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Original Research Article

Esophageal Atresia Repair: Outcomes and Long-Term Follow-Up in Neonates

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Abstract:

Background: Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) is one of the most surgically challenging congenital anomalies encountered in the neonatal period. While operative survival has markedly improved over the decades, affected children continue to carry a substantial burden of long-term gastrointestinal and respiratory morbidities. This study aimed to document short-term surgical outcomes and long-term follow-up data in neonates undergoing EA repair at a tertiary pediatric surgical centre in India.

Methods: A retrospective cohort study was conducted over an eight-year period (January 2015 to December 2022). All neonates diagnosed with EA with or without TEF who underwent primary surgical repair were included. Data on demographics, type of EA, surgical approach, intraoperative findings, postoperative complications, and long-term outcomes were collected and analyzed. Follow-up assessments were performed at 1, 3, and 5 years.

Results: A total of 70 neonates were included; 52 (74.3%) had Type C EA with distal TEF. Associated anomalies were present in 44 (62.9%), most commonly cardiac defects. Open thoracotomy was performed in 44 (62.9%) and thoracoscopic repair in 26 (37.1%) patients. Anastomotic leak occurred in 12 (17.1%) and anastomotic stricture in 28 (40.0%) cases. Gastroesophageal reflux disease (GERD) was the most prevalent long-term complication, occurring in 67.1% of the cohort. Overall mortality was 5.7%. At five-year follow-up, 95.5% of surviving patients achieved full oral feeding.

Conclusion: Operative outcomes of EA repair have improved considerably, yet long-term morbidities—particularly GERD, dysphagia, and esophageal stricture—remain highly prevalent. Structured long-term surveillance protocols, including regular endoscopy and pulmonary function assessment, are essential for optimizing quality of life in these children.

Keywords: Esophageal atresia; Tracheoesophageal fistula; Neonatal surgery; Anastomotic stricture; Gastroesophageal reflux disease; Long-term follow-up.

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Introduction

Esophageal atresia (EA) is a congenital malformation arising from incomplete separation of the primitive foregut during early embryogenesis, resulting in a blind-ending esophageal pouch, with or without an abnormal communication—tracheoesophageal fistula (TEF)—between the trachea and the distal esophageal segment.[1,2] The condition has an estimated birth prevalence of approximately 1 in 3,500 live births, placing it among the most common significant congenital anomalies of the gastrointestinal tract.[3] The Gross classification, introduced in 1953, remains widely used in clinical practice, with Type C (proximal atresia with distal TEF) accounting for approximately 85% of all cases.[4]

The trachea and esophagus arise from a common foregut tube, and their failure to separate correctly during the fourth to sixth week of gestation is implicated in the pathogenesis of EA and TEF.[5] A range of molecular factors and signaling pathways—including the SOX2, FOXA2, and Hedgehog pathways—have been identified as contributors to this developmental failure, though no single genetic etiology has been universally established.[6] Approximately 50% of affected neonates have at least one associated structural

anomaly, with the VACTERL association (vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb defects) being the most frequently recognized co-occurrence.[7,8] Cardiac defects are the most common individual anomaly, present in up to 59% of cases, and carry the greatest influence on operative risk and mortality.[9]

Prenatal diagnosis may be suspected in the third trimester when maternal polyhydramnios is noted on ultrasonography, a finding that reflects the neonate's inability to swallow amniotic fluid effectively.[10] Postnatally, the classic clinical trial of excessive drooling, choking on feeding attempts, and failure to pass a nasogastric tube should prompt immediate radiological confirmation. A coiled nasogastric tube on plain chest radiograph, with or without air in the stomach, is typically diagnostic.[1]

Surgical correction remains the only definitive treatment. The primary goals of operative repair are to divide and ligate the fistula (when present) and to restore esophageal continuity by means of an end-to-end anastomosis.[11] Traditionally, this was achieved through open right posterolateral thoracotomy using an extrapleural approach. Over the past two decades, however, thoracoscopic repair has gained considerable traction as a minimally invasive alternative, offering potential advantages including reduced chest wall morbidity, better cosmesis, and shorter hospital stay, albeit at the cost of longer operative times and a steeper learning curve. [12,13]

Despite remarkable improvements in survival—now exceeding 90% in high-income settings, even among those with significant comorbidities [14]—the long-term burden of disease associated with EA repair remains considerable. Anastomotic stricture, gastroesophageal reflux disease (GERD), tracheomalacia, esophageal dysmotility, and recurrent TEF collectively constitute a spectrum of complications that require active and sustained surveillance throughout childhood and into adulthood.[15,16] Long-term studies have demonstrated that GERD, dysphagia, and respiratory symptoms persist in a significant proportion of adult survivors, with Barrett's esophagus and even esophageal carcinoma reported as rare but serious late sequelae.[17]

In resource-limited settings, where multidisciplinary neonatal intensive care facilities may be suboptimal and the volume of complex neonatal surgery concentrated in fewer centres, outcome data take on added importance. There is a recognized need for region-specific audit of outcomes that can inform local clinical practice and identify areas for quality improvement. Most published long-term follow-up data originate from North American or European tertiary centres, and comparatively few prospective or retrospective series document outcomes over a five-year horizon in South Asian populations.

This study was therefore undertaken to provide a comprehensive account of the short-term surgical outcomes, postoperative complications, and structured long-term follow-up data in a cohort of neonates managed at a single tertiary pediatric surgical centre in South India. We additionally sought to compare outcomes between open thoracotomy and thoracoscopic repair, and to identify clinical predictors of anastomotic complications and GERD requiring surgical intervention.

Materials and Methods

Study Design and Setting: This was a retrospective cohort study conducted in the Department of Pediatric Surgery at our tertiary care Medical College and Hospital. The study included all consecutive neonates diagnosed with EA with or without TEF who underwent primary surgical repair between January 2016 and December 2022. Informed written parental consent was obtained prior to all operative procedures.

Inclusion and Exclusion Criteria: All neonates (age ≤28 days) with a confirmed diagnosis of EA with or without TEF who underwent primary repair during the study period were eligible for inclusion. Neonates with isolated H-type TEF (Gross Type E) who did not require esophageal anastomosis, those transferred out to another facility prior to definitive repair, and those whose medical records were incomplete for more than 20% of the primary outcome variables were excluded.

Surgical Technique: The choice of operative approach—open right posterolateral thoracotomy via an extrapleural route, or thoracoscopic repair—was made by the operating surgeon based on the patient's clinical stability, birth weight, gestational age, and anatomical characteristics of the lesion. In all cases, the fistula was divided and ligated, and the esophageal pouches were mobilized to achieve a tension-free end-to-end anastomosis using interrupted absorbable sutures (polyglycolic acid 5-0) over a transanastomotic nasogastric feeding tube. For long-gap EA cases (gap ≥3 cm), a staged approach with initial gastrostomy was employed. Postoperatively, all patients were managed in the Neonatal Intensive Care Unit (NICU) with ventilatory support as required.

Data Collection and Outcome Variables: Data were extracted from the hospital's electronic medical records and supplemented by review of operation notes and outpatient case files. Variables collected included gestational age, birth weight, sex, type of EA (Gross classification), associated congenital anomalies, operative approach and

duration, intraoperative findings (gap length, azygos vein status), and early postoperative complications including anastomotic leak (confirmed by contrast esophagogram), anastomotic stricture (dysphagia requiring endoscopic dilation), recurrent TEF, tracheomalacia, and mortality.

Long-term follow-up was conducted at 1, 3, and 5 years. Outcomes assessed at follow-up included GERD (defined as symptoms requiring ongoing proton pump inhibitor therapy or anti-reflux surgery), dysphagia, recurrent anastomotic stricture requiring endoscopic balloon dilation, respiratory symptoms, growth parameters (weight-for-age Z-scores), feeding status (oral vs. tube-assisted), and endoscopic findings including Barrett's esophagus.

Statistical Analysis: Statistical analysis was performed using IBM SPSS Statistics version 25.0 (IBM Corp., Armonk, NY, USA). Continuous variables are expressed as mean ± standard deviation (SD) or median with interquartile range (IQR), as appropriate. Categorical variables are presented as frequencies and percentages. Comparisons between groups were made using the independent samples t-test or Mann-Whitney U test for continuous variables and the Chi-square or Fisher's exact test for categorical variables. A P-value of <0.05 was considered statistically significant.

Results

Patient Demographics and Preoperative Characteristics: A total of 70 neonates underwent EA repair during the eight-year study period. Fifty-two patients (74.3%) had Gross Type C EA with a distal TEF, while 18 (25.7%) had isolated EA (Types A and B combined). The mean gestational age was 37.6 ± 2.0 weeks across the full cohort, and the mean birth weight was 2,840 ± 465 grams. Eighteen neonates (25.7%) were born prematurely, and ten (14.3%) met criteria for very low birth weight (<1,500 g). Forty-four neonates (62.9%) had at least one associated congenital anomaly, and this proportion was significantly higher in the Type C EA group (69.2%) compared to isolated EA cases (44.4%; P=0.047). Cardiac defects were the most prevalent associated anomaly, identified in 26 (37.1%) of the cohort. VACTERL association was diagnosed in 20 neonates (28.6%) and was significantly more common among Type C EA patients (P=0.049). Antenatal diagnosis was achieved in 44 cases (62.9%) based on ultrasound findings of polyhydramnios and absent gastric bubble. (Table 1).

Table 1: Patient Demographics and Preoperative Characteristics

Characteristic	Type C EA/TEF (n=52)	Isolated EA (n=18)	P-value
Gestational Age (weeks), mean ± SD	37.4 ± 2.1	38.1 ± 1.8	0.14
Birth Weight (grams), mean ± SD	2,790 ± 480	2,950 ± 420	0.12
Male Sex, n (%)	30 (57.7%)	10 (55.6%)	0.87
Premature (<37 weeks), n (%)	14 (26.9%)	4 (22.2%)	0.71
VLBW (<1500 g), n (%)	8 (15.4%)	2 (11.1%)	0.68
Antenatal Diagnosis, n (%)	32 (61.5%)	12 (66.7%)	0.68
Associated Anomalies, n (%)	36 (69.2%)	8 (44.4%)	0.047*
– Cardiac, n (%)	22 (42.3%)	4 (22.2%)	0.12
– Vertebral, n (%)	14 (26.9%)	2 (11.1%)	0.17
– Renal, n (%)	10 (19.2%)	2 (11.1%)	0.43
VACTERL Association, n (%)	18 (34.6%)	2 (11.1%)	0.049*

SD = standard deviation; VLBW = very low birth weight; *statistically significant (P<0.05)

Operative Findings and Surgical Approach: Open repair via right posterolateral thoracotomy was performed in 44 patients (62.9%), while thoracoscopic repair was attempted in the remaining 26 (37.1%). Of the latter group, five (19.2%) required intraoperative conversion to open thoracotomy due to respiratory instability or technical difficulty in achieving adequate exposure. Primary esophageal anastomosis was achieved in 64 patients (91.4%), with the remaining six (8.6%) requiring staged repair for long-gap EA. The mean gap length was 1.9 ± 0.9 cm in the open group and 1.8 ± 0.8 cm in the thoracoscopic group. Operative time was significantly longer in the thoracoscopic group (198.6 ± 41.7 min vs. 145.2 ± 32.4 min; P<0.001). Median ICU stay was 8 days (IQR 5–14) for open and 7 days (IQR 4–12) for thoracoscopic repair, while median total hospital stay was 22 and 20 days, respectively. No statistically significant differences were identified between the two surgical approaches in terms of primary anastomosis rate, complication rates, or hospital stay duration. (Table 2)

Table 2: Operative Findings and Surgical Approach Comparison

Variable	Open Repair (n=44)	Thoracoscopic Repair (n=26)
Age at Surgery (days), median (IQR)	2 (1–3)	2 (1–4)
Operative Time (min), mean ± SD	145.2 ± 32.4	198.6 ± 41.7
Primary Anastomosis, n (%)	40 (90.9%)	24 (92.3%)
Gap Length (cm), mean ± SD	1.9 ± 0.9	1.8 ± 0.8
Long-Gap EA (>3 cm), n (%)	6 (13.6%)	4 (15.4%)
Azygos Vein Preserved, n (%)	18 (40.9%)	14 (53.8%)
Chest Drain Placed, n (%)	40 (90.9%)	22 (84.6%)
Conversion to Open, n (%)	N/A	5 (19.2%)
ICU Stay (days), median (IQR)	8 (5–14)	7 (4–12)
Hospital Stay (days), median (IQR)	22 (16–34)	20 (15–30)

IQR = interquartile range; ICU = intensive care unit; N/A = not applicable; *P<0.001 for operative time comparison between groups

Postoperative Complications and Mortality: Postoperative complications were recorded in 52 of 70 patients (74.3%). Anastomotic leak, confirmed by contrast esophagogram on postoperative day 7, occurred in 12 patients (17.1%), with the majority managed conservatively with prolonged transanastomotic tube feeding and broad-spectrum antibiotic therapy. Anastomotic stricture was the single most common early complication, developing in 28 patients (40.0%); all required endoscopic balloon dilatation, and eight (11.4%) required more than three dilatation sessions. Gastroesophageal reflux disease was documented in 47 patients (67.1%) and represented the most prevalent long-term morbidity; among these, 18 (25.7% of the total cohort) ultimately required Nissen fundoplication. Tracheomalacia was identified in 18 patients (25.7%) and esophageal dysmotility in 34 (48.6%). Recurrent TEF occurred in five patients (7.1%), all requiring re-operative repair. There were four in-hospital deaths (5.7%), all attributable to severe congenital cardiac disease in combination with sepsis, and none were directly related to the esophageal anastomosis or operative technique. (Table 3)

Table 3: Postoperative Complications and Mortality

Complication	Open (n=44)	Thoracoscopic (n=26)	Total N=70, n (%)
Anastomotic Leak	7 (15.9%)	5 (19.2%)	12 (17.1%)
Anastomotic Stricture	18 (40.9%)	10 (38.5%)	28 (40.0%)
Recurrent TEF	3 (6.8%)	2 (7.7%)	5 (7.1%)
Gastroesophageal Reflux Disease	30 (68.2%)	17 (65.4%)	47 (67.1%)
Tracheomalacia	12 (27.3%)	6 (23.1%)	18 (25.7%)
Pneumonia / Respiratory Complications	14 (31.8%)	8 (30.8%)	22 (31.4%)
Esophageal Dysmotility	22 (50.0%)	12 (46.2%)	34 (48.6%)
Nissen Fundoplication Required	12 (27.3%)	6 (23.1%)	18 (25.7%)
Mortality	3 (6.8%)	1 (3.8%)	4 (5.7%)

GERD = gastroesophageal reflux disease; TEF = tracheoesophageal fistula. Percentages are of group totals.

Long-Term Follow-Up Outcomes: Of the 66 surviving neonates, structured follow-up was completed in 60 (90.9%) at one year, 52 (78.8%) at three years, and 44 (66.7%) at five years. GERD on proton pump inhibitor therapy declined progressively over follow-up, from 56.7% at one year to 40.9% at five years, reflecting some natural resolution alongside the effect of surgical anti-reflux procedures. Dysphagia, however, remained persistently prevalent, affecting approximately 46% of patients at all three time points. Recurrent anastomotic stricture requiring repeat endoscopic dilation was present in 16 patients (26.7%) at one year, reducing to 6 (13.6%) at five years. Barrett's esophagus was detected endoscopically in four patients (9.1%) by the five-year follow-up, all of whom had long-standing GERD. Growth was normal (weight-for-age Z-score ≥-2 SD) in 42 patients (70.0%) at one year, improving to 36 (81.8%) at five years. Full oral feeding without tube supplementation was achieved in 48 patients (80.0%) at one year and in 42 patients (95.5%) by five years, indicating progressive and meaningful functional recovery in the majority of survivors. (Table 4)

Table 4: Long-Term Follow-Up Outcomes at 1, 3, and 5 Years

Long-Term Outcome Parameter	1-Year Follow-Up n=60	3-Year Follow-Up n=52	5-Year Follow-Up n=44
GERD (on medication), n (%)	34 (56.7%)	26 (50.0%)	18 (40.9%)
Dysphagia, n (%)	28 (46.7%)	24 (46.2%)	20 (45.5%)
Recurrent Anastomotic Stricture, n (%)	16 (26.7%)	10 (19.2%)	6 (13.6%)
Endoscopic Dilation Required, n (%)	20 (33.3%)	14 (26.9%)	8 (18.2%)
Respiratory Symptoms (wheezing/cough), n (%)	22 (36.7%)	18 (34.6%)	14 (31.8%)
Normal Growth (weight-for-age ≥-2SD), n (%)	42 (70.0%)	38 (73.1%)	36 (81.8%)
Normal Oral Feeding (without tube), n (%)	48 (80.0%)	46 (88.5%)	42 (95.5%)
Barrett's Esophagus (endoscopy), n (%)	0 (0%)	2 (3.8%)	4 (9.1%)
Surgical Re-intervention, n (%)	10 (16.7%)	6 (11.5%)	4 (9.1%)

GERD = gastroesophageal reflux disease; SD = standard deviation. Follow-up denominators reflect surviving patients attending clinic at each time point.

Discussion

This study provides a comprehensive longitudinal account of surgical outcomes and long-term morbidity in neonates undergoing EA repair at a single tertiary centre over an eight-year period. The overall survival of 94.3% compares favourably with contemporary reports from high-income countries, where survival rates typically range from 90% to 97% depending on the severity of associated anomalies,[14] and reflects the impact of improving neonatal intensive care infrastructure and increasing surgical experience.

The predominance of Type C EA (74.3%) in our cohort is consistent with the established distribution in the global literature, where distal TEF accounts for the large majority of cases.[1,3] The high prevalence of associated anomalies (62.9%), with cardiac defects being the most common, mirrors findings from large multi-institutional series and further underscores the importance of pre-repair echocardiography and multidisciplinary team involvement in the perioperative management of these neonates.[7,9] All four deaths in this cohort were attributable to complex cardiac disease rather than operative failure, a finding consistent with published data indicating that cardiac anomalies remain the principal determinant of mortality in EA.[5]

The anastomotic leak rate of 17.1% observed in this series is within the range reported in the literature, which varies from 10% to 25%.[4,11] Anastomotic stricture, occurring in 40.0% of patients, was the most prevalent early complication, consistent with rates documented in other institutional series and systematic reviews.[13,15] The risk factors for stricture formation—anastomotic tension, anastomotic leak, and GERD—are well recognized. In our cohort, the co-occurrence of GERD was closely linked to stricture development, supporting the pathophysiological mechanism of acid-induced scar formation at the anastomotic site.[16] The majority of strictures responded to repeated endoscopic balloon dilatation, with a small minority requiring surgical revision, an observation broadly consistent with published series.[13]

The 67.1% prevalence of GERD in this cohort is consistent with the extensive literature documenting that GERD is an almost universal accompaniment of EA repair. Studies from tertiary centres have reported GERD rates ranging from 40% to 95% depending on the definition used, the length of follow-up, and whether pH-metry or symptom-based criteria are applied.[11,16] The pathophysiology of GERD following EA repair is multifactorial, encompassing intrinsic esophageal dysmotility (which we observed in 48.6% of our cohort), disruption of the gastroesophageal junction during dissection, and absence of a competent lower esophageal sphincter mechanism.[17] The anti-reflux surgery rate of 25.7% in our series is consistent with published rates of 20–35% reported in the literature.[18]

The detection of Barrett's esophagus in 9.1% of patients by the five-year follow-up point is a clinically significant finding. Long-term adult follow-up studies have documented Barrett's esophagus in 10–15% of EA survivors, with isolated reports of esophageal adenocarcinoma.[15,17] The emergence of this finding even within the first five years of life highlights the importance of initiating endoscopic surveillance early, particularly in children with persistent symptomatic GERD, and of maintaining transition pathways into adult gastroenterology services.[15]

Regarding surgical approach, no statistically significant differences in complication rates, mortality, anastomotic leak, or stricture formation were identified between open and thoracoscopic repair groups. This is broadly consistent with the findings of large multi-institutional studies, including data from the American College of Surgeons NSQIP-Pediatric database[12] and comparative analyses from the Midwest Pediatric Surgery Consortium, which similarly found no difference in perioperative outcomes by approach other than operative time.[1] The conversion rate of

19.2% for thoracoscopy in our cohort is somewhat higher than the 9–12% reported in specialized Western centres, likely reflecting the evolving learning curve at our institution during the study period.[13]

Long-term functional outcomes were encouraging. Full oral feeding was achieved in 95.5% of surviving patients at five years, and growth parameters normalized progressively over the follow-up period, with 81.8% achieving normal weight-for-age Z-scores by five years. These findings underscore that, while the operative period is associated with significant morbidity, most children achieve meaningful functional recovery over time with appropriate multidisciplinary support. However, the persistence of dysphagia in approximately 46% of patients across all three follow-up time points, driven largely by esophageal dysmotility and recurrent stricture, highlights the ongoing need for coordinated speech therapy, dietetic, and gastroenterological input throughout childhood. [5,19]

The findings from this study have practical implications for the design of follow-up protocols. We recommend a minimum surveillance schedule of upper gastrointestinal endoscopy at six months post-repair and at one, three, and five years, with pH-impedance testing or manometry reserved for patients with persistent symptoms or GERD refractory to medical therapy. Pulmonary function testing and bronchoscopy should be considered for children with recurrent lower respiratory tract infections or persistent wheeze, given the high prevalence of tracheomalacia and esophageal dysmotility contributing to aspiration risk in this population. [12,20]

Limitations

This study has several limitations that warrant acknowledgment. The retrospective design carries inherent risks of selection and information bias. The relatively modest cohort size from a single centre limits generalizability and statistical power for sub-group analyses. Loss to follow-up at five years (33.3% of survivors) may introduce attrition bias, potentially under-representing patients with severe morbidity or adverse outcomes. The absence of standardized quality-of-life instruments limits interpretation of functional outcomes. pH-impedance studies and esophageal manometry were not uniformly performed across the cohort, which may have led to underestimation of GERD and dysmotility prevalence.

Conclusion

EA repair can be performed with acceptable short-term outcomes and low operative mortality in a tertiary pediatric surgical centre. However, long-term morbidity—particularly GERD, anastomotic stricture, dysphagia, and esophageal dysmotility—remains highly prevalent and requires sustained, structured surveillance throughout childhood. The emergence of Barrett's esophagus even within five years further reinforces the critical importance of early and ongoing endoscopic monitoring. Both open thoracotomy and thoracoscopic repair offer comparable outcomes when performed by experienced surgical teams.

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