Esophageal Atresia: Recent Five Years' Mortality and Morbidity

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Abstract- Esophageal atresia (EA) is a rare congenital anomaly that may be isolated or associated with other anomalies requiring prompt medical and surgical planning for optimal result. This study was conducted *to* show our recent experience on the outcome of treated patients in two hospitals affiliated to Tehran University of Medical Sciences (TUMS). From January 2008 to May 2013 records of 43 neonates patients (23 male) with EA admitted in 2 children centers and all related data including demographic, diagnostic associated anomalies, surgical approaches, birth weight, mortality, and complications were collected. Inability of feeding and swallowing was the most common symptoms (in 90,6%), associated CHD (44%), and Type C (EA) was the most common type of EA observed in 86% patients, The mortality rate was 4.7% and most common complication was anastomotic stricture (AS) in 60% of patients. Our study showed that despite improvements in management and survival of an infant with (EA), still sepsis, aspiration pneumonia, prematurity, and low birth weight and severe Congenital Heart Disease (CHD) were independent etiology of death and birth weight < 2.500 gr has a significant effect on the occurrence of postoperative complications.

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Keywords: EA; Outcome; Birth-weight; Congenital heart disease; Operation

Introduction

EA (EA) with or without a TEF (TEF) is a known correctable congenital malformation, with the prevalence of around of 2.4 per 10 000 births (2). EA may be isolated or associated with other anomalies, requiring prompt diagnosis and planning for investigation, medical and surgical intervention for the optimal outcome (1, 2). The prognosis of EA has improved significantly to approximately 95% in recent decades. Waterston's risk classification based on the risk factors consist of birth weight, presence of associated anomalies and pneumonia, and later Spitz's classification is based on the infant's birth weight more or less than 2 kg, absence or presence and severity of CHD has been very useful to predict prognosis, outcome and to design approaches of treatment and follow up of infants with EA. Recent studies indicate that: infants with lower birth weight (< 1500 g), prematurity, major Congenital Heart Disease (CHD), severe associated anomalies and ventilator dependency, and a long gap, still have high mortality and morbidity, but improvements in prenatal diagnosis, neonatal intensive care, anesthesiology, surgical procedures, and parenteral nutrition were effective for better prognosis and requiring to revision of these classifications (3-10). The aim of this study was to demonstrate our recent experience on survival, outcome, and effects of some risk factors such as the patient's weight and cardiac anomalies on treated patients.

Materials and Methods

In this study, all patients diagnosed with EA at Children's Medical Center Hospital and Bahrami

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Hospital affiliated to Tehran University of Medical Sciences from January 2008 to December 2013 included. Patients with incomplete files and patients who did not undergo primary surgery at our center and the one who died before operation were excluded from the study. Based on diagnostic criteria, 43 patients were identified and treated. All data including patient demographics', age at admission time, sex, weight (>2500 and <2500 g), types of EA, associated CHD, length of the esophageal gap (EG) (>2 and <2cm), surgical approach, mortality and complications were collected. We divided infants into two groups related to birth weight less and more than 2.500 g and their relations to mortality and morbidity, overall and for every post-operative complication and also relations to Esophageal Gap EG and CHD) were calculated. Data analysis was done by using SPSS software version 11 and chi-square commands for qualitative variables and T-test for quantitative variables. *P* was defined at a level of 0.05%.

Results

Patient characteristics' are shown in table 1 including 43 patients (23 male and 20 female), the average age at the time of admission was 8 days (2 to 22 days). Based on birth weight 27 (62%) cases had a birth weight more than 2.5 kg, and 16 patients had birth weight less than 2.5 kg, among this group, 5 cases had birth weight less than 1500 g. Inability in feeding and swallowing were the most common symptoms observed in 40 patients (93%), followed by dysphagia, regurgitation (9.06%), weight loss in 13 patients (30.2%) and respiratory infection in 10 patients (23.3%).

Table 1. Characteristics of 43 patients

Demographic		No (%)
Total no: 43		M: 23 F :20
Age: at admission		(2 to 22days) Average age: 8days 4 (9.3%): first 24 hrs 9 (20.9%): second 24 hrs 6 (13.9%): at 3 days 22 (51.1%): > 6 d
Birth weight	**> 2.500 g**	27(62%)
	<2.500 g	16
	<1500g	5
Presentation	**Unable to feed and swallow**	40 (93%)
	Dysphagia	39 (90.6%)
	Regurgitation	39 (90.6%)
	Weight loss	13 (30.2%)
	Respiratory infection	10 (23,3%)
CHD : PDA and VSD		19 (44%)
(EG)		5(11.62%) >2cm 35(83.37%)<2cm
Gross Type of atresia		C (87%), A (6.9%) E (6.9%)

Table1 also shows the distribution of the types of atresia, length of the esophageal gap, and associated congenital heart anomalies, that most common of them were PDA and VSD.

Esophageal Gap (EG)

For emergency medical managing and planning of surgery, all patients routinely were admitted at NIC (neonate intensive care) and investigated for other congenital anomalies as (VACTERL) by radiologist, cardiologist, anesthesiologist, and others necessary consultations. Diagnosis of patients with EA types A and C was with the identification of a coiled gavage radiopaque tube by chest radiography. And H-type TEF by contrast esophagography confirmed by bronchoscopy and passing a ureteral catheter through the TEF. Most

patients (37(86%)) were EA type C, followed by type A (7%), and type E (7%). Surgical approaches were primary esophageal anastomosis with TEF ligation in 35 infants of type C (72.9%) with the lengths of EG less than 2 cm, staged operation was performed in 5 (11.62%) infants including 2 patients with type C and 3 patients with type A with length of EG more than 2 cm. Three patients with EA type H underwent transcervical fistula division and ligation.

The most complication which was observed in our series was Anastomotic Stricture (AS) that occurred in 26 (60%) patients followed by GERD in 23 (53.5%)patients, anastomotic leak (AL)in 16 patients (37.2%), TEF recurrence 3(7%). The mortality rate in our studied patients was 6.97% (3 patients). Causes of death were sepsis in the first patient, aspiration pneumonia in the second patient, the third patient had Low Birth Weight (LBW), prematurity, and cardiac anomaly who died after gastrostomy. Another patient died before performing any surgery and was excluded from the mortality rate (Figure 1).

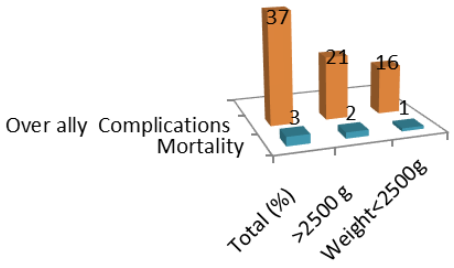

Figure 1. The frequency of (complication overall) and (mortality) relation to weight

Table 2 shows summarized the relationship between the frequency of Outcome (mortality and morbidities overall) and separately for every complication) to two Weight's groups.

Relations of the frequency of complications to risk factors including CHA and also EG length in two weight's group of patients is shown in table 3

Table 2. The relationship between the frequency of outcome (mortality &morbidities) and weight

	Outcome	Weight<2500g	>2500 g	Total (%)	*P*
1	**Mortality**	1 (6.25%)	2 (4.65%)	3 (7%)	0/706
2	**Overall complications**	16 (100%)	21 (77%)	37 (86%)	0/042
2.1	**AL**	7 (43.7%)	9 (33%)	16 (37.2%)	0/001
2.2	**AS**	11 (68%)	16 (59.2%)	27 (62.7%)	0/207
2.3	**TEF recurrence**	2 (12.5%)	1 (3.7%)	3 (7%)	0/136
2.4	**GER**	11 (67.8%)	11 (40.7%)	22 (51.1%)	0/001

Anastomotic leakage (AL), Anastomotic Stricture (AS) ,Gastroesophageal reflux (GER)

Table 3. The frequency of complications in relation to CHA and EG length in two weight's group

Post-operative complications	birth weight	< 2500 g	> 2500 g	Total	*P*
		16 (33%)	21 (77%)	37 (86%)	0/042
CHD and complications		17 (89.4%)	2 (10.6%)	19 (100%)	0/453
Length of (EG) and	**Long gap**	4 (80%)	1 (20%)	5 (100%)	0/248
Complications	**Short gap**	30 (78%)	8 (22%)	38 (100%)	0/248

Esophageal Gap (EG)

Discussion

In our study, the mortality rate was (6.97%) and although the relationship between births weight< 2,500 gr was not significant, but three patients died; first due to sepsis, the second patient due to aspiration pneumonia and the third one had LBW, prematurity, and cardiac anomaly and died after gastrostomy.

Studies by Waterston's and Spitz (22,23) showed that major cardiac malformations and LBW (<1500 g) are two predictors for the high rate of mortality. In a similar study, performed in Iran mortality rate was 56% (24) that was higher than the rate seen in our study. In another large cohort study, performed in the Netherlands the mortality of patients born with EA decreased from 61% to 11% since 1947 (11). Our study and others' reports showed that improvements in surgical techniques, neonatal intensive care, and parenteral and

enteral nutrition, and anesthesia experience had decreased mortality of the patients (23).

In our study, there was a significant relationship between birth weight and postoperative complications overall. Especially there was a significant relationship between birth weight and AL (*P*=0.001) and occurrence of GER but was not with other complications (*P*=0.001). In the present study, although relation of birth weight <2.500 gr and frequency of AS was not significant, but because of complications and AL and GERD that both are most causes of AS, both affect the occurrence of stricture, that next studies with multi vibrate analysis are required (Table2).

In this study, there was not a significant relationship between CHD. (*P*=0.453), length of EG (*P*=0.248) with post-operative complications. This may be different from some reports, and so next studies are required (Table 3).

ALwas observed in 37.2% of our patients which was higher compared with the reported rate by Deurloo (9%) and Davari (17.1%) (11). It has been shown that ALsignificantly is caused by anastomotic tension as a higher risk (11).

Complication of AS developed, in (60%) of our patients. In other studies, major surgical complications affected 24 to 78% of children with EA (12,13). An end-to-end repair with minimal tension appears protective. Although boogies were the first tools widely available to dilate AS, balloon dilatation is now thought to be safer and more effective (14). If AS persist, surgical interventions may be necessary.

In our study, GER developed in 53.5% of patients. Based on several documented studies, the occurrence rate of GER varies between 25.5% to 62.9% (13,15,16). So GER should be considered a component of EA and TEF because it is very often diagnosed after neonatal repair of this condition. GER may cause AS, failure to thrive, respiratory problems, and in the long term, esophagitis and esophageal metaplasia (17). Different factors such a suturing technique; EG, the tension between esophageal pouches, AL, and GER primary or secondary to esophageal manipulation related to the operation are involved in the pathogenesis of AS that was most post-operative complication in our study. Therefore to minimize AS formation, aggressive treatment of GER is critical that includes medical therapy and ant reflux surgery

The incidence of recurrent TEF in our series was 7% similar to reports of Spitz (18) and Tsai (19) that were 12% and 10%, respectively. The pathogenesis of recurrent TEF is an anastomotic suture line leak with erosion through the site of the previous repair of the TEF (19). Because recurrent TEFs rarely close spontaneously, It has been advocated reoperation with interposition of a pleural or pericardial flap after fistula division such as our 3 patients (19,20), and slide tracheoplasty that recently has been suggested (21).

Our study showed that despite improvements in management and survival of infants with EA still sepsis, aspiration pneumonia, prematurity, LBW, and severe cardiac anomalies are independent major etiologies of death in our patients.

Birth weight <2.500 gr significantly affected the occurrence of post-operative complications and especially anastomotic leakages and GERD.

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