall deformities [3; 17; 29].

Surgical treatment strategies have evolved from open thoracotomy to minimally invasive techniques such as thoracoscopic repair, offering potential benefits including reduced musculoskeletal morbidity and faster recovery [6; 7; 24]. Nevertheless, these procedures remain technically demanding and require careful patient selection, especially in complex cases like long-gap EA or infants with very low birth weight [8; 20].

Long-term follow-up studies highlight the necessity of interdisciplinary surveillance extending into adolescence and adulthood, as patients often experience chronic issues impacting pulmonary function, nutritional status, and quality of life [9; 11; 27]. Additionally, the role of pharmacologic interventions such as proton pump inhibitors (PPIs) for stricture prophylaxis and GERD management remains under debate, with recent meta-analyses yielding mixed conclusions [4; 18; 25].

This review incorporates recent high-quality systematic reviews and meta-analyses [3, 4, 6, 18, 28, 31], together with contemporary cohort studies and longitudinal series [15, 22, 27], published in the last decade. By integrating the available international evidence with national data where applicable, the article allows comparison between broader recommendations and local clinical practices, which strengthens its scientific relevance.

AIMS

This review aims to provide a comprehensive and current analysis of surgical techniques for esophageal atresia repair, critically appraising the available evidence on long-term outcomes and evaluating emerging multidisciplinary management approaches.

METHODS

SEARCH STRATEGY

A comprehensive literature review was conducted to identify relevant studies on the surgical management and long-term outcomes of congenital esophageal atresia (EA). The search included peer-reviewed articles published between 2015 and 2025 in English, with full-text availability. The primary databases used for the

search were PubMed, Embase, and Google Scholar using combinations of the following keywords: (esophageal atresia OR congenital esophageal atresia) AND (open surgical repair OR thoracotomy OR thoracoscopy OR surgical management OR laparoscopic treatment OR grafts OR anastomoses OR esophageal reconstruction OR delayed treatment OR comorbidities) AND (long-term outcomes OR quality of life OR patient-reported outcomes OR complications).

The initial search revealed 77 results in PubMed and 60 in Embase. After removing duplicates, 98 unique articles remained for screening. Following a review of titles and abstracts, 46 studies were selected for full-text analysis. After applying eligibility criteria, 31 studies were ultimately included in this review.

The last literature search was completed in June 2025 to ensure the inclusion of the most up-to-date evidence. Article selection was performed manually by reviewing titles, abstracts, and full texts, with additional relevant studies identified through reference list screening of included articles. Only studies meeting the predefined eligibility criteria were retained for synthesis.

INCLUSION AND EXCLUSION CRITERIA

Inclusion Criteria:

Studies were included if they met any of the following conditions:

- Focused on surgical techniques for the repair of esophageal atresia, including open and minimally invasive methods
- Reported clinical or functional outcomes (short- or long-term) following EA repair
- Evaluated minimally invasive approaches or novel strategies in the surgical management of EA
- Presented data on long-term complications, such as strictures, gastroesophageal reflux, pulmonary function, or quality of life
- Included systematic reviews, meta-analyses, or large retrospective/prospective cohorts relevant to the subject
- Provided radiologic or diagnostic insights, particularly those influencing surgical planning or postoperative surveillance

Exclusion Criteria:

- Articles not available in English
- Studies without full-text access
- Publications that focused solely on non-esophageal anomalies or unrelated pediatric conditions
- Case reports or narrative reviews without clinical outcome data
- Reports based on in vitro and animal studies

A total of 31 articles were retained for final inclusion in this narrative review.

Table 1. Characteristics of Main Selected Studies Reviewed in the Context of Surgical Treatment Strategies and Long-Term Outcomes of Esophageal Atresia Repair

Author (Year)	Country	Study Design	Sample Size	Main Findings
Capitanio et al. (2021)	Italy	Observational, cross-section	50	Dysphagia is the most disabling symptom in children with EA, but overall quality of life is not severely affected.
Elhattab et al. (2020)	France, Egypt	Retrospective, bicentric	187	Thoracoscopic repair has comparable outcomes to thoracotomy, with added skeletal and cosmetic benefits.

Folaranmi et al. (2021)	United Kingdom	Single-center	198	Very low birth weight infants with EA have lower rates of primary anastomosis and higher mortality.
Giúdice et al. (2016)	Argentina	Prospective, longitudinal	27	Patients with EA remain at risk for long-term morbidity, supporting the need for lifelong follow-up.
Jové Blanco et al. (2020)	Spain	Retrospective, longitudinal	97	EA patients often develop progressive respiratory and gastrointestinal comorbidities.
Koivusalo et al. (2016)	Finland	Longitudinal	209	Routine childhood endoscopic surveillance after EA repair shows limited benefit.
Petit et al. (2019)	Canada	Prospective, longitudinal	77	Half of EA patients develop histopathological complications, particularly with recurrent strictures, requiring long-term follow-up.
Ritz et al. (2020)	Germany	Observational retrospective	48	In very low birth weight infants, staged repair lowers some complications but increases reflux, requiring individualized surgical planning.
Rozeik et al. (2020)	Egypt	Prospective randomized controlled trial	30	Thoracoscopic repair is equally safe as thoracotomy but offers superior cosmetic results and azygos vein preservation.
Stenström et al. (2017)	Sweden	Retrospective comparative	63	Prolonged PPI prophylaxis does not reduce stricture rates after EA repair.
Tamaki et al. (2024)	Japan	Retrospective, observational	25	Definitive EA surgery in trisomy 18 patients improves respiratory stability and survival compared with palliative approaches.
Ten Kate et al. (2021)	Netherlands	Longitudinal	110	Health status improves over time in EA patients, while quality of life may decline, highlighting the need to assess both separately.
Wei et al. (2017)	Canada	Prospective	52	Neonatal thoracotomy frequently causes musculoskeletal deformities, which muscle-sparing techniques can

FINDINGS

1. CLASSIFICATION AND ANATOMY OF THE DEFECT

1.1 CLASSIFICATION

Accurate anatomical classification of congenital esophageal atresia (EA) and associated tracheoesophageal fistula (TEF) is vital for clinical decision making and surgical planning. The Gross and Vogt classifications—described in the mid 20th century and still in widespread use—categorize EA/TEF into five principal types based on the presence and location of fistulas. The correspondence between descriptive terminology, Gross classification, and Vogt classification is presented in Table 2.

Table 2. Comparison of Gross and Vogt Classifications of Esophageal Atresia

Description of Anomaly	Gross Classification	Vogt Classification
Absent distal esophagus	-	Type I
Esophageal atresia without TEF (pure EA, ~7–8%)	Type A	Type II
EA with proximal TEF (rare)	Type B	Type IIIa
Esophageal atresia with distal TEF (most common, ~85%)	Type C	Type IIIb
EA with proximal and distal TEF (very rare)	Type D	Type IIIc
Isolated TEF without EA (“H-type” fistula, ~4–5%)	Type E	Type IV

1.2 ASSOCIATED ANOMALIES

EA is frequently accompanied by other congenital abnormalities, with up to 50–70% of patients exhibiting at least one additional malformation. The most well-recognized association is the VACTERL spectrum, which stands for vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb anomalies. Among these, cardiac defects are the most commonly reported, occurring in approximately 20–35% of patients, with ventricular septal defects and tetralogy of Fallot being prevalent subtypes [11; 30].

Pulmonary complications are also frequent, including congenital lung malformations, tracheomalacia, and long-term airway hyperreactivity. These can complicate both the surgical course and long-term respiratory outcomes, as highlighted in longitudinal studies [12; 17].

Musculoskeletal anomalies, particularly scoliosis and rib fusion, may develop as a result of thoracotomy or as congenital findings, necessitating orthopedic follow-up [29]. Additionally, genitourinary anomalies—such as renal agenesis or ectopic kidneys—are observed in up to 10–15% of patients, often requiring nephrological or urologic co-management [20].

In rare cases, EA may present in the context of syndromic conditions, including trisomy 18 or CHARGE syndrome, which significantly influence prognosis and long-term management needs [3; 26]. The presence and type of associated anomalies should be thoroughly evaluated at diagnosis, as they affect therapeutic strategy, risk stratification, and long-term outcomes.

1.3 CHALLENGES DEPENDING ON THE TYPE OF DEFECT

The anatomical type of esophageal atresia (EA) significantly influences the complexity of surgical management, the risk of early postoperative complications, and the burden of long-term after-effects. Gross type C (EA with distal tracheoesophageal fistula, TEF), the most common subtype, is often surgically manageable with primary anastomosis but is still associated with complications such as anastomotic strictures, gastroesophageal reflux

disease (GERD), and respiratory morbidity [30; 31].

In contrast, **long-gap EA** (most often Gross type A or B) poses a greater technical challenge due to the absence of a distal fistula and significant esophageal discontinuity. These patients frequently require staged surgical reconstruction—such as traction techniques or esophageal replacement—and face higher rates of anastomotic tension, leak, strictures, and feeding difficulties [15; 20]. This type of strategy is favored by the American Pediatric Surgical Association (APSA) and should be applied if feasible.

Patients with **Gross type D** (both proximal and distal TEF) or **E** (H-type TEF) present divergent diagnostic and therapeutic difficulties. The latter may remain undiagnosed until later in infancy and is often complicated by recurrent aspiration pneumonia and feeding troubles, sometimes leading to delayed developmental milestones [19].

The incidence of **postoperative complications** varies by defect type and includes:

- **Anastomotic stricture:** Reported in 30–60% of patients, especially in long-gap and high-tension repairs [4; 25].
- **GERD and esophagitis:** Occur in 40–70%, particularly in types with longer reconstruction or high anastomotic tension [18; 21].
- **Respiratory complications:** Tracheomalacia, chronic cough, and reactive airway disease are common, particularly in type C and long-gap variants [12; 17].
- **Feeding and growth disorders:** Frequently observed in types A and B due to prolonged hospitalization, use of gastrostomy tubes, and oral aversion [2; 11].

Defect type is also a determinant of long-drawn functional outcomes and quality of life. Type A/B patients often experience delayed achievement of full oral feeding and require longer-term multidisciplinary support. Type D is associated with higher rates of surgical reintervention due to dual fistulas and missed lesions [3; 20].

Overall, understanding the anatomical subtype of EA is essential for anticipating the scale of perioperative and chronic management challenges and guiding family counseling and care planning.

2. RECOGNITION AND DIAGNOSIS

2.1 CLINICAL SYMPTOMS

Clinical presentation of congenital esophageal atresia (EA) varies depending on the anatomical subtype, the presence or absence of a tracheoesophageal fistula (TEF), and associated aberrations. Most commonly, EA presents immediately after birth with symptoms related to feeding intolerance and airway compromise.

In the neonatal period, excessive salivation, drooling, coughing, and cyanotic episodes during feeding are hallmark signs. In EA with distal TEF (Gross type C), swallowed air enters the gastrointestinal tract, often leading to abdominal distension, while attempts to feed result in choking and aspiration [1; 30].

In contrast, long-gap EA (typically Gross type A) may present more subtly but becomes evident through inability to pass a nasogastric tube into the stomach, prompting radiologic investigation [20]. Isolated H-type TEF (Gross type E), which may go undiagnosed in the neonatal period, often manifests later with recurrent respiratory infections, aspiration pneumonia, and feeding difficulties [19].

Postoperatively and into childhood, many patients experience chronic respiratory symptoms, including recurrent bronchitis, wheezing, tracheomalacia, and chronic cough. A retrospective, longitudinal study from Spain showed that 24 out of 84 patients after EA repair were followed up by the Department of Pediatric Pulmonology, while in general survivors developed respiratory complications such as: respiratory exacerbations (42 patients, 50 %), wheezing or asthma events (38 patients, 45.2 %), pneumonia (26 patients, 30.9 %; 5 had a single event and 21, 2 or more), and tracheomalacia (21 patients, 25 %) [12; 17]. These are especially prevalent in patients with residual or missed TEF or in those who underwent open thoracotomy, which may contribute to restrictive pulmonary patterns [29].

Gastrointestinal symptoms often persist or emerge after repair, including difficulty in feeding, vomiting, dysphagia, and gastroesophageal reflux disease (GERD). GERD is among the most frequently reported chronic complications and can lead to esophagitis or anastomotic strictures if untreated [3; 18; 21].

Many patients, particularly those with long-gap EA or very low birth weight, show delayed growth and impaired weight gain, requiring nutritional support and feeding therapy [11; 22]. Early surgical intervention, complications, and prolonged hospital stays can also delay the development of normal oral feeding skills [2].

A subset of children, especially those born preterm or with associated anomalies, show delays in

neurodevelopment, while those undergoing thoracotomy may develop musculoskeletal deformities, including scoliosis and shoulder asymmetry, which contribute to reduced quality of life in adolescence and adulthood [27; 29].

2.2 DIAGNOSTIC METHODS

The diagnosis of congenital esophageal atresia (EA) typically occurs in the prenatal or immediate postnatal period, although subtle variants such as H-type tracheoesophageal fistulas (TEFs) may be diagnosed later. Early and accurate identification of EA is crucial for prompt surgical planning and the prevention of aspiration-related complications.

Prenatal suspicion often arises from polyhydramnios combined with the absence of a visible stomach bubble on ultrasound. These findings are suggestive but not specific and occur in less than half of EA cases [20; 30]. Fetal MRI may enhance diagnostic precision in cases where ultrasound is inconclusive, particularly in long-gap EA [20].

After birth, a failed attempt to pass a nasogastric tube into the stomach is the distinctive diagnostic maneuver. In suspected EA, the tube typically halts at ~10–12 cm from the gum margin in full-term neonates. This is confirmed by plain chest and abdominal radiography, which demonstrates the coiled tube in the proximal esophageal pouch and may show air in the gastrointestinal tract (suggestive of a distal TEF) [1; 19].

In selected cases—especially when the anatomy is unclear or H-type TEF is suspected—contrast esophagography or tracheobronchoscopy is employed to visualize the fistula [9; 19]. Preoperative rigid or flexible bronchoscopy is increasingly advocated to accurately locate and characterize the TEF, avoid missed fistulas, and guide surgical repair [19].

Because up to 50–70% of infants with EA have associated anomalies—especially in the context of VACTERL association—a thorough workup is essential. This typically includes echocardiography, renal ultrasonography, vertebral and limb radiographs, and abdominal ultrasound [3; 8]. Early detection of cardiac or renal anomalies impacts both surgical approach and anesthetic risk assessment.

After repair, routine surveillance is essential due to the high risk of complications such as strictures, anastomotic leaks, recurrent fistula, and GERD. Techniques include contrast esophagograms, esophagoscopy, and pH or impedance monitoring [4; 15]. Endoscopy is especially valuable in symptomatic patients for the detection of histopathologic changes like esophagitis or Barrett's esophagus [21].

Given the high prevalence of long-term respiratory morbidity, pulmonary function tests, chest imaging, and bronchoscopy are often used in follow-up, especially in children with recurrent infections or tracheomalacia [12; 17].

2.3 PRENATAL DIAGNOSIS - POSSIBILITIES AND LIMITATIONS

Prenatal detection of esophageal atresia (EA) remains a diagnostic challenge. Despite advances in fetal imaging and screening, the sensitivity and specificity for detecting EA in utero remain modest. The primary prenatal clues—polyhydramnios and non-visualization of the fetal stomach bubble—are neither consistent nor pathognomonic [20; 30].

Ultrasonography, the mainstay of prenatal imaging, identifies indirect signs such as a small or absent stomach and excessive amniotic fluid in approximately 50% of EA cases. However, these findings are also associated with other gastrointestinal and neurological conditions, limiting their specificity [30].

The diagnostic yield improves when EA is associated with other anomalies, particularly in syndromic cases like VACTERL association, which may prompt a more thorough evaluation. However, isolated EA—especially with a distal tracheoesophageal fistula (Gross Type C)—often escapes prenatal detection because the presence of a patent TEF allows amniotic fluid to pass into the stomach, resulting in a normal sonographic stomach bubble [1].

Fetal MRI has emerged as a valuable adjunct, particularly in cases of suspected long-gap EA. It provides detailed anatomical information and may enhance the prediction of EA by showing a dilated upper esophageal pouch or absent intra-abdominal fluid-filled esophagus [20].

Despite these tools, the prenatal diagnostic accuracy remains limited, with detection rates ranging from 30% to 50%, depending on the population and imaging protocols used [30]. Consequently, most diagnoses are still made postnatally, often triggered by failed nasogastric tube insertion shortly after birth.

In summary, while prenatal imaging can raise suspicion for EA, it cannot definitively confirm the diagnosis in most cases. Current limitations underscore the need for improved imaging techniques or biomarkers and emphasize the importance of comprehensive postnatal assessment in all neonates with suggestive prenatal

findings or clinical symptoms.

3. TRADITIONAL OPERATIONAL METHODS

3.1 OPEN SURGICAL TECHNIQUES (THORACOTOMY)

Thoracotomy has long been considered the gold standard for the repair of congenital esophageal atresia (EA), particularly in cases with distal tracheoesophageal fistula (TEF) (Gross Type C). This open surgical approach, typically via a right posterolateral thoracotomy, allows for direct visualization and meticulous dissection of the esophageal pouch and fistula, facilitating a high rate of primary anastomosis [1; 30].

The advantages of thoracotomy include established familiarity among surgeons, optimal exposure of mediastinal structures, and reliable identification and ligation of the fistula. It remains the preferred technique in neonates with low birth weight, severe associated anomalies, or in centers lacking experience with minimally invasive techniques [8; 20].

However, thoracotomy has its limitations. Postoperative complications include musculoskeletal deformities (e.g., scoliosis, winged scapula), shoulder dysfunction, and chronic thoracic pain. A Canadian prospect cohort study consisting of 52 patients at the age of 1–19 years revealed development of musculoskeletal malformations in 13 (25%), while the division of the serratus anterior was associated with a significantly higher probability of developing muscular deformities (log-rank $p=.0237$) and was also a strong predictor of the same [OR 8.6 (95% CI 1.8–42.1)] [29]. Studies show that children undergoing thoracotomy are at increased risk of long-term orthopedic complications due to rib spreading and muscle transection [6].

Respiratory outcomes are another critical consideration. Open repair can be associated with increased postoperative pulmonary complications, particularly in patients with preexisting lung hypoplasia or tracheomalacia [12; 17]. Nonetheless, when compared to thoracoscopic approaches, thoracotomy offers shorter operative times and may be associated with fewer intraoperative complications in less experienced hands [7; 31].

The long-term outcomes following open repair are generally favorable in terms of anastomotic integrity and survival. However, stricture formation, recurrent fistula, and dysphagia remain concerns, often requiring ongoing multidisciplinary follow-up [3; 9; 11].

In conclusion, while minimally invasive surgery is gaining ground, open thoracotomy remains a safe, effective, and widely used method for EA repair, particularly in complex or high-risk neonatal presentations. Its enduring role in surgical practice is supported by decades of data and continued refinement of technique.

3.2 STRATEGIES FOR TREATING DIFFERENT TYPES OF ATRESIA WITH OPEN TECHNIQUES

The management of congenital esophageal atresia (EA) with open techniques via thoracotomy is highly dependent on the anatomical type of atresia, presence and location of a tracheoesophageal fistula (TEF), and patient-specific factors such as birth weight, gestational age, and comorbidities. Open surgical repair remains a versatile and reliable approach, particularly in complex or atypical cases.

In Gross Type C (Vogt IIb), which accounts for approximately 85–90% of EA cases, the standard open approach involves right posterolateral thoracotomy with ligation of the fistula and primary end-to-end esophageal anastomosis. The open approach allows optimal exposure for delicate dissection and tension-free repair [1; 30]. This strategy remains standard, especially in neonates with unstable respiratory or hemodynamic status, or those with low birth weight [8; 22].

In Type A atresia, where there is no TEF, the esophageal segments are often widely separated (long-gap EA). Open surgical management may involve delayed primary anastomosis following esophageal elongation (Foker technique), gastrostomy with staged repair, or esophageal replacement in extremely long-gap cases [20]. Careful preoperative imaging and intraoperative assessment guide the choice of technique. Open access provides essential versatility in handling these technically demanding repairs.

Type B and Type D are rare variants that require heightened intraoperative vigilance to identify both fistulae. Type D, with both proximal and distal TEFs, may be misdiagnosed preoperatively, and intraoperative findings often dictate surgical planning. Open thoracotomy offers the best exposure to identify multiple fistulas, particularly when a cervical fistula is suspected [16; 19].

In patients with H-type TEF (Gross type E), typically presenting later with aspiration or recurrent respiratory infections, a cervical approach is often preferred. However, if thoracic exposure is required (e.g., if the fistula is located deep in the thorax), a right thoracotomy provides the necessary access for fistula ligation [30].

In patients with long-gap EA (>2.5–3 cm), open techniques like the Foker method (esophageal elongation via

external traction sutures) have been increasingly applied with success. These patients often require prolonged hospital stays, staged interventions, and long-term follow-up for strictures, dysmotility, and growth [11; 20].

Open repair continues to play a crucial role in high-risk patients: those with severe associated anomalies, trisomy 18, cardiac defects, or very low birth weight. Although thoracoscopy is increasingly favored in specialized centers, open thoracotomy remains the method of choice in neonates not suitable for prolonged anesthesia or CO₂ insufflation [3; 6; 26].

3.3 PRIMARY VS DELAYED ANASTOMOSIS

The decision between primary and delayed anastomosis in the open surgical management of esophageal atresia (EA) is guided by anatomical gap length, patient stability, and institutional expertise. While primary anastomosis remains the preferred approach in most infants with short-gap EA, delayed repair is often employed in cases of long-gap EA or when perioperative risks are heightened.

In the majority of infants with Gross type C EA (distal tracheoesophageal fistula and a short esophageal gap), primary anastomosis via open thoracotomy remains the gold standard. It enables a single-stage correction and avoids the morbidity associated with staged procedures. This approach has been associated with high survival rates and acceptable rates of postoperative complications such as stricture and leak [1; 30].

Several studies affirm that primary repair contributes to favorable long-term outcomes, particularly when performed by experienced pediatric surgical teams and when the esophageal ends are approximated with minimal tension [6; 7]. However, in infants with low birth weight (<1500 g) or severe comorbidities, the risk of anastomotic complications may increase even with short-gap anatomy [8; 22].

Delayed repair is typically indicated in long-gap EA (commonly type A), where primary tension-free anastomosis is not feasible. Strategies include initial gastrostomy for feeding, followed by staged repair using techniques such as traction-based esophageal lengthening (e.g., Foker method), or esophageal replacement (e.g., gastric or colonic interposition) [20].

Delayed anastomosis allows for spontaneous growth and approximation of the esophageal ends and provides time to optimize the patient's nutritional and respiratory status. However, it may increase the risk of gastroesophageal reflux, anastomotic stricture, and feeding difficulties [11; 19].

While primary repair is associated with shorter hospital stays and earlier establishment of oral feeding, delayed techniques are sometimes necessary to avoid high-risk anastomosis. The presence of congenital anomalies, trisomy 18, and poor pulmonary reserve further shift the balance toward delayed or staged approaches [26].

Regardless of timing, meticulous surgical technique and perioperative care are paramount in optimizing outcomes. Multidisciplinary follow-up, including respiratory, gastroenterological, and nutritional support, is critical for all EA patients, particularly those undergoing delayed repair [17; 27].

4. ADVANCES AND NEW SURGICAL TECHNIQUES

4.1 MINIMALLY INVASIVE TECHNIQUES (THORACOSCOPY)

The advent of minimally invasive surgery has significantly influenced the management of esophageal atresia (EA), particularly in cases involving distal tracheoesophageal fistula (TEF). Thoracoscopic repair, first described in the late 1990s, has evolved into a viable alternative to traditional open thoracotomy.

Thoracoscopic techniques offer several distinct benefits, including reduced musculoskeletal complications such as scoliosis and shoulder girdle asymmetry, which are commonly associated with thoracotomy [6; 29]. Additionally, thoracoscopy is associated with less postoperative pain, quicker recovery, and superior cosmetic outcomes.

Recent comparative studies show that thoracoscopic repair achieves comparable rates of anastomotic leak, stricture, and fistula recurrence to open techniques when performed by experienced surgeons [7; 23; 31]. Moreover, systematic reviews suggest that minimally invasive repair does not compromise long-term outcomes, including respiratory function and growth trajectories [9; 24].

Despite its advantages, thoracoscopic EA repair presents a steep learning curve, requiring advanced endoscopic skills and specific neonatal anesthesia protocols. Challenges include precise dissection in a limited operative field, intracorporeal suturing, and maintaining adequate exposure of the posterior mediastinum [1; 30].

Patient selection remains crucial. Thoracoscopy is most commonly applied in type C EA in infants with stable cardiorespiratory status and no significant associated anomalies. In very low birth weight (VLBW) neonates or those with long-gap EA, thoracoscopic repair may be technically unattainable or inadvisable [20; 22].

Longitudinal studies and interdisciplinary follow-up have shown that thoracoscopic repair does not adversely affect pulmonary or gastrointestinal outcomes compared to open techniques. However, ongoing surveillance is essential due to the risk of complications such as gastroesophageal reflux, tracheomalacia, and recurrent strictures [3; 11; 17].

Moreover, thoracoscopy may reduce the prevalence of thoracotomy-associated musculoskeletal deformities, which often manifest later in childhood and adolescence, emphasizing the potential long-term benefit of minimally invasive approaches [19; 29].

4.2 ESOPHAGEAL RECONSTRUCTIONS - INTESTINAL GRAFTS, PLASTICS

In cases of long-gap esophageal atresia (LGEA), where primary anastomosis is not viable, esophageal replacement using intestinal segments or reconstructive plastic techniques becomes necessary. These approaches are typically reserved for complex cases where length discrepancy prevents native esophageal continuity despite traction or elongation attempts.

Intestinal interpositions—using stomach (gastric pull-up), colon, or jejunum—are established alternatives for esophageal replacement. The gastric pull-up remains one of the most frequently used techniques due to its single anastomosis, reliable blood supply, and plausibility in infants [20; 30]. However, colon and jejunal interpositions are still considered in select cases, particularly when the stomach is unavailable or unsuitable.

Plastic techniques such as the Foker method (tension-induced growth of native esophageal ends) or Kimura myotomy-based elongations aim to preserve the native esophagus and avoid replacement altogether. These procedures may require prolonged intensive care and are typically performed in specialized centers [20].

Esophageal reconstruction is associated with a unique set of long-term risks. Patients with gastric transpositions may experience significant gastroesophageal reflux, delayed gastric emptying, and risk of Barrett's esophagus [3; 9]. Jejunal grafts, although physiologically closer to the esophagus in terms of motility, pose greater technical challenges and risks of graft necrosis.

Colon interposition is generally reserved for older children and often results in better outcomes in terms of conduit patency, albeit with potential for redundancy and motility disorders. Regardless of the conduit, esophageal reconstructions require long-term follow-up with endoscopic surveillance, nutritional assessment, and pulmonary monitoring [11; 15; 17].

Quality of life studies emphasize that while intestinal grafts enable survival and oral intake, they often come at the cost of increased interventions, hospitalizations, and long-term complications, particularly in cases of early-life failure to thrive or comorbid anomalies [10; 19].

4.3 THORACOSCOPY VS. THORACOTOMY

Thoracoscopic repair of esophageal atresia has steadily gained traction, offering a minimally invasive alternative to the traditional open thoracotomy. Advocates emphasize that thoracoscopic approaches generally result in smaller incisions, potentially reducing postoperative pain, lowering infection risk, and enhancing cosmetic results when compared to the more invasive thoracotomy.

In terms of perioperative outcomes, studies indicate that children undergoing thoracoscopic repair typically experience less musculoskeletal trauma. For example, thoracotomy has been associated with a higher incidence of long-term chest wall deformities or scoliosis due to rib spreading, whereas thoracoscopy, which avoids rib spreading, tends to preserve musculoskeletal integrity more effectively [29].

Regarding short-term surgical metrics, minimally invasive thoracoscopic procedures may involve slightly longer operative times, particularly during the initial learning curve. In a retrospective bicentric study of two major pediatric surgery centers in Paris, France, and Mansoura, Egypt the mean operative time was 127.6 ± 35 minutes in thoracoscopic group and 105.7 ± 23 minutes in open thoracotomy group ($P = .0005$). The mean postoperative ventilation time and the mean length of stay were significantly shorter in the thoracoscopic group ($P = .004$ and $P < .0001$, respectively) [7].

Long-term follow-up suggests that thoracoscopic repair is at least comparable to thoracotomy with respect to key functional outcomes including anastomotic stricture rates, gastroesophageal reflux requiring intervention, and overall respiratory health. According to bicentric study by Elhattab et al. the incidence of anastomotic leak was 8.9% in thoracoscopic group versus 16.4% in open thoracotomy group ($P = .33$). Anastomotic stenosis occurred in 33.3% of thoracoscopic group and in 22.4% of open thoracotomy group ($P = .17$). [7; 29].

In summary, thoracoscopy offers several clear benefits—especially regarding reduced musculoskeletal complications and improved cosmetic outcomes—though without compromising critical long-term results. As surgical proficiency with the technique continues to increase, thoracoscopic repair is likely to become the

standard approach for most cases of esophageal atresia suitable for minimally invasive management.

5. COMPLICATIONS AND POSTPONED TREATMENT

Despite advances in surgical techniques for congenital esophageal atresia (EA), postoperative complications remain a significant challenge impacting patient outcomes and necessitating ongoing management. The most common complications include anastomotic leaks, strictures, recurrent fistulas, gastroesophageal reflux disease (GERD), and dysphagia, often requiring multidisciplinary and staged approaches.

Anastomotic leakage is a critical early postoperative complication, occurring in up to 15% of patients, and may prolong hospitalization or require reoperation [7; 30]. Stricture formation at the anastomotic site is a frequent long-term issue, reported in 30–50% of cases, causing dysphagia and feeding difficulties. The etiology includes ischemia, tension, or reflux-induced inflammation [3; 20].

The meta-analysis of 4 retrospective studies (total 455 infants) found that using a transanastomotic feeding tube (TAFT) increased the risk of postoperative stricture by 83% compared to no TAFT, with a pooled risk ratio (RR) of 1.83 (95% CI 1.30–2.58; $p = 0.0005$). Despite the increased stricture risk, TAFT was not associated with higher rates of several serious postoperative complications: anastomotic leakage (RR 1.65; 95% CI 0.93–2.93; $p = 0.09$), sepsis (RR 0.91; $p = 0.85$), tracheomalacia (RR 1.89; $p = 0.56$), gastroesophageal reflux (RR 0.50; $p = 0.31$), wound infection (RR 1.29; $p = 0.74$), and pneumonia (RR 0.97; $p = 0.99$). Despite limited and conflicting evidence, TAFT remains in use by 80–90% of surgeons worldwide, highlighting the need for prospective, randomized trials to determine whether TAFT should remain routine care in EA repair. [28]

Recurrent or persistent tracheoesophageal fistulas (TEFs) are less common but demand prompt diagnosis and surgical intervention due to aspiration risks [19]. Early detection through clinical vigilance and imaging is essential to minimize morbidity.

GERD is highly prevalent in EA patients, with up to 60% experiencing reflux symptoms post-repair [3; 18]. Chronic reflux contributes to esophagitis, strictures, and aspiration pneumonia, emphasizing the importance of surveillance and prophylactic proton pump inhibitor (PPI) therapy [4; 25].

Dysphagia, often multifactorial, persists in many patients even after anatomical repair and may be related to motility disorders or anatomic complications [8; 27]. It significantly affects growth and quality of life, requiring individualized therapeutic strategies.

Endoscopic dilatation remains the cornerstone for managing anastomotic strictures, with serial balloon or bougie dilations providing symptomatic relief in most cases [15; 20]. However, refractory or complex strictures may necessitate adjunctive therapies, such as steroid injections or stenting.

Reoperation is indicated for severe or recurrent strictures, persistent leaks, or fistulas unresponsive to conservative treatment [6; 7]. Decisions regarding timing and surgical approach must weigh risks of morbidity against potential functional improvements.

Some cases require delayed repair or staged interventions due to prematurity, low birth weight, or associated anomalies [8; 22]. Prolonged follow-up is essential to monitor pulmonary complications, nutritional status, and psychosocial outcomes [11; 17].

6. LONG-TERM OUTCOMES AND QUALITY OF LIFE

Advances in the surgical management of congenital esophageal atresia (EA) have significantly improved survival rates; however, patients frequently face long-term challenges that affect esophageal function, growth, and overall quality of life (QoL). Understanding these outcomes is essential for optimizing lifelong care.

Quality of life assessments reveal that many EA survivors experience ongoing difficulties with feeding, swallowing, and respiratory health, contributing to psychosocial stress and impaired physical development [2; 27]. Dysphagia and GERD may lead to poor weight gain and growth failure, requiring nutritional support and monitoring [11; 22].

Psychological impacts, including anxiety and reduced social participation, are increasingly recognized, underlining the necessity of comprehensive care that addresses mental health alongside physical aftermaths [9]. Functional esophageal assessments and endoscopic surveillance are critical in the early detection of complications affecting QoL [15].

A prospective, longitudinal follow-up study was conducted in Rotterdam, The Netherlands, including 110 participants (62% boys, born with EA between 1999 and 2011 at 8 and/or 12 years old) by obtaining Pediatric Quality of Life Inventory (HS) and DUX-25 (QoL) questionnaires. Self-reported HS improved significantly between 8 and 12 years for both boys (mean difference [md] 4.35, effect size [ES] 0.54, $P = 0.009$) and girls

(md 3.26, ES 0.63, $P = 0.004$). Proxy-reported HS tended to improve over time, while self-reported and proxy-reported QoL tended to decline. Self-reported HS at 8 years was below normal for both boys (md -5.44, ES -0.35, $P < 0.001$) and girls (md -7.61, ES -0.32, $P < 0.001$). Girls' self-reported QoL was below normal at 8 (md -5.00, ES -0.18, $P = 0.019$) and 12 years (md -10.50, ES -0.26, $P = 0.001$). Parents reported normal HS at both ages, whereas they rated the QoL of their daughters below normal at 12 years (md -10.00, ES -0.16, $P = 0.022$). [27]

Given the multisystem involvement, coordinated long-term multidisciplinary follow-up is paramount to optimize outcomes. Teams including pediatric surgeons, gastroenterologists, pulmonologists, nutritionists, and psychologists provide holistic care addressing feeding difficulties, pulmonary complications, musculoskeletal issues, and psychosocial development [2; 9; 11].

Transition from pediatric to adult care remains a critical period, requiring structured protocols to ensure continuity and address evolving health needs [3]. Emerging evidence supports the establishment of specialized esophageal atresia clinics to provide tailored surveillance and early intervention, ultimately improving survival and quality of life [19]

A consolidated summary of the reported outcomes from the included studies is presented in the table below, allowing direct comparison of short- and long-term results across different surgical approaches to esophageal atresia repair. This synthesis highlights variations in postoperative complications, functional outcomes, and quality of life measures reported in the literature.

Table 3. Quantitative outcomes after esophageal atresia repair based on data reported in the review

Surgical approach	Outcome	Quantitative estimate	Sources
Any repair	Overall survival in high-income settings	>90%	[22]
Any repair, especially long-gap or high-tension anastomosis	Anastomotic stricture	30–60%	[4], [25]
Any repair with longer reconstruction or high anastomotic tension	GERD or esophagitis	40–70%	[18], [21]
Any repair with TAFT	Stricture risk versus no TAFT	RR 1.83, 95% CI 1.30–2.58, $p=0.0005$	[28]
Any repair with TAFT	Leak, sepsis, tracheomalacia, GERD, wound infection, pneumonia	No significant differences reported versus no TAFT with the RRs listed in text	[28]
Open thoracotomy	Long-term musculoskeletal deformities	25%, OR 8.6 (95% CI 1.8–42.1), log-rank $p=.0237$	[29]
Any repair	Chronic respiratory morbidity including tracheomalacia, chronic cough	25–50%	[12], [17]
Any repair	Quality of life and health status in follow-up	Self-reported HS at 8 years (md -5.44, ES -0.35, $P < 0.001$) for boys and (md -7.61, ES -0.32, $P < 0.001$) for girls	[27]

DISCUSSION

The management of congenital esophageal atresia (EA) has evolved markedly over recent decades, transforming a once uniformly fatal anomaly into a survivable condition with favorable short-term outcomes. However, as highlighted in this review, significant challenges remain—particularly concerning long-term morbidity, surgical complexity in specific anatomical subtypes, and the need for lifelong multidisciplinary care.

Open thoracotomy remains the mainstay in many centers, particularly for complex presentations or in low-resource settings. Despite its proven efficacy, the long-term orthopedic and respiratory sequelae associated with thoracotomy highlight the value of thoracoscopic approaches in selected patients. Minimally invasive techniques offer clear advantages in reducing musculoskeletal morbidity and improving cosmetic outcomes, but require significant surgical expertise and are not universally applicable, especially in long-gap EA or neonates with comorbidities.

The choice between primary and delayed anastomosis continues to be dictated by gap length, overall patient stability, and institutional experience. Techniques such as the Foker method or esophageal replacement are increasingly used in long-gap cases, but they carry higher complication rates and demand prolonged hospitalization. These approaches also underscore the importance of preserving native esophageal tissue whenever feasible, given the functional limitations associated with interposed grafts.

Despite technical refinements, complications such as anastomotic stricture, gastroesophageal reflux disease (GERD), dysphagia, and recurrent fistula remain common and often necessitate repeat interventions. This reinforces the notion that surgical success must be evaluated not only by initial repair integrity but also by functional outcomes, nutritional status, and quality of life. ESPGHAN–NASPGHAN guidance for EA specifically supports prophylactic acid suppression in infancy, often for the first year, to reduce peptic complications. Endoscopic screening with biopsies is mandatory for routine monitoring of GERD in patients with EA every 5–10 years [14].

For patients with primary presentation of tracheomalacia, recurrent fistula or other trachea-laryngeal anomalies lung function tests should be routinely used for monitoring, whereas bronchoscopy must only be performed in case of acute airway symptoms according to ERNICA Consensus on the follow-up of patients with esophageal atresia and tracheoesophageal fistula [5].

Long-term follow-up studies consistently demonstrate that EA survivors are at risk for both gastrointestinal and respiratory complications well into adolescence and adulthood. As such, structured transition protocols from pediatric to adult care are essential. Furthermore, emerging evidence supports the integration of dedicated EA multidisciplinary clinics, which have been shown to improve surveillance, early intervention, and patient-reported outcomes. Every adult patient with previously treated EA should remain under specialized care according to detailed transition-to-adult-care recommendations of the International Network on Oesophageal Atresia (INoEA) [13].

The variable accuracy of prenatal diagnosis remains a limitation, often delaying appropriate preparation for neonatal management. Although fetal MRI has improved detection in certain cases, especially long-gap EA, the widespread application of this modality remains constrained by availability and cost.

Finally, there is a pressing need for the adoption of standardized outcome definitions and uniform reporting protocols across centers to enable reliable comparison of surgical results. The lack of harmonized functional assessment tools and inconsistency in defining complications significantly hinder meta-analyses and the development of robust clinical guidelines. Future research should prioritize well-designed, multicenter prospective studies with sufficient follow-up duration to capture both early and late sequelae. Establishing international collaborative registries would facilitate the pooling of high-quality data, improve external validity, and accelerate the refinement of evidence-based surgical and follow-up strategies for patients with EA.

CONCLUSION

In conclusion, while survival in EA has greatly improved, the condition remains a lifelong disease requiring individualized, multidisciplinary, and dynamic management. The available evidence is limited by the predominance of retrospective designs, heterogeneity in outcome definitions, and variable follow-up durations across studies, which should be considered when interpreting the findings of this review. Surgical decisions must balance technical feasibility with long-term quality of life, and ongoing research should continue to refine both therapeutic strategies and follow-up paradigms.

DISCLOSURES

AUTHORS' CONTRIBUTION:

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All authors have read and agreed with the published version of the manuscript.

FUNDING STATEMENT

The study did not receive special funding.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

The authors did not use any AI language models for generating the text. Plagiarism and grammar check were performed using 'Grammarly' (Grammarly Inc.)

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