

Twenty-four years of esophageal atresia management: Evolution of practice and outcomes in a tertiary pediatric surgery center

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Esophageal atresia (EA) is a congenital anomaly that continues to pose significant challenges in both diagnosis and management. Despite advancements in medical technology and surgical techniques, EA remains a complex condition often associated with considerable morbidity and mortality, particularly in cases involving associated anomalies or long-term complications such as esophageal dysmotility and gastroesophageal reflux disease (GERD). Recent progress in prenatal imaging, neonatal intensive care, and minimally invasive surgical techniques has improved survival rates and overall patient outcomes. However, significant challenges persist in optimizing care for patients with complex presentations or severe comorbidities.

This retrospective observational study aimed to present the clinical experiences of a high-volume referral center over a 24-year period. By analyzing operative approaches, postoperative outcomes, and long-term complications, we sought to highlight the progress achieved, the challenges that persist,

Abstract

Objectives: This study aims to evaluate clinical outcomes, operative approaches, and associated challenges in esophageal atresia (EA) patients treated over 24 years at our institution.

Patients and methods: This retrospective observational study included 97 EA patients (60 males, 37 females, mean age 5.7 years; range, 4 month to 17.2 years) managed between January 2000 and January 2024. Patient data, including demographics, prenatal findings, EA types, associated anomalies, operative details, and complications, were collected from hospital records. Cases without complete data were excluded from analysis.

Results: The male-to-female ratio was 1:0.65. The median birth weight was 2400 g, and the median gestational age was 37 weeks. Prenatally, polyhydramnios was present in 33 patients, with definitive diagnosis in 17. Esophageal atresia types included type A (n=11), type B (n=1), type C (n=83), and type D (n=2). Primary esophageal repair was achieved in 81 cases. Anastomotic strictures (15%) and GERD requiring intervention (20%) were the most common complications. Mortality was 16%, predominantly related to severe cardiac anomalies and pulmonary hypertension.

Conclusion: Advances in neonatal intensive care, pediatric anesthesia, and surgical techniques have significantly improved EA outcomes at our institution. Nevertheless, significant morbidity persists due to associated anomalies and long-term complications such as dysmotility, strictures, and gastroesophageal reflux disease. Enhanced interdisciplinary collaboration and targeted research remain critical to further optimize management strategies and patient care.

Keywords: Esophageal atresia, foker, long-term outcomes, surgical management, thoracoscopic repair.

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and the critical factors influencing the prognosis of children with EA.

PATIENTS AND METHODS

This retrospective observational study included patients diagnosed with EA who were treated at İstanbul University-Cerrahpaşa, Cerrahpaşa

Faculty of Medicine, Department of Pediatric Surgery between January 2000 and January 2024. A total of 135 patients were managed during the study period; however, complete clinical data were available for 97 patients (60 males, 37 females, mean age 5.7 years; range, 4 month to 17.2 years), and only these cases were included in the analysis. Data for the remaining 38 patients could not be accessed due to incomplete records and were therefore excluded from the analysis. Written informed consent was obtained from the parents of all participants. The study protocol was approved by the İstanbul University Cerrahpaşa Non-Interventional Clinical Research Ethics Committee (Date: 22.01.2025, No: 2025/125). The study was conducted in accordance with the principles of the Declaration of Helsinki.

Patient data were collected from hospital medical records and included demographic characteristics, prenatal findings, EA classification, associated anomalies, operative approach, postoperative complications, and clinical outcomes. The classification of EA types was based on the Gross classification system. Associated anomalies were also recorded, with special attention to VACTERL association. Cases without fistula, with isolated proximal fistulas, and those in which the distance between the ends under traction was greater than 1 cm after the complete mobilization of both ends were considered as long-gap EA. Patients with H-type tracheoesophageal fistula were excluded from the study due to their distinct clinical characteristics and the presence of intact esophageal continuity.

All cases were reviewed for operative strategy, timing of surgical repair (primary *vs.* delayed), and need for esophageal replacement. Since 2020, thoracoscopic exploration has been employed in patients over 2000 g of birth weight in our institution. The criteria for esophageal replacement included long-gap EA and failure of primary anastomosis. Techniques such as the Foker procedure, colon interposition, and gastric pull-up were used depending on anatomical and clinical conditions.

Postoperative complications were classified as early (anastomotic leak, sepsis, renal failure, and vocal cord paralysis) or late (recurrent fistula, anastomotic stricture, and GERD). Long-term

follow-up data included the need for dilatation, fundoplication, or reintervention. Mortality was defined as any death occurring before hospital discharge or within the first year of life following the initial surgery related to esophageal atresia.

Statistical analysis

Descriptive statistics were used to summarize the data. The Shapiro-Wilk test was applied to assess the normality of distribution. Continuous variables were expressed as medians with minimum and maximum values due to nonnormal distributions. Categorical variables were summarized as counts and percentages.

RESULTS

Polyhydramnios was identified in 33 cases, and a definitive prenatal diagnosis was established in 17. The male-to-female ratio was 1:0.65. The median birth weight was 2400 g (range, 905 to 3950 g), and the median gestational age was 37 weeks (range, 27 to 40 weeks). According to Gross classification, 11 patients had type A, 83 had type C, one had type B, and two had type D EA (Table 1). A total of 62 associated anomalies were recorded, among which VACTERL

TABLE 1		
Types of esophageal atresia according to Gross classification		
Gross classification	n	%
Type A	11	11
Type B	1	1
Type C	83	86
Type D	2	2

TABLE 2		
Associations and anomalies		
Associations and anomalies	n	%
Cardiovascular	49	50
Urinary	16	16
Anorectal malformation	15	15
Vertebral	10	10
Limb	7	7
VACTERL	20	20
Duodenal obstruction	6	6

association was observed in 20 cases (Table 2). The most common concomitant anomalies were cardiovascular (n=49).

Bronchoscopy was used at the beginning of the procedure in the most recent 57 cases. Primary esophageal repair was achieved in 82 cases. Of these, 78 underwent neonatal primary repair, while delayed primary repair was performed in four patients. The Foker technique was utilized in two of these delayed cases, while remaining two patients underwent repair at three months of age. Esophageal replacement was required in 12 cases. Due to hemodynamic instability, three cases underwent only gastrostomy without further surgical intervention. Surgical outcomes and flowchart are summarized in Figure 1.

In the most recent 12 cases, initial surgical exploration was attempted using thoracoscopy in nine neonates weighing over 2000 g. In the first three cases, conversion to open surgery was required after fistula ligation due to inadequate visualization, and primary anastomosis was then completed via thoracotomy. Among the remaining six cases, primary anastomosis was achieved in

four patients with distal fistulas. Of the other two patients with isolated EA, one underwent delayed primary repair, and the other required esophageal replacement after an unsuccessful Foker attempt.

Seventeen cases were classified as long-gap esophageal atresia. Among these, 11 were isolated, one had proximal fistula, and five had distal fistula. Delayed primary anastomosis was performed in two cases of isolated atresia. The Foker procedure was applied to five patients. Among these, staged primary repair was achieved in two cases as previously mentioned. In two other cases, anastomosis could not be performed, necessitating the creation of an esophagostomy and gastrostomy, followed by colon interposition. In the last case, during the second exploration, gastric pull-up was performed while preserving the cardia and distal esophagus. Overall, esophageal replacement was required in 12 cases. Eleven patients underwent esophagostomy with gastrostomy. Among these, two died before replacement could be performed, and two were lost to follow-up. Colon interposition was performed in seven cases. Three patients died before definitive surgical intervention due to cardiac issues.

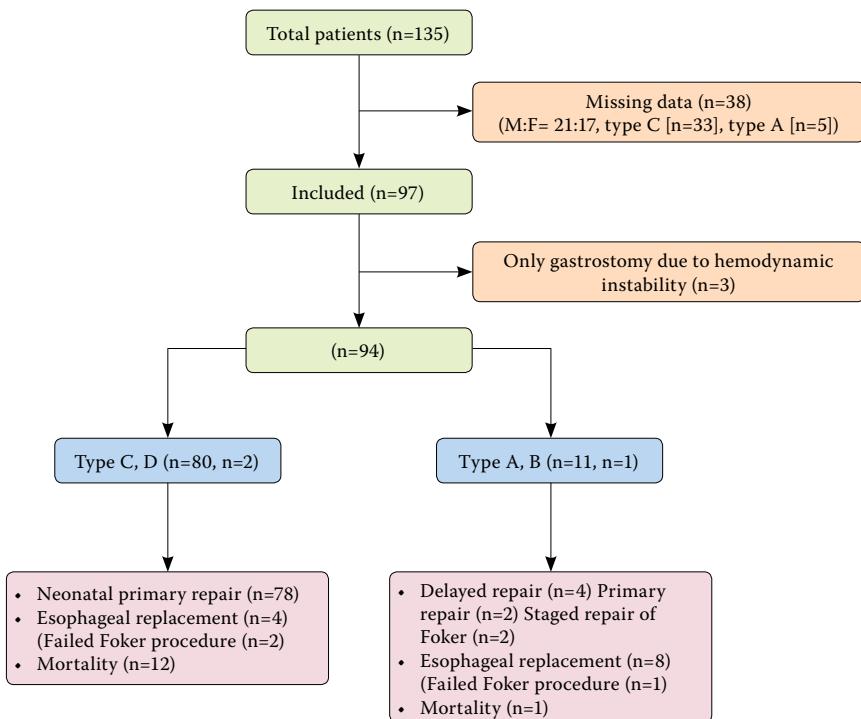


Figure 1. Patient inclusion and management flowchart by EA type.

EA: Esophageal atresia.

TABLE 3
Complications

Complications	n	%
Anastomotic leak	18	18
GERD	42	43
Anastomotic stricture	15	15
Fistula recurrence	1	1
Delayed diagnosed proximal fistula	1	1

GERD: Gastro-esophageal reflux disease.

TABLE 4
Secondary surgical procedures

Surgical management	n	%
Fundoplication	22	22
Balloon dilatation of anastomotic stricture	15	15
Need to gastrostomy following primary anastomosis	12	12
Colostomy	8	8
Colon interposition	7	7
Duodeno-duodenostomy	6	6
Tracheostomy	3	3
Proximal fistula repair (cervical approach)	1	1
Gastric pull-up	1	1
Total	97	100

Early postoperative complications included anastomotic leakage in 18 cases, 16 of which resolved spontaneously. A total of 27 cases were treated for sepsis, including those with leaks. Temporary acute renal failure occurred in two patients with a solitary kidney. One case of vocal cord paralysis required tracheostomy.

Late complications included the identification and surgical repair of a previously undiagnosed proximal fistula via a cervical approach in one patient. Another patient developed recurrent tracheoesophageal fistula. Patients with a recurrent fistula died during the late postoperative period.

Anastomotic stricture developed in 15 cases during follow-up and was managed successfully via endoscopic dilatation and steroid injection. Gastroesophageal reflux disease was controlled medically in 22 cases, while 20 underwent fundoplication. Thirty-nine patients remained asymptomatic for GERD (Table 3). Secondary surgical procedures are summarized in Table 4.

A total of 16 patients died during follow-up, primarily due to severe cardiac anomalies or pulmonary insufficiency (Table 5). All cardiac-related deaths were associated with severe pulmonary hypertension. These included four patients with complex anomalies such as atrioventricular valve atresia and complete atrioventricular septal defects, four with large ventricular septal defects and patent ductus arteriosus, and one with isolated severe pulmonary hypertension. Pulmonary insufficiency accounted for four deaths. Other causes included prematurity related severe intraventricular hemorrhage, uncontrolled hyperammonemia, and neonatal sepsis.

The median postoperative follow-up duration was 60.3 (range, 4 to 210) months, and the median length of hospital stay was 20 (range, 5 to 180) days. For patients managed with thoracotomy or thoracoscopy and anastomosis, the median chest tube duration was 7 (range, 6 to 80) days.

DISCUSSION

Esophageal atresia is a condition that presents significant challenges in neonatal care and necessitates timely and appropriate surgical intervention. Over the years, substantial progress has been made in the diagnosis and management of EA, owing to improvements in neonatal intensive care, prenatal imaging, and surgical techniques. The present study summarized this evolving process.

In the current cohort, a decrease in the number of EA cases was observed compared to our previous series.^[1] However, consistent with the literature, there has been a marked decline in both morbidity and mortality. This trend is attributed to advancements in diagnostic and therapeutic techniques, increasing clinical experience, and improvements in neonatal intensive care unit conditions.^[2] In our center, high-frequency oscillatory ventilation and advanced medical treatment options for premature infants are available. However, extracorporeal membrane oxygenation support for neonates is not currently accessible. If it had been available, it might have made a difference in the course of some patients.^[3] Among the losses, the majority occurred due to pulmonary hypertension associated with cardiac anomalies and bronchopulmonary dysplasia.

TABLE 5						
Causes of death in cases of esophageal atresia						
No	Sex	Birth weight (g)	Gestational age (weeks)	Gross classification	Cause of death	Age at death (days)
1	F	2100	37	C	Pneumonia and respiratory failure (anastomotic leak, under control)	13
2	M	2220	36	C	Pulmonary hypertension and cardiac failure due to congenital heart defect (left atrial isomerism, single AV valve, pulmonary atresia), sepsis	64
3	M	3950	37	C	Pneumonia and respiratory failure	38
4	F	2750	35	C	Pulmonary hypertension and cardiac failure (VSD, large PDA)	12
5	M	2000	35	C	Pulmonary hypertension and cardiac failure (VSD, ASD, large PDA)	16
6	M	1600	31	A	Severe intraventricular hemorrhage (CPR at birth, perinatal asphyxia)	21
7	M	2630	37	C	Pulmonary hypertension and cardiac failure (right atrial isomerism, AV valve atresia)	16
8	F	2630	38	C	Pulmonary hypertension and respiratory failure (VSD, large PDA, recurrent TEF diagnosed at three months postoperative)	180
9	F	3150	37	A	Pulmonary hypertension and cardiac failure (large PDA)	8
10	M	1390	30	C	Hyperammonemia (uncontrolled)	16
11	F	1190	29	C	Pneumonia and respiratory failure (anastomotic leak, under control)	21
12	M	2405	36	C	Pulmonary hypertension and cardiorespiratory failure (AVSD, large PDA)	81
13	M	1990	34	A	Pulmonary hypertension and respiratory failure (suspected pulmonary hypoplasia, uncontrolled PH, BPD)	109
14	F	1525	36	C	Pulmonary hypertension and cardiorespiratory failure (right atrial isomerism, AV valve atresia)	90
15	M	1370	31	D	Pneumonia and sepsis	17
16	M	905	29	C	Early neonatal sepsis	5

AV: Atrioventricular; VSD: Ventricular septal defect; PDA: Patent ductus arteriosus; ASD: Atrial septal defect; CPR: Cardiopulmonary resuscitation; TEF: Tracheoesophageal fistula; AVSD: Atrioventricular septal defect; PH: Pulmonary hypertension; BPD: Bronchopulmonary dysplasia.

Prenatal diagnosis plays an important role in improving the prognosis of infants with esophageal atresia. Early diagnosis allows for timely intervention, optimization of perioperative care, and informed decision-making by both healthcare providers and families. This proactive approach enables medical teams to prepare in advance for specialized care, thereby contributing to improved outcomes and enhanced quality of life for affected children.^[4] In our earlier study, the mean time to surgery was 60 h, and 14.7% of the cases

were home births.^[1] However, when evaluating the current cohort independently, the findings do not support this association. Among the 17 cases with a prenatal diagnosis, 13 were diagnosed in the presence of additional anomalies such as cardiac, renal, anorectal, and cleft lip/palate anomalies, and four (23%) of these patients were lost. Of the 33 cases in which polyhydramnios was present but EA was not prenatally diagnosed, four (12%) died. In the group of 47 patients whose prenatal follow-up was unremarkable, eight (17%) were lost.

It is reasonable to expect a poorer prognosis in cases with multiple anomalies.^[5]

Associated anomalies have been reported with a frequency ranging from 50 to 73% in various series.^[6,7] Beyond VACTERL-associated anomalies, duodenal atresia and trisomies occur more frequently in EA cases.^[6-9] The detection of patent foramen ovale or patent ductus arteriosus, which may still be physiological in neonates, by preoperative echocardiography could contribute to higher reported prevalence of cardiac anomalies. In our study, control echocardiography findings for patients with patent foramen ovale and/or patent ductus arteriosus were reviewed, and this possibility was ruled out. Moreover, anorectal malformations and duodenal obstructions were observed more frequently in our series compared to the literature.^[10] In terms of classification, the incidence of atresia without fistula was higher in our series than rates reported in the literature.^[10] These findings reflect the referral center status of our institution, where more complex and challenging cases are managed. This aligns with the higher number of cases requiring esophageal replacement and the greater frequency of mortality due to cardiac and pulmonary complications compared to current series.

Significant advancements have been made in surgical approaches in recent years. Preoperative bronchoscopy, thoracoscopy, and elongation techniques for patients with long-gap atresia have become prominent components of contemporary surgical practice.^[11,12] Preoperative bronchoscopy enables the preoperative assessment of tracheomalacia, proximal fistula, the level of the distal fistula, and tracheobronchial anomalies. Anatomical findings obtained from bronchoscopy can alter the surgical strategy and also provide critical insights for the evaluation of postoperative complications. The routine use of thoracoscopy in practice has revolutionized surgical approach by offering a minimally invasive alternative to traditional open procedures. These advances provide significant benefits, including reduced tissue trauma, improved surgical visualization, and potentially lower complication rates, thereby enhancing patient outcomes and improving the quality of life for both children and their families.^[11,12] To avoid complications associated

with esophageal replacement, there has been a growing interest in elongation procedures in recent years.^[12,13] Decisions regarding open surgery and esophageal replacement options are now made with much greater selectivity and deliberation compared to the past.

In our series, prior to 2012, the proximal esophageal pouch was commonly assessed using a pouchogram, while bronchoscopy was not routinely performed. During this period, cases of chemical pneumonitis secondary to contrast aspiration were observed, despite the use of nonionic contrast agents. To prevent such complications, the use of pouchograms was discontinued. However, during a short interval before routine bronchoscopy was implemented, a proximal fistula remained undiagnosed in one case. This patient, who was diagnosed late and developed pulmonary insufficiency, prompted a change in our institutional practice. Since then, all procedures have been initiated with bronchoscopy, regardless of birth weight. To date, no complications associated with this approach have been observed.

Our experience with thoracoscopic EA repair is based on a limited number of patients. Thoracoscopic primary anastomosis was achieved in five cases. Based on our limited experience, we can state that patients without anastomotic leakage after thoracoscopy demonstrated a remarkably rapid and uneventful recovery. On the other hand, in cases where leakage occurred, hospital stay and chest tube duration were prolonged, and broad-spectrum antibiotics were required to control sepsis. Observationally, the postoperative course in these cases appeared to be more turbulent than that typically observed in patients who develop leaks following open extrapleural repair. With advancements in thoracoscopic instrumentation and increasing surgical experience, a growing number of studies report improved outcomes and suggest that this approach can be safely applied, even in infants with lower birth weights.^[13,14] However, data on long-term outcomes remain limited. From a technical standpoint, thoracoscopic repair differs from open surgery in several ways. We suggest that thoracoscopy should be the first choice for suitable patients. Nonetheless, surgeons should adhere to the principle that “the best esophagus is the patient’s own esophagus” and patient safety

must always come first. If sufficient skills and experience in both anesthetic management and thoracoscopic technique are lacking, any attempts that could jeopardize a safe primary anastomosis should be strictly avoided.

Long-gap EA is studied as a subcategory with varying anatomical definitions. In most cases of isolated atresia, proximal fistula, and in some cases with a distal fistula, primary anastomosis cannot be performed. These cases may require various approaches, including the timing and content of the operation. The common current approach for atresia without a fistula involves early gastrostomy, followed by surgery around eight to 12 weeks of age. Exploration by thoracotomy or thoracoscopy, if feasible, aims for primary anastomosis. If primary anastomosis is not feasible, secondary exploration following internal or external traction is a widely accepted practice with high success rates.^[13] Adopting the same approach for exploring distal fistula atresia cases in the neonatal period is another viable alternative. If traction fails and primary anastomosis cannot be performed safely during the initial exploration, direct esophagostomy/gastrostomy, direct gastric pull-up or gastric tube for esophageal replacement are potential alternatives. In our series, there were 17 cases of long-gap esophageal atresia, and different surgical management approaches were used over the years. Our series includes experiences with all these approaches.

The Foker technique is a pivotal approach in managing long-gap EA, particularly in cases where the upper and lower esophageal segments cannot be directly anastomosed using conventional surgical methods. This technique offers several advantages. Primarily, it facilitates natural esophageal growth, reducing the need for complex grafting or replacement procedures. Additionally, the gradual approximation process may lower the risk of complications such as anastomotic leaks and strictures compared to immediate repair.^[15] However, traction-based techniques are not without limitations. The process is often prolonged, requiring careful monitoring and frequent adjustments of traction sutures over time. Successful implementation also demands significant surgical expertise, particularly in managing the complexities of tissue elongation.^[16] Our clinical experience reflects both the

benefits and challenges of using the Foker technique. In our series, primary anastomosis was achieved in two cases through thoracotomy at three months. However, in the remaining three cases, anastomosis could not be performed. Intraoperative findings in these cases revealed tissue erosion and signs of infection at the traction suture sites. Despite satisfactory approximation of the esophageal ends during secondary exploration, anastomosis was not deemed feasible. Two patients underwent esophagostomy and gastrostomy in the neonatal period, while one underwent gastric pull-up at eight weeks. At the time of this writing, all three patients were fully orally fed without the need for gastrostomy or nasogastric support. These observations suggest that while the Foker technique facilitates esophageal approximation, it does not guarantee successful anastomosis, particularly during early exploration of EA with fistula in neonates.

Postoperative surgical outcomes have improved over the years due to advancements in surgical and medical practices in neonatal care.^[17] However, despite a reduction in mortality, it is still not possible to achieve universally satisfactory outcomes for all cases of EA.^[18-20] While conditions for patient care have improved, morbidity has increased with the survival of higher-risk patients, and mortality has not been entirely eliminated.^[21] When examining the causes and risk factors for mortality, prematurity, birth weight, major anomalies, and pulmonary condition emerge as significant factors. Notably, the association between cardiac anomalies and mortality is well-documented.^[21,22] Similarly, in our series, deaths attributed to cardiac anomalies accounted for the majority of total mortality. Compared to other studies in literature, a unique finding was observed in our study. This finding revealed that the common feature of cardiac anomalies in fatal cases was their association with systemic pulmonary hypertension. Four cases with the tetralogy of Fallot and patent ductus arteriosus requiring invasive intervention after the neonatal period, as well as three cases with ventricular septal defect managed with medical therapy, exhibited uneventful courses without sequelae. Randomized controlled trials are needed to further investigate this observation regarding cardiac anomalies associated with severe pulmonary hypertension.

Ongoing research efforts, coupled with interdisciplinary collaboration and specialized training programs, continue to play pivotal roles in refining outcomes for children undergoing EA repair. This underscores the critical importance of sustained innovation in pediatric surgical care, ensuring that advancements translate into tangible benefits for patients and their families. In our earlier series, the mortality rate was high, with many early deaths.^[1] However, with the improved prognosis, long-term complications are now being observed more frequently. These include GERD, esophageal strictures, tracheomalacia, feeding and nutritional issues, respiratory complications, Barrett's esophagus, and psychological and social issues.^[23,24] Improved intensive care conditions have reduced mortality by enabling the survival of low-birth-weight infants and those with major congenital anomalies. However, the survival of these high-risk and critical patients has also led to an increase in long-term complications.

This study had several limitations. First, this study had a retrospective observational design and did not include comparative or statistical evaluation of risk factors or treatment outcomes. Second, the data were collected over a long time period, during which surgical techniques, neonatal care protocols, and diagnostic methods evolved, introducing potential inconsistencies in patient management. Third, 38 patients were excluded due to incomplete records, which may have introduced selection bias. Additionally, long-term follow-up data were incomplete for some cases, limiting conclusions about late complications and functional outcomes. Lastly, this study was conducted at a single center, and the findings may not be generalizable to other institutions or healthcare settings with differing resources and protocols.

In conclusion, this 24-year retrospective experience from a single tertiary center highlights both the progress and the ongoing challenges in the management of esophageal atresia. Improved survival rates reflect advancements in neonatal intensive care, surgical techniques, and multidisciplinary collaboration. However, the increased survival of high-risk patients has led to a greater burden of long-term complications such as GERD, strictures, and dysmotility. Observational data support the careful use of techniques such

as thoracoscopy and the Foker procedure, while underscoring the importance of early diagnosis and individualized management strategies. Continued efforts in surgical training, long-term follow-up, and collaborative multicenter studies are essential to further enhance outcomes and improve quality of life for patients with EA.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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