ORIGINAL ARTICLE

Mortality in esophageal atresia: Assessment of probable risk factors (10 years' experience)

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Background: Esophageal atresia (EA) is a common congenital anomaly. In this study, we evaluated the mortality and its risk factors in patients born with esophageal atresia. **Methods and Materials:** A database of 206 consecutive patients treated for EA was developed in ST-Zahra hospital of Isfahan between 1994 and 2004. **Results:** In this study, 206 patients were evaluated. The most common type of EA was type C (86.4%). Mortality rate was 56%; it was more common in patients with congenital malformation, in late operation and in low birth weight and premature babies. Sepsis was the most common cause of death. **Conclusions:** For improvement of EA survival, we should improve operation techniques, supportive care, and ICU management. It is also important to operate these patients as soon as possible.

Key words: Esophageal atresia, mortality, morbidity

INTRODUCTION

Esophageal atresia/tracheoesophageal fistula (EA/TEF) is a life-threatening congenital malformation of the esophagus associated with significant neonatal morbidity and mortality. Because of the major anatomical and physiological derangement, nearly all infants with this anomaly exhibit a high degree of morbidity, including feeding intolerance, failure to thrive, and prolonged duration of hospitalization. EA/TEF survival has improved significantly since the initial successful surgical correction performed by Cameron height in the 1940. The initial mortality in patient with this condition was high, but with improvement in surgical technique, suture materials, neonatal intensive care, and antibiotics[1,2] majority of these infants now survive. The patients who are treated nowadays for EA/TEF in a pediatric surgical center are born earlier, weight less, and have more associated anomalies than those treated 50 years ago; therefore, periodic evaluation of factors associated with mortality in these patients will be important. The purpose of this study was to evaluate the factors associated with mortality in neonates admitted to intensive care for the management of neonates with EA/TEF.

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MATERIALS AND METHODS

The clinical study was approved by the ethical committee of our hospital. Data were collected from the one the largest neonatal surgery unit in central of Iran (St Zahra Hospital, Isfahan). A retrospective review was performed of an identified neonatal surgery intensive care patient dataset. Participants were all neonates consecutively admitted to the neonatal surgery unit of the study hospital for management of EA/TEF. This study included patients who were managed between January 1994 and January 2004. Neonates who died in the delivery room or those who were not admitted to the neonatal surgery intensive care unit were excluded. The gestational age assignment was based on the best obstetrical estimate prior to the delivery and was recorded as completed weeks. Neonates were diagnosed with EA/TEF based on the physician's inability to pass a 10 French nasogastric tube into the stomach, with the tube coiling in the upper esophageal pouch on Chest X-ray. After a work up that included an echocardiogram and an abdominal ultrasound, thoracotomy and primary anastomosis with fistula ligation was done. In the case of a significant gap between the ends, surgical options included a Livaditis myotomy, anastomosis under tension or neck esophagostomy + gastrostomy with late second procedure (one year after birth). In very premature infants, fistula ligation was performed at

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the first exploration and reconstruction was performed in second stage operation (after patient stability but in the same time of hospitalization). Postoperatively, all infants were placed on antibiotic medication. Collected data included EA/TEF type (A: Atresia alone; B: Atresia + Proximal TEF ; C: Atresia + Distal TEF; D: Atresia + Proximal and distal fistulas; and E: TEF without atresia), associated congenital malformations, and treatment options. For further analysis, the cause of death was categorized into four groups as pulmonary failure, sepsis, pneumothorax, and other causes. Written consent was obtained from the parents after they were told the purpose of the study.

Differences in the characteristics of the patients with EA/TEF who died were compared to those who were discharged home. We compared the two population samples using univariate analysis. Contentious variables were evaluated with 2-tailed *t*-tests. Categorical variables were evaluated with 2-tailed chi-square or exact Fisher's tests. Nonparametric data were assessed by Kruskal-Wallis analysis of variance. The Statistical Package for Social Sciences (SPSS) software (11.5) was used for data analysis. Data was expressed as mean ± SD. *P* values less than 0.05 were considered significant.

RESULTS

The study group consisted of 127 boys (61.7%) and 79 girls (38.3%). The mean birth weight was 2435 g,(1000 - 4000 g); 48 patients (23.3%) were born prematurely with a gestational age less than 37 weeks. From these patients, 12 cases (25%) had a birth weight less than 1500 g. The mean maternal age was 25.6 years old, range 15- 42 years old. In 158 term patients, two patients (1.3%) had low birth weight. Table 1 shows the distribution of the types of atresia.

One hundred twenty-nine patients (62.6%) had no other congenital malformations, the remaining 77 patients (374%) had one or more associated congenital malformation [Table 2].

Pneumonia was present preoperatively in 37 patients (19.2%). An operation was performed in 173 patients. Thirty three patients died after gastrostomy (before main operation). The operations performed are listed in Table 3.

A total of postoperative complications occurred, included right pneumothorax (30.8%), left pneumothorax (8.5%), wound infection (8.2%), anastomotic leak (17.1%), TEF recurrence (4.7%), esophageal stenosis (46.1%), GERD (43.8%), tracheomalacia (38.2%), esophageal dysmotility (5.6%), chest tube infection (4.4%), intestinal adhesions (1.8%), empyema (1.8%), gastrostomy fistula (0.9%), stomach rupture (1.8%), and pyloric stenosis (1.5%).

Table 1: Types of esophageal atresia in 206 patients

Type	Number	%
A	12	5.6
B	0	0
C	177	86
D	14	6.8
E	3	1.1

Table 2: Congenital malformations associated with esophageal atresia

Malformation	Number	%
Vertebral	12	5.8
Anorectal	27	13.1
Cardiac	25	12.1
Renal	13	6.3
Duodenum	5	2.4
Undescended testis	5	2.4
Club foot	5	2.4
Vascular	5	2.4
Ribs	4	1.9
Ambigus genitalia	3	1.5
Cleft lip	2	1
Pulmonary hypoplasia	1	0.5

Table 3: Treatment of 173 patients with esophageal atresia

Operation	Number	%
Primary repair	70	40.4
Primary repair + gastrostomy	12	6.9
Gastrostomy + secondary repair	83	47.9
Colon bypass	8	4.6

In 25 patients, esophageal anastomosis had performed in first two days of birth. Mortality rate was 56%. Death was more common in patients with congenital malformations (68.1% *vs.* 46.7%, $P = 0.004$), patients with late operation (33.1% *vs.* 49.1%, $P = 0.03$),and patients with secondary repair (55.3% *vs.* 31.6%, $P = 0.02$). It also was more in low birth weight ($P < 0.001$) and premature ($P < 0.001$) patients.

Sepsis was the most frequent cause of death (50.5%), followed by pulmonary insufficiency (29%).

DISCUSSION

Congenital EA and/or TEF are common congenital anomalies.[2] Although, the mortality of patients born with EA has decreased from 61% to 11% since 1947,[4] but in our study, death rate was high. In the most recent years, 78% of mortality is caused by associated congenital anomalies. In our study, we also showed this conclusion. The reduction in mortality can be achieved by early treatment of these anomalies, and it seems that our limitations in malformations management cause high mortality in our

population. The increased survival of population of EA seems to be mainly due to improved preoperative and postoperative care and the influence of ICU management. Operation time had significant effect in outcome and it seems that in our society with malformation management limitations, early operation can increase survival. In this study, we showed the significance of LBW (Birth weight <2500g) and prematurity on outcome. This is in sharp contrast to Calisti *et al*. study.[5] In their study, birth weight and week of delivery did not seem to influence outcome. Congenital anomalies can affect as confounding variables in evaluation of with and gestational age effects on death.

There have been multiple reports on the increased incidence of pyloric stenosis after repair of esophageal atresia.[6-8] It is not clear what the cause of this increased incidence is. Vagal nerve lesion, gastrostomy, and transpyloric feeding tubes have been mentioned as possible causes of hypertrophic pyloric stenosis after correction of esophageal atresia.[9]

The incidence of pyloric stenosis in the normal population is 0.1% to 1%. In our study population, pyloric stenosis was as seen in normal population.

Tracheomalacia is a common problem after correction of esophageal atresia. In several large series,[10,11] its incidence had 7 to 16%. In our study, 38.2% of patients had tracheomalacia. It seems that GERD can aggravate tracheomalacia,[11] and in this study, GRED rate was 43.8%.

Anastomotic leak and pneumothorax were more common in our population compared with others.[4] Preventing anastomotic tension and pleural lesion during surgery are important to diminish the risk of postoperative complications. Anastomotic tension causes a significantly higher risk for development of anastomotic leak, and a pleural lesion causes a significantly higher risk for the development of a pneumothorax.

The major aspect of this study that is hopeful is the improvement of the outcome in the second lustrum compared to the first lustrum. Factors that might have contributed to this improvement were improvements in surgical techniques, suture materials, neonatal intensive care, antibiotics, and the developments in parenteral and enteral nutrition, and a better understanding of developmental embryology and anesthesia.

In summary, this study shows that congenital malformations, operation time, and birth weight and age are important risk factors for EA treatment outcome. We should improve operation techniques, supportive care, and aggressive intensive care treatment of postoperative morbidity to decrease mortality after correction of esophageal atresia.

REFERENCES

1. Burjonrappa SC, Youssef S, St-vil D. What is the incidence of Barrett's and gastric metaplasia in esophageal atresia/tracheoesophageal fistula(EA/TEF) patients. Eur J Pediatr Surg 2011;21:25-9.
2. Kovesi T, Rubin S. Long-term complications of congenital esophageal atresia and/or tracheoeosphageal fistula. Chest 2004;126:915-25.
3. Kawahara H, Imvra K, Yagi M, Kubota A. Clinical characteristics of congenital esophageal stenosis distal to associated esophageal atresia. Surgery 2001;129:29-38.
4. Deurloo JA, Ekkelkamp S, Schoorl M, Heij HA, Aronson DC. Esophageal atresia: Historical evolution of management and results in 371 patients. Ann Thorac Surg 2002;73:267-72.
5. Calisti A, Oriolo L, Nanni L, Molle P, Briganti V, D'Urzo C. Mortalily and long term morbidity in esophageal atresia: The reduced impact of low birth weight and maturity on surgical outcome. J Perinat Med 2004;32:171-5.
6. Okada A, Usui N, Inoue M, Kawahara H, Kubota A, Imura K, *et al*. Esophageal atresia in Osaka: A review of 39 years, experience. J pediatr surg 1997;32:1570-4.
7. Kiliç N, Gürpinar A, Kiriştioğlu I, Doğruyol H. Association of oesophageal atresia and hypertrophic pyloric stenosis. Acta Paediatr 2000 ,89:118-9.
8. Zhang ZB. Esophageal atresia/tracheoesophageal fistula complicated by hypertrophic pyloric stenosis: A case report. Zhongguo Dang Dai Er Ke Za Zhi 2010;12:71-2.
9. Oğuzkurt P, Tanyel FC, Haliloğlu M, Hiçsönmez A. An uncommon association of H-type tracheoesophageal fistula with infantile hypertrophic pyloric stenosis. Turk J Pediatr 1999;41:143-6.
10. Lacher M, Froehlich S, von Schweinitz D, Dietz HG. Early and long term outcome in children with esophageal atresia treated over the last 22 years. Klin Padiatr 2010;222:296-301.
11. Tsai J, Berkery D, Wesson S.F. Esophageal atresia and tracheoesophageal fistula: Surgical experience over two decades. Ann Thorac Surg 1997;64:778-83.

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