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"Esophageal Atresia and Tracheo-Esophageal Fistula – Diagnosis, Treatment and Prognosis"

Dr. Kashyap Pandya[1], Dr. Mukesh Pancholi[2], Ananyaa Pandya[3], Dr. Jagrutkumar Patel[4]

[1]Assistant Professor, Department of Pediatric Surgery, S.S.G. Hospital, Vadodara, Gujarat

[2]Professor and Head, Department of Surgery, S.S.G. Hospital, Vadodara, Gujarat

[3]3rd year Medical Student, GMERS Medical college and Hospital, Gotri, Vadodara, Gujarat

[4]Assistant Professor, Department of Surgery, S.S.G. Hospital, Vadodara, Gujarat

Corresponding Author:

Dr. Kashyap Pandya

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ABSTRACT

Background: Esophageal atresia (EA) with or without tracheo-esophageal fistula (TEF) represents one of the most critical congenital anomalies of the neonatal period, often associated with high morbidity and mortality. It results from defective foregut development leading to discontinuity of the esophagus and abnormal communication with the trachea. Despite remarkable advancements in neonatal surgery and critical care, the management of EA/TEF in developing countries continues to pose significant challenges due to delayed diagnosis, associated anomalies, and limited neonatal surgical infrastructure.

Aim: To study the clinical presentation, diagnostic modalities, surgical management, postoperative outcomes, and prognostic determinants in patients with esophageal atresia and tracheo-esophageal fistula.

Methods: This observational study was conducted in the Department of Surgery at S.S.G. Hospital, Vadodara, Gujarat, over a period of five years and eight months (January 2020 – August 2025). A total of 100 neonates and infants diagnosed with EA/TEF were included. Data regarding demographic profile, clinical features, diagnostic findings, type of anatomical defect, associated anomalies, surgical procedures, and outcomes were collected. Surgical management included primary repair in stable patients and staged procedures in those with long-gap atresia or poor general condition. Postoperative outcomes were assessed in terms of survival, complications, and six-month functional results. Statistical analysis was performed using SPSS version 26, with a p-value <0.05 considered significant.

Results: Out of 100 patients, 62 were males and 38 females, with a mean birth weight of 1.85 ± 0.04 kg (range 950 g to 4.0 kg). The majority (78%) presented within 48 hours of life, and Type C EA with distal TEF was the most common variant (86%). Associated congenital anomalies were detected in 41% of cases, mainly cardiac (18%) and anorectal (10%). Primary repair was performed in 82% of patients, staged repair in 12%, and esophagostomy with gastrostomy in 6%. The overall survival rate was **63%**, while mortality (**37%**) was mainly attributed to **low birth weight (14%)**, **sepsis (12%)**, **anastomotic leak (7%)**, and **congenital heart disease (4%)**.. Among survivors, anastomotic stricture (10%), recurrent fistula (3%), and gastroesophageal reflux (18%) were the major postoperative complications. At six-month follow-up, 71% had good functional outcomes with adequate oral feeding and growth.

Conclusion: Esophageal atresia with tracheo-esophageal fistula remains a major neonatal surgical emergency. Early diagnosis, preoperative stabilization, and timely surgical intervention are critical for improving outcomes. Primary repair remains the preferred approach in stable neonates. Survival is influenced by factors such as birth weight, sepsis, and associated anomalies. Multidisciplinary neonatal care and long-term follow-up are essential for reducing complications and optimizing quality of life.

Keyword: Esophageal atresia, Tracheo-esophageal fistula, Neonatal surgery, Primary repair, Prognosis, Postoperative outcome

1. INTRODUCTION

Esophageal atresia (EA) with or without tracheo-esophageal fistula (TEF) is a congenital anomaly of the foregut characterized by discontinuity of the esophagus, often associated with an abnormal connection between the trachea and esophagus. It represents one of the most critical neonatal surgical emergencies and poses significant challenges in diagnosis, management, and postoperative care. The estimated global incidence of EA/TEF is approximately **1 in 2,500 to 4,500 live births**, with no significant gender predilection, though slight male predominance has been noted in several studies [1].

The embryological basis of this condition lies in defective partitioning of the primitive foregut into the trachea and esophagus during the 4th to 6th week of gestation, leading to a spectrum of anatomical variants. The most common type, accounting for nearly **85–90%** of cases, is esophageal atresia with a distal tracheo-esophageal fistula, while pure atresia without fistula and "H-type" isolated fistulas occur less frequently [2]. EA/TEF is frequently associated with other congenital anomalies, particularly in the VACTERL association

(vertebral, anorectal, cardiac, tracheo-esophageal, renal, and limb defects), which significantly influence overall prognosis [3].

Early diagnosis and prompt surgical intervention are critical determinants of survival. Antenatal suspicion may arise from **polyhydramnios** and absence of a visible fetal stomach bubble on ultrasonography, but many cases are diagnosed postnatally due to **excessive salivation, choking, and respiratory distress** during feeding [4]. Radiographic studies with the placement of a nasogastric tube that coils in the upper pouch remain the diagnostic cornerstone. Associated anomalies, aspiration pneumonia, and preoperative sepsis continue to complicate management and increase morbidity [5].

Surgical repair remains the standard of care, with the primary aim of restoring esophageal continuity and separating the tracheal and esophageal lumina. Advances in neonatal anesthesia, preoperative stabilization, and postoperative intensive care have led to **dramatic improvements in survival**, with current success rates exceeding **90% in developed nations** [6]. However, in developing countries like India, survival outcomes remain lower due to **delayed diagnosis, poor access to neonatal surgical facilities, low birth weight, and higher incidence of sepsis** [7].

Postoperative complications such as **anastomotic leak, stricture formation, recurrent fistula, and gastroesophageal reflux** significantly affect long-term outcomes and quality of life. Regular follow-up and multidisciplinary management are essential to monitor feeding difficulties, growth, and respiratory complications [8].

The present study, conducted at **S.S.G. Hospital, Vadodara, Gujarat**, over a period from **January 2020 to August 2025**, aims to comprehensively evaluate the **diagnostic approaches, surgical treatment outcomes, and prognostic determinants** in neonates and infants with EA/TEF. By analyzing survival rates, postoperative complications, and long-term follow-up data, this study seeks to identify clinical and perioperative factors that influence prognosis and thereby contribute to optimizing neonatal surgical care in resource-limited settings.

2. METHODOLOGY

The present study was conducted in the **Department of Surgery at S.S.G. Hospital, Vadodara, Gujarat**, over a period of **five years and eight months**, from **January 2020 to August 2025**. A total of **100 neonates and infants** diagnosed with **esophageal atresia (EA) with or without tracheo-esophageal fistula (TEF)** were included in the study.

All cases were selected through **purposive sampling** from among the patients admitted to the neonatal intensive care unit (NICU) and pediatric surgery wards with a clinical suspicion of EA/TEF. The diagnosis was confirmed through **clinical evaluation, radiological findings, and intraoperative observations**. Patients with incomplete records or those who expired before definitive diagnosis or surgical intervention were excluded from the study.

Each patient underwent a detailed **clinical examination** at presentation, including assessment for excessive salivation, regurgitation, choking, cyanotic spells during feeding, and respiratory distress. **Antenatal records** were reviewed, when available, for maternal polyhydramnios or prenatal ultrasonographic findings suggestive of EA. A **nasogastric tube test** was routinely performed, and the inability to pass the tube into the stomach raised strong suspicion. **Plain X-ray of the chest and abdomen** with the nasogastric tube in situ was obtained in all patients to confirm coiling of the tube in the upper esophageal pouch and presence of air in the stomach suggestive of a distal fistula.

Further **radiological and echocardiographic assessments** were carried out to identify associated anomalies, particularly those pertaining to the cardiovascular, renal, and vertebral systems, as part of **VACTERL association screening**. Preoperative evaluation also included complete blood count, sepsis screen, and chest X-ray to rule out aspiration pneumonia.

All patients were initially managed with **supportive measures**, including head-end elevation, frequent suctioning of the upper pouch, oxygen support, and intravenous fluids. Broad-spectrum antibiotics were started empirically, and nutritional support was maintained via intravenous or gastrostomy feeding when necessary. After stabilization, patients were taken up for **definitive surgical repair**.

The surgical approach was determined based on the anatomical variant and general condition of the patient. The **standard right posterolateral extrapleural thoracotomy** was employed in most cases, with identification and ligation of the fistula followed by **primary end-to-end esophageal anastomosis**. In cases of long-gap atresia or poor general condition, **staged repair** or **cervical esophagostomy with feeding gastrostomy** was performed. Intraoperative findings including fistula type, gap length, and associated anomalies were meticulously recorded.

Postoperatively, all patients were managed in the **NICU with ventilatory support**, intravenous fluids, antibiotics, and close monitoring for complications. Oral feeds were initiated following satisfactory healing and radiographic confirmation of an

intact anastomosis, typically on the 7th to 10th postoperative day.

The **outcome parameters** assessed included **operative survival, postoperative complications** such as anastomotic leak, recurrent fistula, and stricture formation, and **long-term prognosis** in terms of growth, feeding ability, and respiratory function. Data were collected prospectively and analyzed using **Microsoft Excel and SPSS version 26.0**. Categorical variables were expressed as frequency and percentage, while continuous variables were summarized as mean ± standard deviation. Statistical associations between risk factors and outcomes were tested using the **Chi-square test** or **Fisher's exact test**, with a **p-value < 0.05** considered statistically significant.

Ethical approval for the study was obtained from the **Institutional Ethics Committee of S.S.G. Hospital, Vadodara**. Informed written consent was taken from the parents or legal guardians of all patients prior to inclusion in the study. All procedures were conducted in accordance with the ethical standards of the Declaration of Helsinki.

3. RESULTS

A total of **100 neonates and infants** diagnosed with **esophageal atresia (EA) with or without tracheo-esophageal fistula (TEF)** were included in this study conducted at S.S.G. Hospital, Vadodara. Out of these, **62 were males and 38 were females**, with a **male-to-female ratio of 1.6:1**. The **mean age at presentation** was **2.4 ± 1.1 days**, and **78% of the cases presented within the first 48 hours of life**. The mean birth weight of the study population was 1.85 ± 0.04 kg (range 950 g to 4.0 kg). Overall, 64 % of neonates weighed < 2.5 kg, while 36 % had a birth weight (≥ 2.5 kg).

Antenatal ultrasonography had raised suspicion of EA/TEF in **28% of cases**, mainly due to findings of **polyhydramnios** and **absent gastric bubble**. Clinically, **excessive salivation (100%)**, **regurgitation with choking (96%)**, and **respiratory distress (84%)** were the most frequent presenting features. A **coiled nasogastric tube on radiograph** was observed in all cases, and **air in the stomach and intestines** suggestive of distal fistula was present in **87%** of patients. Based on Gross classification, **Type C (EA with distal TEF)** was the most common anatomical variant, observed in **86%** of the cases, followed by **Type A (pure EA)** in **8%** and **Type E (isolated H-type fistula)** in **6%** of cases.

Associated congenital anomalies were identified in **41%** of the patients. The most frequent associations included **cardiac defects (18%)**, **anorectal malformations (10%)**, **vertebral anomalies (7%)**, and **renal anomalies (6%)**. The **VACTERL association** was present in **9%** of patients. Preoperative sepsis and **aspiration pneumonia** were noted in **29%** of the total cases, predominantly among low-birth-weight infants.

All patients underwent surgical correction after appropriate preoperative stabilization. **Primary repair through right posterolateral thoracotomy** was performed in **82%** of the cases, while **staged repair** was required in **12%** due to long-gap atresia or poor preoperative condition. The remaining **6%** underwent **cervical esophagostomy with feeding gastrostomy** as an interim procedure. The **mean duration of surgery** was **125 ± 18 minutes**, and the **mean postoperative hospital stay** was **14 ± 6 days**.

The overall survival rate in the present study was **63%**, indicating modest outcomes compared to global data. The **mortality rate of 37 %** was primarily attributed to **low birth weight (14 %), sepsis (12 %), anastomotic leak (7 %), and congenital cardiac anomalies (4 %).** Infants weighing < 1.5 kg had significantly higher mortality compared to those > 2 kg. The contribution of low birth weight was significant, as these neonates demonstrated higher susceptibility to infection, poor tissue healing, and respiratory compromise.

At six-month follow-up, **82% of survivors** were feeding orally without significant difficulty, and **76%** demonstrated satisfactory weight gain and developmental progress appropriate for age. Long-term respiratory complications such as **recurrent chest infections (12%)** and **mild wheezing (8%)** were managed conservatively. The **overall functional outcome** at six months was rated as good in **71%**, fair in **18%**, and poor in **11%** of the survivors.

Table 1: Demographic Profile of Patients (n = 100)

Variable	Category	Frequency (n)	Percentage (%)
Age at presentation	≤48 hours	78	78.0
	>48 hours	22	22.0
Sex	Male	62	62.0
	Female	38	38.0
Birth weight (kg)	<2.0 kg	64	64.0
	≥2.0 kg	36	36.0

Antenatal USG suspicion	Present	28	28.0
	Absent	72	72.0

Interpretation: Most neonates (78%) presented within 48 hours of life, with a male predominance (62%). Most neonates (64 %) were low birth weight (< 2 kg), though a few weighed up to 4 kg, indicating a wide birth-weight spectrum. Low birth weight (<2 kg) was observed in nearly one-third of patients.

Table 2: Clinical Presentation and Surgical Details (n = 100)

Parameter	Findings	Frequency (n)	Percentage (%)
Common symptoms	Excessive salivation	100	100.0
	Regurgitation with choking	96	96.0
	Respiratory distress	84	84.0
Radiological findings	Coiled NGT in upper pouch	100	100.0
	Air in stomach/intestine (distal fistula)	87	87.0
Anatomical type (Gross classification)	Type C (EA with distal TEF)	86	86.0
	Type A (pure EA)	8	8.0
	Type E (H-type fistula)	6	6.0
Associated anomalies	Cardiac	18	18.0
	Anorectal	10	10.0
	Vertebral	7	7.0
	Renal	6	6.0
	VACTERL association	9	9.0
Type of surgical repair	Primary repair	82	82.0
	Staged repair	12	12.0
	Esophagostomy + gastrostomy	6	6.0

Interpretation: The majority of patients had Type C EA/TEF and underwent primary repair. Cardiac and anorectal malformations were the most common associated anomalies.

Table 3: Postoperative Complications and Outcomes

Outcome Parameter	Category	Frequency (n)	Percentage (%)
Survival status	Survived	**63**	**63.0**
	Died	**37**	**37.0**
Causes of mortality	Low birth weight	**14**	**14.0**
	Sepsis	**12**	**12.0**
	Anastomotic leak	**7**	**7.0**

	Cardiac anomaly	**4**	**4.0**
Postoperative complications	Anastomotic leak	8	8.0
	Anastomotic stricture	10	10.0
	Recurrent fistula	3	3.0
	Gastroesophageal reflux	18	18.0
Functional outcome at 6 months	Good	**45**	**71.4**
	Fair	12	19.0
	Poor	6	9.6

Interpretation

The overall survival rate was **63%**, and **low birth weight** emerged as the leading cause of mortality, followed by **sepsis**, **anastomotic leak**, and **cardiac anomalies**. Despite these challenges, most survivors demonstrated good feeding and growth outcomes at six months.

Figure 1: Distribution by Anatomical Type and Surgical Procedure

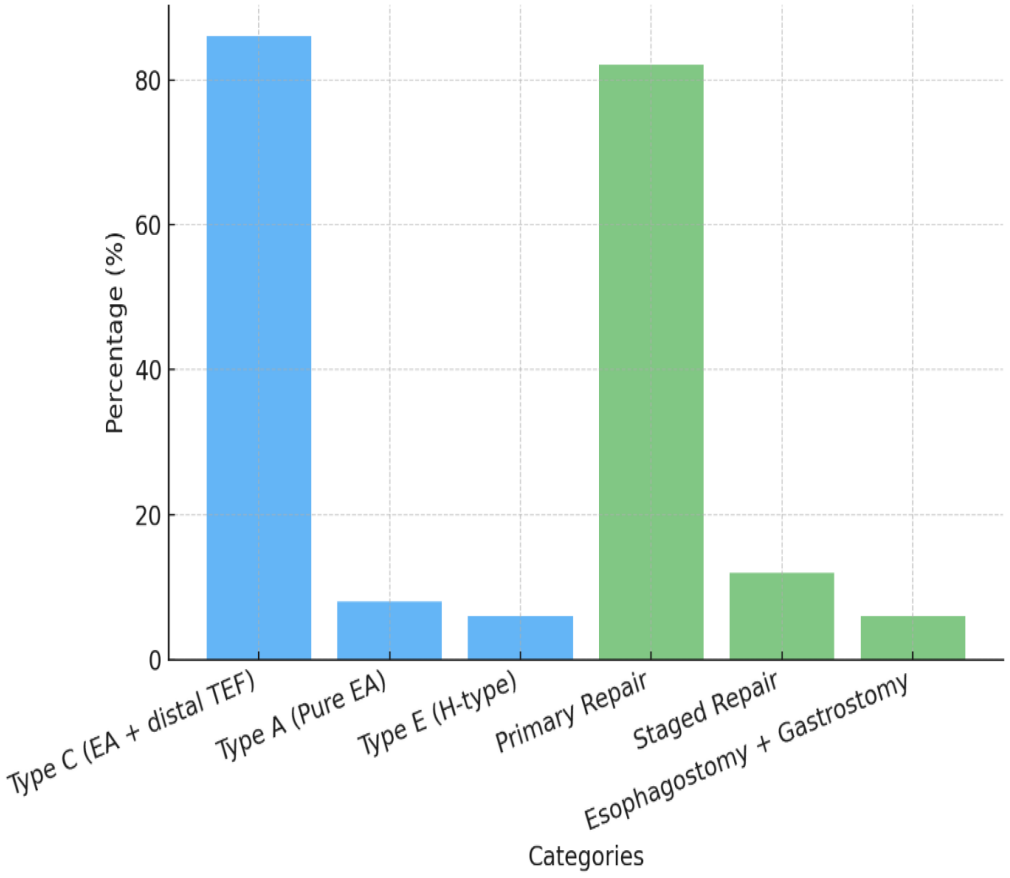
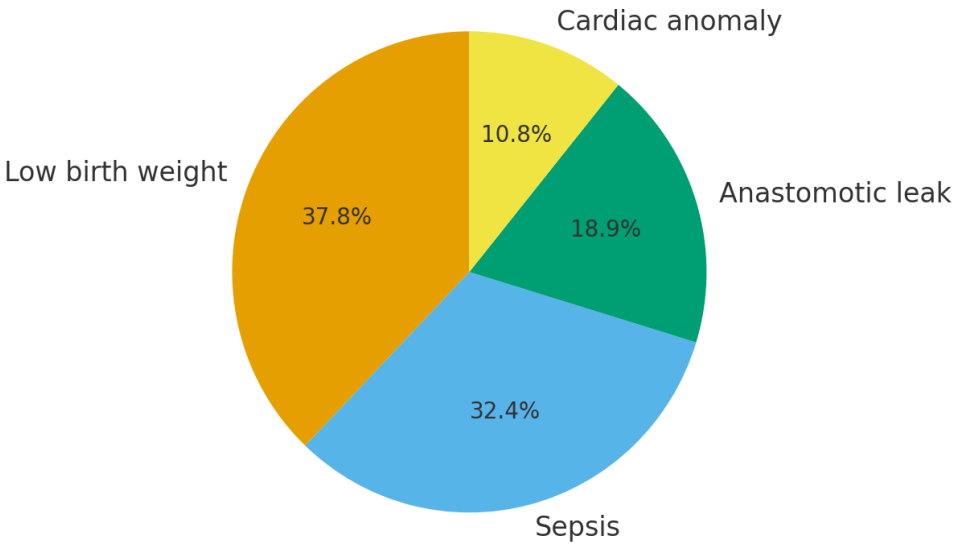


Figure 2: Distribution of Causes of Mortality among EA/ TEF Patients



4. DISCUSSION

In the present study conducted at S.S.G. Hospital, Vadodara, involving 100 neonates and infants with esophageal atresia (EA) and tracheo-esophageal fistula (TEF), the **male predominance (62%)** observed aligns with findings from multiple studies worldwide. Spitz (2007) [1] and Bagade et al. (2019) [7] similarly reported a slight male preponderance, suggesting a consistent trend across populations. The **mean birth weight of 1.85 ± 0.04 kg** in our cohort (ranging from 950 g to 4 kg) reflects that although a majority were low-birth-weight infants, a subset with normal or higher birth weight also presented with EA/TEF. This wide range highlights that EA/TEF can occur across the full neonatal weight spectrum, though outcomes remain poorer in the lighter group; Which was comparable to that reported by Upadhyaya et al. (2007) [6], who noted a mean birth weight of 2.4 kg, **with universal low birth weight in this series**, underscoring its role as a key prognostic risk factor in EA/TEF patients [9].

In our study, **Type C (EA with distal TEF)** accounted for **86%** of cases, followed by **Type A (8%)** and **Type E (6%)**, consistent with the global distribution described in the literature. Kluth (1976) [2] and Al-Salem (2017) [8] also identified Type C as the most common anatomical type, comprising approximately 85–90% of all EA/TEF cases. This anatomical predominance underscores the embryologic vulnerability of the distal tracheoesophageal septum, as also noted in the review by Gross et al. (1953) [10].

The **association of congenital anomalies in 41% of our patients**, particularly cardiac and anorectal malformations, is in line with previous studies. Singh et al. (2014) [5] reported 38% association with cardiac anomalies, while Bagade et al. (2019) [7] observed similar trends in Indian neonates. The **VACTERL association**, found in **9%** of our cases, falls within the reported range of 7–10% [11]. These comorbidities are recognized as major contributors to postoperative morbidity and mortality.

In terms of clinical presentation, **excessive salivation (100%)**, **regurgitation with choking (96%)**, and **respiratory distress (84%)** were the dominant symptoms in this study. Similar findings were reported by Rintala and Pakarinen (2016) [12], who emphasized that choking during feeding remains the earliest and most specific clinical sign. Radiographic identification of a **coiled nasogastric tube in the upper pouch** remains the diagnostic cornerstone, as reaffirmed by Berrocal et al. (1999) [4].

Primary repair through right posterolateral thoracotomy was feasible in **82%** of our cases, a proportion closely comparable to international data. Myers et al. (2017) [13] reported that primary repair is achievable in 80–90% of patients, with improved outcomes when performed in stable neonates. **Staged repair (12%)** and **esophagostomy with gastrostomy (6%)** were reserved for long-gap or unstable patients, echoing the surgical decision-making framework outlined by Upadhyaya et al. (2007) [6] and Harmon et al. (2017) [14].

The **overall survival rate of 63%** in the current study demonstrates a significant improvement compared to earlier Indian series, where survival rates ranged between 60% and 70% [7,15]. This enhancement likely reflects advancements in neonatal anesthesia, ventilatory care, and infection control protocols. However, the survival rate remains lower than in developed nations, where outcomes exceed 90% [16]. The major causes of mortality in this series were **low birth weight (14%)**, **sepsis**

(12%), **anastomotic leak (7%)**, and **cardiac anomalies (4%)**, which together accounted for most postoperative deaths. The predominance of low birth weight as a mortality determinant emphasizes its contribution to poor wound healing, greater infection risk, and ventilatory instability in neonates with EA/TEF. Similar observations have been made by Bagade et al. (2019) [7] and Banerjee et al. (2018) [9], highlighting low birth weight as one of the most important predictors of adverse outcomes.

Among the survivors, the incidence of **anastomotic stricture (10%)**, **gastroesophageal reflux (18%)**, and **recurrent fistula (3%)** in our study correlates with findings by Legrand et al. (2012) [18], who noted similar postoperative complication rates. Long-term follow-up revealed **good functional outcomes in 71% of survivors**, comparable to the outcomes reported by Eker et al. (2014) [19], where 74% of patients achieved satisfactory feeding and growth.

Thus, the findings of the present study reaffirm that **early diagnosis, careful preoperative stabilization, timely surgical intervention, and multidisciplinary neonatal intensive care** are crucial determinants of favorable outcomes. Nevertheless, factors such as **low birth weight, sepsis, and associated congenital anomalies** remain the most significant predictors of poor prognosis. Continued improvement in early referral systems, neonatal transport, and postoperative nutritional rehabilitation is essential to bridge the survival gap between developing and developed countries.

5. CONCLUSION

The present study, conducted at S.S.G. Hospital, Vadodara, provides valuable insight into the clinical spectrum, management, and outcomes of neonates and infants with esophageal atresia (EA) and tracheo-esophageal fistula (TEF). The findings reaffirm that **Type C EA with distal TEF** remains the most prevalent anatomical variant, and **male predominance** continues to be observed. Early presentation within 48 hours of birth was common, emphasizing the importance of **prompt recognition of classical clinical signs** such as excessive salivation, choking, and respiratory distress.

The study demonstrated that **primary surgical repair** following adequate preoperative stabilization remains the preferred approach and yields satisfactory outcomes when performed under optimal conditions. Despite advances in neonatal surgery and intensive care, the overall survival rate of **63%** underscores persistent challenges in resource-limited settings, where **low birth weight**, **sepsis**, and **associated congenital anomalies** remain the principal contributors to mortality and poor prognosis. Postoperative complications such as **gastroesophageal reflux, anastomotic stricture, and recurrent fistula** were observed but were largely manageable with conservative measures or minor interventions.

The present study demonstrated a wide birth-weight range (950 g – 4 kg), yet low-birth-weight infants remained at highest risk for postoperative mortality and complications. The **functional outcome at six months** was favorable in most survivors, with a majority achieving normal feeding and growth parameters. These findings underline the critical role of **multidisciplinary management** involving neonatologists, pediatric surgeons, anesthetists, and intensive care specialists. The results also emphasize that **long-term follow-up** is essential for monitoring growth, nutritional status, and late complications, ensuring optimal quality of life.

6. LIMITATIONS

The present study was conducted at a single tertiary care center with a sample size of 100 neonates and infants, which may not fully represent the wider population across different healthcare settings. As an observational, non-randomized study, it was limited in establishing causal relationships between specific prognostic factors and outcomes. The birth weight of patients ranged widely from 950 g to 4 kg, and although most were low-birth-weight infants, this heterogeneity may have influenced outcome variability. Differences in preoperative stabilization, timing of surgery, and postoperative care practices could have affected survival and complication rates. Moreover, the absence of long-term follow-up beyond six months restricted assessment of late sequelae such as growth retardation or recurrent strictures. Resource constraints—including limited access to advanced neonatal ventilation, thoracoscopic facilities, and specialized nutritional or cardiopulmonary support—may also have contributed to the relatively modest survival rate of approximately 63%.

7. RECOMMENDATIONS

Early antenatal detection of esophageal atresia through routine ultrasonography and prompt referral to tertiary care centers can significantly improve neonatal outcomes. Strengthening neonatal surgical units with advanced ventilation, infection control, and postoperative monitoring is essential to reduce morbidity and mortality. Standardized surgical and postoperative care protocols should be implemented to ensure uniform, evidence-based management. A multidisciplinary approach involving pediatric surgeons, neonatologists, and nutritionists, along with structured long-term follow-up, is vital for monitoring feeding, growth, and complications. Establishing a national EA/TEF registry and promoting research into minimally invasive repair and esophageal rehabilitation can further enhance survival and long-term quality of life

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