MORTALITY RATES AND RISK FACTORS IN NEONATES WITH ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL ATRESIA: A SINGLE CENTER ANALYSIS

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INTRODUCTION

Esophageal atresia with tracheoesophageal fistula (EA/TEF) is a common birth defect that occurs in around 1 in 2500 live births(1) It is often associated with other congenital anomalies, including VACTERL syndrome, which can lead to significant morbidity and mortality in affected infants. Early diagnosis and surgical intervention are critical to minimize adverse outcomes, and most infants with EA/TEF undergo early surgical repair. However, cases with a long gap between the two ends of the esophagus pose a significant challenge for surgical repair(2).

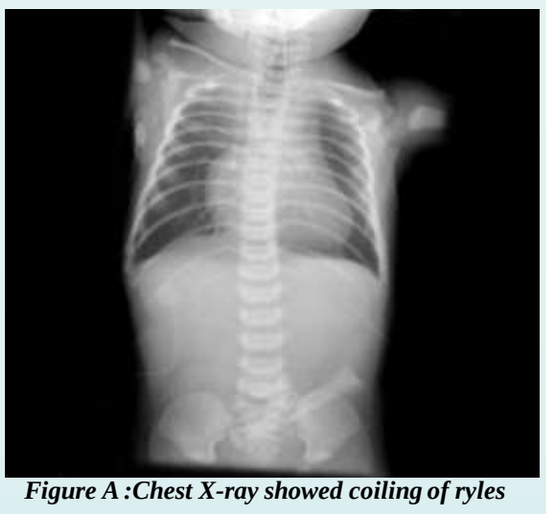

Figure A :Chest X-ray showed coiling of ryles tube in upper esophagus pouch

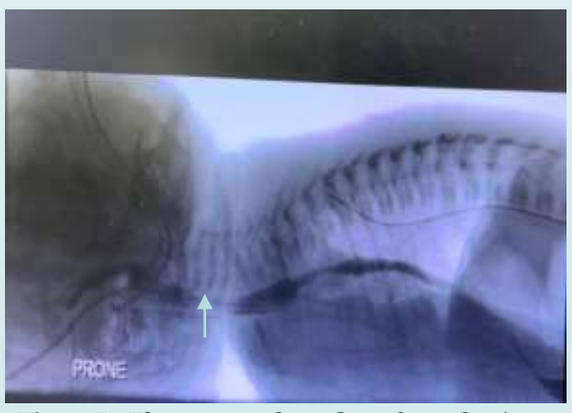

Figure B :Fluoroscopy showed esophageal stricture

MATERIALS AND METHODS

The study was conducted at Hospital Raja Perempuan Zainab II (HRPZII) using prospectively collected data from the past six years (2016-2021) to evaluate the management and outcomes of neonates diagnosed with EA/TEF. Diagnosis was based on the inability to pass a nasogastric tube into the stomach, with the tube coiling in the upper esophageal pouch as shown on chest X-ray. The work-up included echocardiogram prior thoracotomy, and primary anastomosis. In cases with a long gap between the ends, ligation of the fistula and gastrostomy were performed with late second procedure. The collected data included pure esophageal atresia, esophageal atresia with distal fistula, and long gap esophageal atresia, associated congenital anomalies, timing of repair, and complications post-operatively. The primary outcome analyzed was mortality in HRPZII, and the study also evaluated the different types of EA/TEF and the risk factors that affected them.

RESULTS

This study included 21 patients who underwent surgery, consisting of 11 boys (52.4%) and 10 girls (47.6%). The mean birth weight of the patients was 2.4 kg, ranging from 1.1 kg to 3.9 kg. Of the patients, 23.8% were born prematurely with a gestational age of less than 37 weeks, and 9.5% had a birth weight of less than 1.5 kg. The mean of gestational age was 37 week. Only 2 patients (9.5%) had no other congenital malformations, while the remaining 90.4% had one or more concomitant congenital

Malformation	No	Percentage %
Vertebral	4	19.05%
Anorectal	2	9.52%
Cardiac	16	76.19%
Renal	3	14.29%
Duodenum	1	4.76%
Limbs	3	14.29%
others	3	14.29%

Table 1 Congenital malformations associated with esophageal atresia

All 21 patients underwent surgery, but unfortunately, 3 patients died after gastrotomy before the second stage operation could be performed. Table 2 shows distribution of the types of atresia.

Type of Esophageal Atresia	No	%
Pure Esophageal Atresia	1	4.76%
Long Gap	2	9.52%
EA with distal TEF	18	85.71%

Table 2 Types of esophageal atresia in 21 patients

Complication	No	%
Wound infection	1	4.76%
Anastomotic leak	2	9.52%
Gastroesophageal reflux (GER)	3	14.29%
Radiological stricture	3	14.29%
Empyema	3	14 %
Subscapular liver hematoma	1	4.76%

Table 3 Complication postoperatively

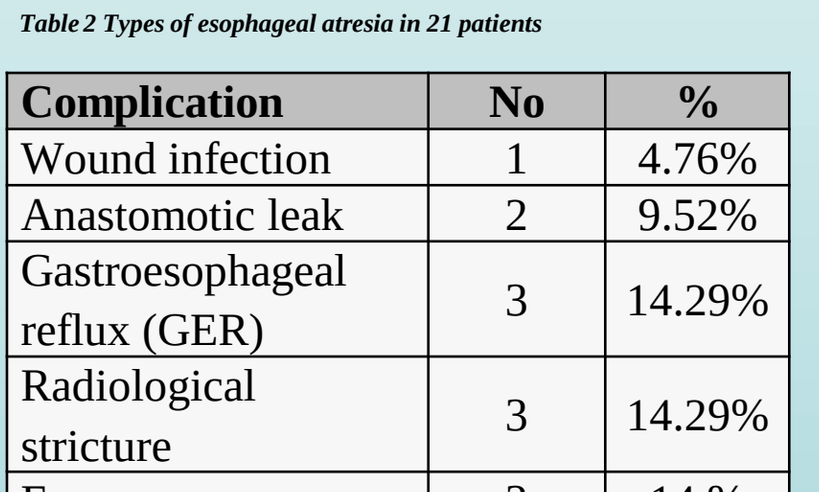

Diagram A showed outcome of mortality and causes of death

The study involved 21 patients who underwent esophageal anastomosis within the first five days of birth, and 1 patient who underwent gastrostomy on day 8 of life.

The complication post operatively are showed as per Table 3 and was diagnosed based on clinical symptoms and radiological imaging. All the complications were treated non-operatively with continuous observation and started on proton pump inhibitor for reflux, dilatation for stricture and transanastomotic feeding for leak.

The number of deaths in the study group was 7 out of 21 patients that was operated, which gives overall mortality rate of 33%. The causes of death were sepsis (3 patients), cardiac causes (1 patients), extreme prematurity with LVH grade 4 (1 patients) and unknown (2 patients) due to missing data. This is due to retrospective data collection with referral to another centre post operatively.

Mortality was found to be more common in patients with congenital malformations (p=0.047), but there was no significant association between mortality and low birth weight(0.169) or prematurity (0.325).

DISCUSSION

1. Our study showed mortality rate of 33%, higher than the US nationwide average of 9% likely due to underestimation of the occurence of EA/TEF in rural areas with notable inconsistency of detection rates ranging 10-50% in rural areas(3). While a separate study at Hospital Sultanah Bahiyyah reported a mortality rate of 23% , which is lower compared to our study likely due to lower percentage of associated congenital anomalies in their study.(4).
2. Cardiac anomalies remains as the most significant risk factors for mortality, consistent with other studies and birth weight did not significantly contribute to mortality, which similar finding can be seen with a study by Calisti et al. In their regression statistical data, it was confirmed that major cardiac anomalies were stronger predictor compared to birth weight according to Montreal criteria.(5)
3. Anastomotic leak had occurred in 9.52% of cases, detected when upper contrast study performed 1 week postoperatively. None of the patients in our cohort required surgical intervention following the discovery of the anastomotic leak. One of the patients, however, developed anastomotic stricture. There was no significant association (p=0.514) between anastomotic leak and mortality rate. The incidence of anastomotic leaks (9.5%) in our studies is lower compared to Indian series leak rate (25%) and has been decreased over time by preventing anastomotic tension during surgery to reduce the risk of anastomotic leak(6).
4. Radiological anastomotic strictures accounted for 14.29% of cases in our study, however only 1 patient required post-operative esophageal dilatation. Reported incidence of stricture post repair ranges from 32 to 59% in the majority of recent studies, which shows that careful handling of the oesophageal ends, preservation of the blood supply, and thorough mucosal inclusion in each suture of the anastomosis can reduce the risk of strictures (1).
5. Post-operative empyema was seen in 1 case (9.52%) which required a long course of antibiotics predisposing patient to nosocominal infection.
6. The discrepancy of the mortality rates may contributed by small sample size and retrospective data. A multicentre study should be done to get better views in pediatric care in Malaysia.(4)

CONCLUSION

The study found that mortality rates in our hospital were higher than international studies and studies in Malaysia and were strongly linked to the presence of accompanying congenital anomalies, emphasizing the need to identify and manage such anomalies to reduce mortality rates in neonates with EA/TEF, with type C being the most common type and long gap TEF having a higher mortality rate.

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