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Leecarlo Millano Lumban Gaol
Department of Surgery, Pediatric Surgery Division, Tarakan General Hospital, Jl. Kyai Caringin No.7, RT.11/RW.4, Cideng, Kecamatan Gambir, Kota Jakarta Pusat, Daerah Khusus Ibukota Jakarta, Indonesia

Yohanes Firmansyah
Department of Surgery, Pediatric Surgery Division, Tarakan General Hospital, Jl. Kyai Caringin No.7, RT.11/RW.4, Cideng, Kecamatan Gambir, Kota Jakarta Pusat, Daerah Khusus Ibukota Jakarta, Indonesia

Melian Anita
Department of Surgery, Pediatric Surgery Division, Tarakan General Hospital, Jl. Kyai Caringin No.7, RT.11/RW.4, Cideng, Kecamatan Gambir, Kota Jakarta Pusat, Daerah Khusus Ibukota Jakarta, Indonesia

Corresponding Author:
Yohanes Firmansyah
Department of Surgery, Pediatric Surgery Division, Tarakan General Hospital, Jl. Kyai Caringin No.7, RT.11/RW.4, Cideng, Kecamatan Gambir, Kota Jakarta Pusat, Daerah Khusus Ibukota Jakarta, Indonesia

Prevalence of esophageal atresia as well as Mortality Outcomes based on Prognostic Criteria at Tarakan General Hospital from 2015 To 2021

Leecarlo Millano Lumban Gaol, Yohanes Firmansyah and Melian Anita

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Abstract
Esophageal atresia (EA) is a disorder that often occurs in infants with an incidence in the world ranging from 0.4 - 3.6 per 10,000 births. The survival rate in developed countries ranges from 85 to 95%. The causes of EA mortality in infants vary widely. The purpose of this study was to see the risk factors for the incidence of EA mortality in infants and the prevalence of EA from all operations in the pediatric surgery division at Tarakan Hospital. Method and Material: Cross sectional study with research samples in the form of EA in infant from April 10, 2021 to April 15, 2021 at the Department of Surgery, Pediatric Surgery Division, Tarakan General Hospital from 2015 to 2021. The study was analyzed using descriptive and analytical statistical analysis. Results: The incidence of esophageal atresia was 1.51% from 2015 to 2021. The demographics of the participants were 16 (51.6%), the average age was 8.00 (2.00 - 19.00) months, the type of atresia esophagus type B was 28 (90.3%) and Waterston C was 9 (29.0%). The mortality rate was detected with sepsis, perforation and aspiration in 16 (51, 6 percent) of respondents. In bivariate analysis, the presence of co-morbidities such as sepsis, leaks, and perforation was found to have contributed significantly to death (p-value 0.001), and the mean age difference between death and survival groups was significant.

Keywords: esophageal atresia; mortality; outcome; sepsis; prognostic

Introduction
Esophageal atresia is a congenital disorder characterized by not connecting the proximal esophagus with the distal esophagus. Esophageal atresia can occur with tracheoesophageal fistula, which is a congenital abnormality in which there is an abnormal connection between the esophagus and the trachea [1–3].

Esophageal atresia is a fairly frequent congenital negligence with an average incidence of about 1 in 2500 to 3000 live births. Incidence of Esophageal atresia in the United States is 1 case per 3000 live births. Worldwide, the incidence varies from 0.4 - 3.6 per 10,000 live births. The highest incidence is in Finland, which is 1 case in 2500 live births [4].

Until now, the teratogen causing this disorder is still unknown [5]. There have been reports linking esophageal atresia in families. There is a 2% risk if you have this disorder. This disorder is also associated with trisomy 21, 13 and 18 [6–8]. The incidence of twins is 6 X more than non-twins. Today, many believe that the development of esophageal atresia is not genetically related. The debate over this embryopathological process continues, but little progress has been made. His old theory held that lateral in folding divides the foregut into the esophagus and trachea, but findings in the field of human embryology do not support this theory [9].

In 1984, O'Rahily stated that there is a fix cephalad point of the tracheoesophageal separation, with elements from the tracheobronchial and esophageal extending caudally. This theory is less suitable for esophageal atresia, but describes TEF as deficiency or failure of the esophageal mucosa, as linear growth of organs in cellular division of the esophageal epithelium. In 1987, Kluth stated that tracheoesophageal septals played an important role in the development of esophageal atresia. Based on the embryopathological process in development even though it is still in its early stages, there has been differentiation between the trachea and the esophagus, where the distance between them is too close so that there is no separation. He also stated that vascular disorders can also play a role in the occurrence of esophageal aresia or fistula.

In 2001 Oxford and others stated that ectopic ventral misposition of the notochord in embryos aged 21 days of gestation can cause gene locus disorders, apoptotic disorders of the foregut and other types of esophageal atresia. This condition can occur due to variations in the influence of teratogens during early gestation such as twins, exposure to toxins, or the possibility of abortion [10–14].

Neonates who undergo surgical correction generally experience different degrees of esophageal dysmotility. The extent of correction affects the severity of complications that arise. Often a stricture occurs at the site of anastomosis which eventually requires dilation. Serial esophagography should be performed at 2 months, 6 months and 1 year of age or if swallowing is difficult. Recurrence of tracheoesophageal fistulas has been reported and requires re-correction. Recurrence occurs most often at the site of the primary anastomosis. Tissue damage due to poor vascularization of the distal esophagus and dissection too close to the trachea are risk factors for fistula recurrence [15-16].

Nearly half of patients who undergo surgical correction of esophageal atresia then experience gastroesophageal reflux (GERD). And half of those with GERD responded well to prokinetic agents, histamine H2 receptor blockers, or both, and the other half required surgical correction. Patients with prolonged GERD will experience changes in the esophageal mucosa such as esophagitis and gastric metapopulation (Barrett's esophagus) [17–20]. Adenocarcinoma of the stomach originating and gastric metapopulation was reported in a patient with esophageal atresia 20 years after neonatal correction. For this reason, it is recommended to follow up with long-term endoscopy [21–23]. In 1962, Waterson *et al* classified babies born with esophageal atresia into 3 groups "with different life expectancies". Classification according to birth weight and other related disorders: According to Birth Weight: Group A:> 2500 gr and good; Group B: Born 1800-2500 gr and good OR Birth weight> high, moderate pneumonia and congenital abnormalities; Group C: High birth weight & severe pneumonia and severe congenital abnormalities [24].

It is clear that a new risk-based classification system is needed in the modern era. The new, risk-based classification includes birth weight and heart malfomations which are responsible for the majority of deaths.

Classification According to Other accompanying disorders. Spitz's classification of safety in esophageal atresia: Group I: birth weight> 1500 g without major heart defects (major); Group II: birth weight <1500 g or with major heart defects; Group III: birth weight <1500 g and major heart defects [25].

The purpose of this study was to see the prevalence of esophageal atresia based on the Waterston classification, the mortality rates for children with esophageal atresia, and the causes of the high mortality rates for children with esophageal atresia.

Methods and Material

This study was a survey research conducted from April 10, 2021 to April 15, 2021, with the sample in the form of all patients registered in the operating room register at Department of Surgery, Pediatric Surgery Division, Tarakan General Hospital from 2015 to 2021. The inclusion criteria in this study were children with esophagus atresia and received surgical therapy at Tarakan Hospital, as well as further treatment at Tarakan Hospital. The exclusion criteria in this study were incomplete medical record data, patients who received surgical therapy elsewhere, and patients who refused to participate in the study. The sampling method in this study was total sampling. This research procedure begins with the preparation of a research proposal, submitting ethical clearance at the Ethical Clearance department of Tarakan Hospital, requesting permission from several related departments, data collection, data analysis, and data presentation. The variables in this study were the esophageal classification based on the Waterston and Gross systems, age of the child, mortality rate, and cause of death. Data analysis used in this research is descriptive analysis in which numerical data is presented in the form of a centralized data distribution in the form of mean and standard deviation for normal data distribution, as well as median, minimum-maximum for abnormal data distribution. Categorical data will be presented in the form of proportions and percentages.

Results

There were 31 cases of esophageal atresia recorded in the Department of Surgery, Pediatric Surgery Division, Tarakan General Hospital from 2015 to 2021. In other words, the incidence of esophageal atresia is 1.51%. There were 31 cases of esophageal atresia that met the inclusion criteria during the 2014-2021 period at Tarakan Hospital. The demographic characteristics of respondents were dominated by 16 (51.6%) men, the average age was 8.00 (2.00 - 19.00) months, the type of esophageal atresia type B was 28 (90.3%) respondents, and Waterston C by 9 (29.0%) respondents. The mortality rate was found in 16 (51.6%) respondents with etiologies such as sepsis, perforation, and aspiration (Table 1).

Table 1: Demographic Characteristics

Parametric	N (%)	Mean (SD)	Med (Min – Max)
Sex • Male • Female	 16 (51, 6%) 15 (48, 4%)	-	-
Age	-	8,23 (4,88)	8,00 (2,00 – 19,00)
Type Of Esophageal Atresia • Esophageal Atresia Type A • Esophageal Atresia Type B • Esophageal Atresia Type C • Esophageal Atresia Type D • Esophageal Atresia Type E	 3 (9, 7%) - 28 (90, 3%) - -	-	-
Esophageal atresia according to Waterston • Waterston A • Waterston B • Waterston C	 6 (19, 4%) 16 (51, 6%) 9 (29, 0%)	-	-
Mortality/ Died		-	-

• Yes • No	16 (51, 6%) 15 (48, 4%)		
Dead cause • Sepsis • Leakage • Perforation	11 (73, 3%) 1 (6, 7%) 3 (10, 0%)	-	-

Statistical analysis using Fisher's Exact found that there was no significant relationship between gender, Type of Esophageal Atresia, and Waterston on the outcome of esophageal atresia (p-value> 0.05). However, there was a strong correlation between the etiology of sepsis, leakage, and perforation on the outcome of esophageal atresia (p-value <0.001; r: 0.915). On the other hand, the mean age played an important role in the outcome in patients with esophageal atresia with a weak correlation (p-value 0.045; r: 0.362). (Table. 2)

Table 2: Cross tabulation test of gender, age, etiology, esophageal atresia type and waterston type on output results

Parametric		Output				r	*P-value*
		Died		Survival			
		N	%	N	%		
Sex	Male	8	50	8	50	0,033	0,853
	Female	8	53,3	7	46,7		
Etiology	Sepsis	12	100	-	-	0,915	< 0,001
	Leakage	1	100	-	-		
	Perforation	3	100	-	-		
Type of Esophageal Atresia	Type-A	2	66,7	1	33,3	0,098	0,583
	Type-B	-	-	-	-		
	Type-C	14	50	14	50		
Waterston	A	4	66,7	2	33,3	0,044	0,816
	B	7	43,8	9	56,3		
	C	5	55,6	4	44,4		
Age		10,00 (5,18)jo		6,33 (3,85)		0,362	0,045

Discussion

Research in Serbia in 2015 entitled Analysis of Prognostic Factors and Mortality in Children with Esophageal Atresia describes that the survival rate for infants with esophageal atresia is 85-90% and higher in developed countries by up to 95%. This research is a survey research with a total sample of 60 babies (40 boys and 20 girls), found The birth weight, mean Apgar score (AS) value, gestational age, and duration of birth-operative treatment all had a significant effect on treated patients' mortality ($p < 0.05$). Thirty five percent (35%) of newborns had aspiration pneumonia at the time of hospitalization, and 86.7 percent were operated on within the first 48 hours. The presence of associated anomalies had a significant effect on the death rate of patients treated ($p < 0.05$). While the incidence of postoperative complications was comparable to that in developed countries, total mortality was significantly higher (28.3 percent), with sepsis as the leading cause of death. Postoperative complications and sepsis had a significant effect on patient mortality ($p < 0.05$) [26].

A 2017 Chinese study entitled A scoring system to predict mortality in infants with esophageal atresia A case-control study, with 198 infants taken from medical records of patients with esophageal atresia from March 2004 to June 2016, found that The overall mortality rate was 18.1% (36/198). The nonsurvivor group had a significantly higher rate of prematurity, low birth weight, long gap, anastomotic leak, respiratory failure, postoperative sepsis, respiratory distress syndrome, pneumothorax, and septic shock than the survivor group ($P < 0$, 05). Anastomotic leak (OR: 10.75, 95% CI: 3.113–37.128), respiratory failure (OR: 4.104, 95% CI: 2,292–7,355), postoperative sepsis (OR: 3,564, 95% CI: 1,516–8,375), and low birth weight (OR: 8,379, 95% CI: 3,357–20,917) were all associated with a high mortality rate in the logistic regression analysis. At a 2-point cutoff, a scoring model with a sensitivity of 0.861, a specificity of 0.827, a positive predictive value of 0.524, and a negative predictive value of 0.963 was developed. The area under the receiver-operating characteristic curve for death from EA was 0.905 (95 percent confidence interval [CI], 0.863–0.948, P =0.000). As the scores increased, the mortality rate increased rapidly, and all patients with scores of 5 were edited. Anastomotic leak, respiratory failure, postoperative sepsis, and low birth weight are all independently associated with death in EA. Infants with a predictive score of 5 had a greater than 50% chance of dying [27].

Research in China in 2020 entitled Prognostic Factors on Low-Birth-Weight Infants of Atresia and / or Tracheoesophageal Fistula: a Cohort Study, with a study sample of 81 patients, found that the overall prevalence of preoperative, intraoperative, and postoperative complications was significantly higher in low-birth-weight patients (P = 0.002, P = 0.001, P = 0.001), but OS was lower (75.9 percent vs 92.3 percent, p = 0.027). The postoperative OS of patients with stress hyperglycemia was lower than that of patients with euglycemia (81.0 percent vs 93.6 percent, p = 0.03) [28].

Another study in India in 2008, entitled Esophageal atresia: Factors influencing survival - Experience at an Indian tertiary center with a total sample size of 127 samples collected from February 2004 to May 2006, explained that In 117 cases, EA with TEF was the most common type (92 percent). Associated congenital anomalies were present in 52 (41 percent) of the patients, with cardiac anomalies being the most common, followed by gastrointestinal anomalies. VACTERL was discovered in 6 (5%) of the cases. Prematurity, associated congenital anomalies, esophageal gap, and preoperative respiratory status were all significant predictors of survival (P = 0.001). The majority of patients underwent primary extrapleural repair. In 46 cases, the Azygos vein was preserved, and no retropleural drainage was used in 27 cases. In 19 cases, including 6 cases of isolated esophageal atresia, staged procedures were performed. The most common early

postoperative complications were pneumonia and sepsis (42 percent). The most common causes of death were hypoxia and cardiorespiratory arrest (11 cases). Anastomotic leaks complicated 13 cases, 9 of which were major leaks and 4 of which were minor leaks. Seven people died as a result of a major leak followed by sepsis. Survival according to Waterston criteria was 100% in group A, 83 percent in group B, and 22 percent in group C [29].

Conclusion

From 2015 to 2021, the incidence of esophageal atresia was 1.51% in the Department of Surgery, Pediatric Surgery Division, Tarakan General Hospital. The demographic characteristics of respondents were dominated by 16 (51.6%) men, the average age was 8.00 (2.00 - 19.00) months, the type of esophageal atresia type B was 28 (90.3%) respondents, and Waterston C was 9 (29.0%) respondents. The mortality rate was discovered in 16 (51.6%) of the respondents with etiologies such as sepsis, perforation and aspiration. In bivariate analysis, it was discovered that the presence of comorbidities such as sepsis, leakage, and perforation significantly contributed to mortality (p-value 0.001), and there was a significant mean age difference between the groups who died and survived.

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