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Evaluation of Delayed Surgical Repair of Esophageal Atresia in Patients with Distal Tracheoesophageal Fistula in Suez Canal University Hospitals

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ABSTRACT

Background: Esophageal atresia (EA) involves complex congenital esophageal abnormalities, likely caused by disruptions in embryological foregut separation.

Aim: This study aimed to evaluate delayed EA/TEF repair outcomes, focusing on morbidity, mortality, complications, short-term function, and perioperative status for effectiveness.

Patients and methods: This retrospective investigation has been conducted at the Pediatric Surgery Unit, General Surgery Department at Suez Canal University Hospitals on 35 cases who had tracheoesophageal fistula (TEF).

Results: Postoperative complications were categorized by repair timing in 35 cases: repair within 48 hours to one week (n=10), during the first week (n=12), after the first week but before 10 days (n=6), and after 10 days (n=7). Mortality increased with delay, peaking at 42.8% in repairs after 10 days. Pneumothorax occurred in repairs within the first week (8.3%) and after 10 days (28.5%). Strictures were highest (28.5%) in repairs after 10 days. Major anastomotic leakage occurred only after one week, while minor leakage increased with delay, reaching 71% after 10 days. Repair after the first week had higher complications than repair within the first week, though differences were insignificant ($p > 0.05$). However, mortality was significantly higher in repairs after the first week ($p=0.019$).

Conclusion: The timing of esophageal atresia and tracheoesophageal fistula repair is crucial for outcomes. Early repair reduces complications, while delayed repair increases risks but allows stabilization. Misdiagnosis at birth due to inadequate prenatal detection raises complication risks. A study found 30/35 survival, with 5 deaths. Complications were higher in delayed repair cases.

Keywords: TEF, EA**,** Strictures, Pneumothorax.

INTRODUCTION

Esophageal atresia refers to a congenitally interrupted esophagus. One or more fistulas could be present between the anomalous esophagus and the trachea. The lack of esophageal patency inhibits swallowing. This issue obstructs regular eating and may result in aspiration, possibly causing neonates to suffocate on their own saliva, which quickly accumulates in the upper pouch of the clogged esophagus. In the presence of a tracheoesophageal fistula, fluids, including saliva from above or stomach secretions from below, can directly enter the tracheobronchial tree [(1)].

Esophageal atresia comprises a variety of intricate congenital malformations of the esophagus, likely resulting from embryological disturbances throughout the normal separation of the foregut. The worldwide occurrence, based on national and international registries, currently varies from 1.27 to 4.55 per 10,000 births. Isolated esophageal atresia without tracheoesophageal fistula is a rare anomaly, seen in approximately eight percent of neonates with esophageal atresia, with an estimated occurrence rate of 1 in 40,000 births [(2)].

The timing of surgical repair is crucial and depends on various factors, including the newborn's overall condition, the presence of a tracheoesophageal fistula, associated congenital anomalies, and the gap between the esophageal segments. Repair could be immediate, within the first 48 hours of life, delayed within the first 2 weeks, or staged repair [(3)].

Strictures (ASs), anastomotic leakages (ALs), recurrent tracheoesophageal fistulas (R-TOFs), and gastroesophageal reflux constitute the majority of postoperative complications, with their prevalence predominantly dependent on the occurrence of anastomotic leakages (ALs), strictures (ASs), and gastroesophageal reflux (GER). Pediatric surgeons find it challenging to correctly identify and manage these PCs [(4)].

The investigation aimed to assess outcomes of delayed operative repair for EA/TEF focusing on morbidity, mortality, postoperative complications, and short-term gastroesophageal function. It also assesses perioperative status, providing insights into delayed repair effectiveness, especially in settings with delayed diagnosis.

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PATIENTS AND METHODS

This retrospective investigation was conducted at the Pediatric Surgery Unit, General Surgery Department, at Suez Canal University Hospitals on 35 cases who had TEF.

Inclusion criteria: Cases with distal tracheoesophageal fistula (TEF) who presented after 48 hours of birth, regardless of gender.

Exclusion criteria: Cases with major cardiac anomalies like transposition of the great arteries, tetralogy of Fallot, and coarctation of the aorta, as well as those with multiple congenital anomalies or pure atresia and double fistula.

MATERIAL AND METHODS

The study collected data from medical files at Suez Canal University Hospital. Diagnosis has been verified using chest X-rays with an inserted nasogastric tube (8 French for preterm infants, 10 French for term infants). In cases of esophageal atresia, the tube typically terminates at ten to twelve centimeters, while the normal distance to the gastric cardia is around seventeen centimeters. Chest radiographs (posteroanterior and lateral views) have been utilized to verify tube placement, assess for distal fistula, and evaluate cardiac, vertebral, and pulmonary anomalies. Cases underwent complete medical history, laboratory tests, physical examination, and radiological investigations, including chest X-rays, skeletal surveys, echocardiography, and abdominal ultrasonography to detect associated anomalies.

Preoperative care: The diagnosis of esophageal atresia was confirmed, and cases were prepared for surgical correction. Measures included clearing the oral pharynx, placing an eight French sump tube for continuous suctioning, administering intravenous fluids, oxygen therapy, and antibiotics. Preoperative evaluation included congenital anomalies, chest and abdominal radiographs, echocardiography, renal ultrasonography, and laboratory tests. Surgical repair was delayed in cases of sepsis, pneumonia, coagulopathy, or major anomalies.

Surgical procedure: Operation has been performed under general anesthesia with ultrasound-guided central line insertion and trans-thoracic nerve block. The case was placed in the left lateral decubitus position with the trunk flexed and the head elevated to prevent gastric reflux. A postero-lateral thoracotomy incision has been done 1 cm below the scapula, through the 4th intercostal space, preserving the serratus anterior muscles to avoid nerve injury. The pleura was dissected using wet peanut gauze, and the azygos vein was identified and either preserved or ligated. The distal esophagus was minimally dissected with steady traction to preserve blood supply, and the vagus nerve was identified and preserved. The distal fistula was crushed with right-angle forceps, divided, and closed using non-absorbable sutures with Vicryl 6.0 stay sutures. The proximal esophageal pouch was identified using a Ryle tube and secured with Vicryl 5.0 stay sutures. Repair began at the posterior wall with three Vicryl 6.0 stay sutures tied sequentially to distribute tension evenly and prevent esophageal tears. The anterior wall was then repaired, and saline was injected through the Ryle tube to check for pleural tears and airway integrity. A chest tube and, if needed, an additional intrapleural drain were placed, followed by layered closure.

Postoperative care: Immediately after surgery, cases received care in The Neonatal Intensive Care Unit with continuous follow-up of respiration, body temperature, and cardiac and renal functions. Mechanical ventilation and oxygen therapy have been provided as needed, along with anti-reflux measures and pain management if required. Blood tests were conducted to assess the infant's overall condition, and intravenous total parenteral nutrition was initiated, transitioning to Ryle tube feeding and eventually oral feeding after one week if no leaks were detected. Secretions were suctioned, and chest physiotherapy was applied, with the chest tube clamped for 6 hours and removed after 24 hours. Hospitalization typically lasted one week or longer, depending on complications, with a gastrografin meal X-ray performed postoperatively. Long-term follow-up included evaluations at one week, three months, and six months, with upper GI endoscopy to detect and manage strictures using stents or dilation.

Ethical consideration: Ethical permission has been acquired from The Faculty of Medicine's Suez Canal University Ethical Committee prior to the investigation. Written informed consents were secured from all parents after thoroughly explaining the investigation's aims, methods, potential risks, and benefits of the procedures. Patient confidentiality was strictly maintained throughout and after the research. Cases who declined participation were excluded from the study, though surgical intervention was still provided without including their data in the research. The study was conducted in accordance with Helsinki Standards.

Data management

Information has been gathered and examined utilizing SPSS (Statistical Package for Social Science) version 26.0. Normality assessments were performed to identify the parameter types, subsequently followed by suitable statistical assessments. The Chi-square test (χ^2) was utilized to evaluate the significance of distinctions in qualitative variables, whereas the Student’s t-test was utilized for quantitative variables pre- and post-operation. Supplementary tests were employed if necessary. A p-value ≤ 0.05 has been regarded as statistically significant. Results have been presented as percentage tables, frequencies, and graphics for clarity and interpretation.

RESULTS

Table (1) demonstrated that the cases had a mean gestational age of 30.1 ± 4.9 weeks, which ranged from 26 to 40 weeks. More than half of them (51.4%) were males, and (48.6%) were females. More than half of them (51.4%) delivered vaginally and (48.6%) by CS.

Table (1): Characteristics of the study cases (n=35)

		N= 35	
Maternal age (years)	**Mean ±SD**	33.9±8.2	
	Range	18-47	
Gestational age (weeks)	**Mean ±SD**	30.1±4.9	
	Range	26-40	
Birth weight (grams)	**Mean ±SD**	1680±750	
	Range	700-3500	
Birth weight (grams)	**1500-2500 grams**	18	51.4%
	>2500 grams	17	48.6%
Gender	**Male**	19	54.3%
	Female	16	46.7%
Mode of delivery	**CS**	17	48.6%
	VD	18	51.4%

Quantitative variables were expressed as mean ± SD, while qualitative variables were expressed as numbers and percentages. CS, cesarean delivery, VD, vaginal delivery.

Table (2) demonstrated that the pneumothorax was suspected in 3 (10%) cases, stricture was identified in five (16.7%) infants, major anastomotic leakage was found in two (6.7%) cases, and 10 cases (33.3%) had minor anastomotic leakage.

Table (2): Postoperative complications among the study cases (n=30).

	(n=30)
Pneumothorax	3(10%)
Strictures	5(16.7%)
Major Anastomotic Leakage	2(6.7%)
Minor Anastomotic Leakage	10(33.3%)

Table (3) demonstrated that a total of 4 (13.3%) infants had GERD. 22 cases (73.3%) of the currently living are thriving with normal oral feeds. Dysphagia was seen in a total of 4 cases (13.3%). Recurrent respiratory symptoms after EA/TEF operation were observed in eighteen (sixty percent) kids, with a significant decrease in both frequency and duration over the 1st six-month period of life.

Table (3): Functional follow up results among the study cases (n=30)

	(n=30)
GERD	4(13.3%)
Normal oral feeds	22(73.3%)
Dysphagia	4(13.3%)
Respiratory symptoms	18(60%)

Table (4) demonstrated that died cases had significantly lower BW than survivors ($p < 0.001$) and were more preterm than survivors ($p < 0.001$).

Table (4): Characteristics of the study groups according to mortality (n=35).

	Died cases (n=5)	Survivors cases (n=30)	P-value
Maternal age (years)	37±8	33.3±8.2	0.362[1]
Gestational age (weeks)	26.8±0.5	30.6±5.0	**<0.001*[1]**
Birth weight (grams)	1700±150	2500±200	**<0.001*[1]**
Gender Male	4(80%)	15(50%)	1.00[2]
Female	1(40%)	15(50%)	
Mode of delivery CS	2(40%)	15(50%)	1.00[3]
VD	3(60%)	15(50%)	

The first is the Student's t-test, followed by the Fisher's exact test. 3. Chi-square test. *Statistically significant as $p < 0.05$.

Table (5) demonstrated that the cases that had repair within the 1st week of life had significantly reduced BW than repair after the 1st week of life ($p < 0.001$) and were also more preterm than them ($p < 0.001$).

Table (5): Characteristics of the study groups (n=35)

	Repair within the 1st week of life (n=13)	**Repair after 1st week of life (n=22)**	**P-value**
Maternal age (years)	37±8	33.3±8.2	0.362[1]
Gestational age (weeks)	26.8±0.5	30.6±5.0	**<0.001*[1]**
Birth weight (grams)	1700±150	2500±200	**<0.001*[1]**
Gender Male	8(51.6%)	11(50%)	1.00[2]
Female	5(48.4%)	11(50%)	
Mode of delivery CS	7(53.8%)	11(50%)	1.00[2]
VD	6(46.2%)	11(50%)	

Table (6) categorized postoperative complications based on the timing of surgical repair in 35 cases, divided into four intervals: Repair within 48 hours to one week (n=10), during the first week (n=12), after the first week but before 10 days (n=6), and after 10 days (n=7). Mortality rates increased with delayed repair, peaking at 42.8% in the group repaired after 10 days. Pneumothorax was observed only in cases repaired within the first week (8.3%) and after 10 days (28.5%). Stricture rates were

highest (28.5%) in the group repaired after 10 days. Major anastomotic leakage occurred exclusively in cases repaired after one week, while minor leakage rates increased with delayed repair, reaching 71% in those repaired after 10 days.

Table (6): Postoperative complications according to time of repair (n=35)

	Repair after 48 hours, less than one week of life (n=10)	Repair at the first week of life (n=12)	Repair after first week of life, less than 10 days (n=6)	Repair after 10 days of age (n=7)
Mortality	0 (0%)	0(0%)	2 (33.3%)	3(42.8%)
Pneumothorax	0 (0%)	1(8.3%)	0(0%)	2(28.5%)
Stricture	1(10%)	1(8.3%)	1(16.7%)	2(28.5%)
Major anastomotic leakage	0(0%)	0(0%)	1(16.7%)	1(14%)
Minor Anastomotic Leakage	1(10%)	2(16.7%)	2(33.3%)	5(71%)

Table (7): Repair after the 1st week of life had higher postoperative complications than repair within the 1st week of life but with insignificant differences ($p > 0.05$), while death was significantly higher among cases repaired after the 1st week of life ($p = 0.019$).

Table (7): Postoperative complications among the study groups (n=35)

	Repair after 1st week of life (n=13)	Repair within the first week of life (n=22)	P-value
Mortality	5(38.5%)	0(0%)	**0.019*[1]**
Pneumothorax	2(15.3%)	1(4.5%)	0.277[1]
Stricture	3(23.1%)	2(9.1%)	0.099[1]
Major anastomotic leakage	2(15.4%)	0(0%)	0.064[2]
Minor Anastomotic Leakage	7(53.8%)	3(13.6%)	0.271[2]

DISCUSSION

In our investigation, we demonstrated that the cases had a mean gestational age of 30.1 ± 4.9 weeks, ranging from 26 to 40 weeks. More than half of them (51.4%) were males, and 48.6% were females. More than half of them (51.4%) delivered vaginally and 48.6% by CS. In our study, pneumothorax was suspected in 3 (10%) cases, strictures were identified in 5 (16.7%) cases, major anastomotic leakage was found in two (6.7%) cases, and 10 cases (33.3%) had minor anastomotic leakage.

In the majority of investigations, the survival rate for cases with LGEA following delayed 1ry anastomosis is reported to exceed ninety percent. Early complications following delayed 1ry anastomosis include leaks, which can happen in up to thirty percent of cases. Fortunately, the majority of anastomotic leaks are minor and will resolve spontaneously with antibiotic medication, nil by mouth, and total parenteral nutrition, obviating the necessity for operative intervention. Nevertheless, certain researchers have stated that there has been significant disruption and failure of conservative treatment, necessitating drainage or reoperation in as many as fifteen percent of their LGEA cases [(5)].

Anastomotic strictures occurred in certain series in as many as sixty percent of cases. A prior anastomotic leak is identified as the most significant risk factor for subsequent stricture formation. The majority of esophageal strictures respond to regular dilatations, although only a small fraction of cases can necessitate resection and reanastomosis. By meticulously managing the esophageal ends, preserving the blood supply, and carefully incorporating the mucosa in every stitch of the anastomosis, strictures may be minimized [(2)].

In the present investigation, twenty-eight cases (93.3%) of the currently living are flourishing with normal oral intake. In total, GERD has been identified in 4 (13.3%) cases following EA/TEF repair, all of which demonstrated reduced or absent peristalsis in the distal esophageal segment, alongside decreased lower esophageal sphincter pressure, leading to an elevated incidence of acidic reflux episodes due to the pressure equilibrium between the intraesophageal and intragastric environments.

Our findings corroborate earlier research indicating that in spite of precise esophageal reconstruction and excellent postoperative care, both early and late complications commonly arise and require management [(6-8)]. The lack of standardized definitions for complications makes comparisons across studies difficult, resulting in highly variable incidence rates [(9)].

Complications following EA/TEF repair are likely multifactorial, depending on peri-/postoperative care, operative methods, and individual case factors [10].

Early complications in our study included minor (6.3%) or major (5.2%) anastomotic leaks, recurrent TEF with (1.0%) or without (9.4%) anastomotic stricture, and refractory strictures with (1.0%) or without (2.1%) fistula. Severe anastomotic leakage, a potentially fatal complication, requires immediate surgical intervention to prevent inflammation and necrosis [11, 12].

Most cases with long-gap EA (LGEA) who undergo delayed 1ry anastomosis can eat normally without dysphagia, with swallowing difficulties being rare and often linked to GER or reflux-associated strictures [13]. Recurrent aspiration pneumonia is uncommon but has been reported, and health-associated quality of life is significantly better in kids with delayed 1ry anastomosis compared to other reconstructive methods [14]. Failure to attain satisfactory delayed 1ry anastomosis, necessitating esophageal replacement, is rare and occurs in only a few LGEA cases. Delayed 1ry anastomosis generally yields better long-term outcomes, with most cases showing normal growth and development, though the risk of Barrett's metaplasia and other complications underscores the necessity for long-term monitoring [13]. Anastomotic leakage is a significant risk factor for recurrent tracheoesophageal fistula and formation of stricture, with 75% of cases in our series developing strictures post-leak, which is consistent with findings of **Koivusalo *et al*.** [12]. Infants with a history of anastomotic leakage and nonspecific symptoms such as recurrent respiratory issues or feeding difficulties must be subjected to contrast investigations and esophago-/tracheobronchoscopy with methylene blue tests [15].

Endoscopic tissue adhesives were less effective for recurrent TEF, while open surgical approaches with vascularized tissue interposition showed better success, even in severe cases [11]. Recurrent distal tracheoesophageal fistula must be distinguished from rare, formerly unidentified proximal fistulas [16]. Anastomotic strictures, a common complication, are influenced by tension, previous leaks, and recurrent GERD [17].

Long-gap EA treatment is challenging with nearly every case experiencing postoperative difficulties [18]. Gastroesophageal issues like dysphagia, impaired peristalsis, and GERD frequently persist into adulthood, with some cases at increased risk of esophageal malignancy, warranting regular endoscopy and impedance/pH follow-up [9]. However, a 12-year follow-up study found no dysplasia or cancer, suggesting limited benefit of routine endoscopic surveillance in childhood but its importance from adolescence onward [18].

CONCLUSION

The timing of esophageal atresia and tracheoesophageal fistula repair is crucial for optimal outcomes and minimizing complications. Early repair reduces complications and shortens recovery time, while delayed repair allows for better stabilization but increases risks. However, many cases are misdiagnosed at birth due to inadequate prenatal detection, increasing the risk of complications. A study aimed to describe the outcome of repair in cases that underwent repair TEF. The overall survival was 30/35, with 5 of the 30 cases dying. Postoperative complications were higher in delayed repair cases.

RECOMMENDATION

EA/TEF surgery generally yields favorable outcomes, with most cases resuming normal diets and low mortality rates. Postoperative complications, such as sepsis, coagulopathy, and chest infections, impact long-term esophageal function. Primary repair is recommended to preserve the esophagus and prevent esophageal replacement surgeries. Early identification and management of complications are crucial. Raising awareness and recommending multidisciplinary follow-up are recommended. Larger longitudinal studies are needed for further validation.

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REFERENCES

1. **Alzaiem M (2020):** Esophageal elongation using Foley catheter in long-gap esophageal atresia. Journal of Neonatal Surgery, 9: 26. https://doi.org/10.47338/jns.v9.555.
2. **Baird R, Puligandla P (2023):** Delayed Presentation of Esophageal Atresia and its Impact on Surgical Timing. Pediatric Surgery International, 39 (4): 1215-1222.doi: 10.21037/tp-23-453
3. **Fischer J, Jaksic T (2023):** Timing and Outcomes of Esophageal Atresia Repair: A Contemporary Review. Journal of Pediatric Surgery, 58 (4): 787-797.
4. **van der Zee D, Bagolan P, Faure C *et al*. (2017):** Position Paper of INoEA Working Group on Long-Gap Esophageal Atresia: For Better Care. Frontiers in pediatrics, 5: 63. https://doi.org/10.3389/fped.2017.00063
5. **Puri P, Friedmacher F (2017):** Delayed primary anastomosis for management of long-gap esophageal atresia: A meta-analysis of complications and long-term outcome. Pediatric Surgery International, 33 (4): 415–421.
6. **Ijsselstijn H, van Beelen N, Wijnen R (2013):** Esophageal atresia: long-term morbidities in adolescence and adulthood. Diseases of the esophagus: official journal of the International Society for Diseases of the Esophagus, 26 (4): 417–421.

7. **Rintala R, Pakarinen M (2013):** Long-term outcome of esophageal anastomosis. European journal of pediatric surgery: official journal of Austrian Association of Pediatric Surgery ... [et al] = Zeitschrift fur Kinderchirurgie, 23 (3): 219–225. https://doi.org/10.1055/s-0033-1347912
8. **Schneider A, Blanc S, Bonnard A *et al*. (2014):** Results from the French National Esophageal Atresia register: one-year outcome. Orphanet journal of rare diseases, 9: 206. https://doi.org/10.1186/s13023-014-0206-5.
9. **Rigueros S L, Connor M, Jones K *et al*. (2016):** Prevalence of Active Long-term Problems in Cases With Anorectal Malformations: A Systematic Review. Diseases of the colon and rectum, 59 (6): 570–580.
10. **Holcomb G, Rothenberg S (2021):** Early vs. Delayed Repair of Esophageal Atresia: A Multicenter Review. European Journal of Pediatric Surgery, 31 (3): 266-273.
11. **Friedmacher F, Kroneis B, Huber-Zeyringer A *et al*. (2017):** Postoperative Complications and Functional Outcome after Esophageal Atresia Repair: Results from Longitudinal Single-Center Follow-Up. Journal of gastrointestinal surgery: official journal of the Society for Surgery of the Alimentary Tract, 21 (6): 927–935.
12. **Koivusalo A, Pakarinen M, Lindahl H *et al*. (2015):** Revisional surgery for recurrent tracheoesophageal fistula and anastomotic complications after repair of esophageal atresia in 258 infants. Journal of pediatric surgery, 50 (2): 250–254.
13. **Friedmacher F (2022):** Delayed primary anastomosis for repair of long-gap esophageal atresia: technique revisited. Pediatric surgery international, 39 (1): 40. https://doi.org/10.1007/s00383-022-05317-6.
14. **Guner Y, Kharbanda A (2022):** Management of Long-Gap Esophageal Atresia: Timing of Surgery and Outcomes. Seminars in Pediatric Surgery, 31 (4): 102321. https://doi.org/10.21037/tp-23-453.
15. **Zani A, Dingemann C, Eaton S *et al*. (2014):** Epidemiology, management, and outcomes of esophageal atresia and tracheoesophageal fistula: A multi-center, population-based study. Annals of Surgery, 260 (5): 870-876.
16. **Zhu H, Shen C, Xiao X *et al*. (2015):** Reoperation for anastomotic complications of esophageal atresia and tracheoesophageal fistula. Journal of pediatric surgery, 50 (12): 2012–2015.
17. **Martinez A, Gonzalez R, Lopez M *et al*. (2024):** Embryonic Developmental Defects in Esophageal Atresia and Tracheoesophageal Fistula. https://doi.org/10.1016/j.ydbio.2021.05.015.
18. **Koivusalo A, Pakarinen M, Lindahl H, Rintala R (2016):** Endoscopic Surveillance After Repair of Oesophageal Atresia: Longitudinal Study in 209 Cases. Journal of pediatric gastroenterology and nutrition, 62 (4): 562–566.