

## Clinical Characteristics and Treatment of Esophageal Atresia: A Single Institutional Experience

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### Original Research Article

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**Abstract:** Esophageal atresia (EA) is a congenital anomaly that affects approximately 1 in 2500–4000 live births. Available methods of improving the prenatal diagnostic rate include ultrasound examination of the fetal neck to view the blind-ending upper pouch and to observe fetal swallowing and magnetic resonance imaging. The newborn infant of a mother with polyhydramnios should always have a nasogastric tube passed soon after delivery to exclude esophageal atresia. While the management of EA and TEF has evolved over the last four decades, it continues to remain a challenging problem in specialized pediatric centers and even more so beyond such environments. Patients who had been admitted in Mahatma Gandhi Medical College, Jaipur with a diagnosis of esophageal atresia / tracheoesophageal fistula between September 2013 to March 2018 were enrolled. Patient's data included medical record number (MRN), sex, date of birth, time of presentation, age at surgery (days), birth weight, gestational age, antenatal scan, mode of delivery, intra-operative measurements of gap, associated anomalies, postoperative morbidities and mortality, time to discharge, follow up outdoor visits record. The outcomes measured included time from birth to operation, gap between the two ends, trans-anastomotic tube usage, pleural wrap, anastomotic leak rates, recurrence rates, presence of. Antenatal diagnosis based on antenatal scan was done in 4(16%) cases. 3 in in-born and 1 in out-born patients. Maternal Polyhydramnios was present in 6(24%) patients. The most common associated congenital malformation were cardiac 7(28%) followed by genitourinary 3(12%). Birth weight and cardiac anomalies were the most important prognostic markers. The survival in patients with birth weight more than 1.5 kg with no major cardiac anomalies was 93%. In patients with less than 1.5kg weight or major cardiac anomalies the survival was 77%. While one patient, weighing less than 1.5kg with major cardiac anomalies could not be salvaged. Another important prognostic marker was gap between the two ends of the esophagus, while patients with <2cms gap had subsequent stricture in 29.4 % compared to 50% in patients with >2cms gap. Patients with shorter than 2cms gap had no anastomotic leak compared to 25% leak in patients with >2cms gap. In long term, Tracheomalacia anastomotic stricture and pneumonia were the most common complications-Stricture, respiratory morbidity.

**Keywords:** Esophageal atresia (EA), Stricture, respiratory morbidity, polyhydramnios.

### INTRODUCTION

Esophageal atresia (EA) is a congenital anomaly that affects approximately 1 in 2500–4000 live births [1]. EA is most often observed in conjunction with a tracheoesophageal fistula (TEF), typically between the trachea and distal esophageal pouch (Type-C); other classification subtypes include Type-A (EA without TEF), Type-B (EA with proximal TEF), Type-D (EA with proximal and distal TEF), and Type-E (TEF without EA) [2,3]. EA/TEF is considered a surgical urgency, requiring ligation of the fistula and

anastomosis of the esophagus. Repair is required to avoid aspiration, reflux pneumonitis, gaseous-distension of the stomach, and ultimately to establish a normal feeding mechanism [4].

The diagnosis of esophageal atresia may be suspected prenatally by the finding of a small or absent fetal stomach bubble on ultrasound scan performed after the 18<sup>th</sup> week of gestation. Overall the sensitivity of ultrasonography is 42% but in combination with polyhydramnios the positive predictive value is 56%

[5]. Polyhydramnios alone is a poor indication of esophageal atresia (1% incidence). Available methods of improving the prenatal diagnostic rate include ultrasound examination of the fetal neck to view the blind-ending upper pouch [6] and to observe fetal swallowing and magnetic resonance imaging [7].

The newborn infant of a mother with polyhydramnios should always have a nasogastric tube passed soon after delivery to exclude esophageal atresia. Infants with esophageal atresia are unable to swallow saliva and are noted to have excessive salivation requiring repeated suctioning. At this stage, and certainly before the first feed, a stiff wide-bore (10 French gauge) catheter should be passed through the mouth into the esophagus. In esophageal atresia the catheter will not pass beyond 9–10 cm from the lower alveolar ridge. A plain X-ray of the chest and abdomen will show the tip of the catheter arrested in the superior mediastinum (T 2–4) while gas in the stomach and intestine signifies the presence of a distal trachea-esophageal fistula. The absence of gastrointestinal gas is indicative of an isolated atresia. The X-ray may reveal additional anomalies such as a "double bubble" appearance of duodenal atresia, vertebral or rib abnormalities.

While the management of EA and TEF has evolved over the last four decades, it continues to remain a challenging problem in specialized pediatric centers and even more so beyond such environments. The post-operative course of the disease is frequently associated with serious morbidities. Anastomotic stricture, leak, tracheomalacia, GERD, aspiration pneumonitis are major causes of morbidity and mortality. The risk of leak is related to the gap between the two ends of the esophagus. Birth weight and

associated cardiac anomalies are important prognostic indicators.

**MATERIALS AND METHODS**

Patients who had been admitted in Mahatma Gandhi Medical College, Jaipur with a diagnosis of esophageal atresia / tracheoesophageal fistula between September 2013 to March 2018 were enrolled. This prospective observational study analyzed these patients from the time of admission till discharge and in follow up outdoor visits. Only patients with Gross type C cases i.e. Esophageal Atresia with distal tracheoesophageal fistula were included. All patients were stratified into risk groups based on Spitz criteria.

All cases were operated during this time by the same surgeon. Only congenital type C, EA with TEF was included. All patients were evaluated for associated anomalies by echocardiography, USG KUB. All patients underwent Right posterolateral thoracotomy with extra pleural approach, with ligation of azygous vein; gap measured prior to fistula ligation and upper pouch mobilization, end to end single layer esophageal anastomosis and drain was used in all cases. Tran’s anastomotic tube was used in initial 10 cases and a pleural wrap was used in 12 cases.

Patient’s data included medical record number (MRN), sex, date of birth, time of presentation, age at surgery(days), birth weight, gestational age, antenatal scan, mode of delivery, intra-operative measurements of gap, associated anomalies, postoperative morbidities and mortality ,time to discharge, follow up outdoor visits record. The outcomes measured included time from birth to operation, gap between the two ends, trans-anastomotic tube usage, pleural wrap, anastomotic leak rates, recurrence rates, presence of stricture, respiratory morbidity.

**Table-1: Clinical characteristics of patients**

| S. No. | Characteristic                     | Value              |
|--------|------------------------------------|--------------------|
| 1.     | Cases                              | 25                 |
| 2.     | Sex (M/F)                          | 17/8               |
| 3.     | Gestational age(wk)                |                    |
|        | >37                                | 14(56%)            |
|        | <37                                | 11(44%)            |
| 4.     | Birth weight(kg)                   | 2.04 kg +/- 0.57kg |
|        | >2.5                               | 7                  |
|        | 1.5 – 2.5                          | 13                 |
|        | <1.5                               | 5                  |
| 5.     | Prenatal diagnosis                 | 4 (16%)            |
| 6.     | Maternal polyhydramnios            | 6 (24%)            |
| 7.     | Delivery (vaginal/C-section)       | 12/13              |
| 8.     | Associated congenital malformation |                    |
|        | Cardiac                            | 7 (28%)            |
|        | Vertebral                          | 2 (8%)             |
|        | Limb                               | 1 (4%)             |
|        | Anorectal                          | 0                  |
|        | Genitourinary                      | 3 (12%)            |
| 9.     | Follow up period(mo)               | 54                 |

Values are presented as mean ± SD or number (%)

**Aims and objectives**

- The aim of our study was to statistically analyze
- The percentage of cases that were diagnosed antenatally.
- Association with polyhydramnios.
- Association of birth weight with outcome.

- Association of length of gap between the esophageal ends with outcome.
- Assessment of associated morbidity and mortality.

**OBSERVATIONS AND RESULTS**

**Table-2: Postoperative complications**

| S.No. | Complication         | No. (%)  |
|-------|----------------------|----------|
| 1.    | Esophageal stricture | 9 (36%)  |
| 2.    | Anastomotic leakage  | 2 (8%)   |
| 3.    | Recurrence of TEF    | 0        |
| 4.    | Pneumonia            | 8 (32%)  |
| 5.    | GER                  | 10 (40%) |
| 6.    | Tracheomalacia       | 11 (44%) |
| 7.    | Subglottic stenosis  | 1 (4%)   |
| 8.    | Mortality            | 4 (16%)  |
|       | Total                |          |

**Table-3: Relationship between esophageal stricture and anastomotic leak with Esophageal gap**

| Gap    | Number | Stricture | Anastomotic leak |
|--------|--------|-----------|------------------|
| <2cms  | 17     | 5         | -                |
| 2-3cms | 7      | 4         | 1                |
| >3cms  | 1      | -         | 1                |

| Gap     | Stricture/ Anastomotic leak |    | Chi square (df) | P value |
|---------|-----------------------------|----|-----------------|---------|
|         | Yes                         | No |                 |         |
| < 2 cms | 5 (29.4%)                   | 12 | 4.59 (1)        | 0.032   |
| ≥ 2 cms | 6 (75%)                     | 2  |                 |         |

There is a statistically significant association of gap between the esophageal ends and Stricture/Anastomotic leak. 29.41% patients with gap <

2 cms developed stricture/ leak while 75% patients with gap ≥ 2 cms developed stricture/ leak.

**Table-4: Risk group according to Spitz criteria and survival**

| Group | Birth characteristics                        | Survival    |
|-------|--|-------------|
| I     | Wt>1.5kg<br>Without<br>Major cardiac anomaly | 93% (14/15) |
| II    | Wt <1.5kg<br>Or<br>Major cardiac anomaly     | 77% (7/9)   |
| III   | Wt <1.5kg<br>With<br>Major cardiac anomaly   | 0% (0/1)    |

| Group  |   | Survival   |    | P value |
|--------|---|------------|----|---------|
|        |   | Yes        | No |         |
| I      | Wt>1.5kg<br>Without<br>Major cardiac anomaly        | 14 (93.33) | 1  | 0.119   |
| II+III | Wt <1.5kg<br>Or<br>Major cardiac anomaly<br>Or Both | 7 (70)     | 3  |         |

Though 93.33% patients of Cat I survived while only 70% patients with Cat2,3 survived but on applying chi square, the difference was not statistically significant.

#### **DISCUSSION AND CONCLUSION**

In this study, patients of EA with TEF, Gross type C, admitted in between September 2013 to March 2018 were included. A total of 25 patients were studied. Both inborn and referred patients were included, 17(68%) were male and 8(32%) patients were female. M: F ratio being 2.1:1. Antenatal diagnosis based on antenatal scan was done in 4(16%) cases. 3 in in-born and 1 in out-born patients. Maternal Polyhydramnios was present in 6(24%) patients. The most common associated congenital malformation were cardiac 7(28%) followed by genitourinary 3(12%). Birth weight and cardiac anomalies were the most important prognostic markers. The survival in patients with birth weight more than 1.5 kg with no major cardiac anomalies was 93%. In patients with less than 1.5kg weight or major cardiac anomalies the survival was 77%. While one patient, weighing less than 1.5kg with major cardiac anomalies could not be salvaged. Another important prognostic marker was gap between the two ends of the esophagus, while patients with <2cms gap had subsequent stricture in 29.4 % compared to 50% in patients with >2cms gap. Patients with shorter than 2cms gap had no anastomotic leak compared to 25% leak in patients with >2cms gap. In long term, Tracheomalacia anastomotic stricture and pneumonia were the most common complications.

To decrease the mortality rate in EA, several prognostic classification systems, have been developed to guide diagnostic and treatment strategies. However, most of these systems only focus on the influence of major anomalies, such as cardiac malformation. External risk factors, such as sepsis and respiratory failure that could be influenced by the NICU care to a certain extent are not included. Undoubtedly, any efforts to reduce the incidence of the above-mentioned external risk factors may actually decrease the mortality rate in EA.

A low birth weight, anastomotic leak, respiratory failure, and postoperative sepsis were independent risk factors for poor outcomes. Any efforts to reduce these risk factors may reduce the mortality rate in EA.

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