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Surgical Outcome of 65 Cases of Congenital Esophageal Atresia with Tracheoesophageal Fistula: Experience of 5 Years in Two Institutes

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Abstract

Background Congenital esophageal atresia and tracheoesophageal fistula are well-known congenital anomalies which affect 1 in 2400 – 4500 live births. The survival rate has dramatically improved in the last decades due to the advances in the neonatal intensive care, anesthetic management, ventilatory support and the surgical techniques.

Objective To evaluate the surgical outcomes and postoperative complications in patient with esophageal atresia and tracheoesophageal fistula who were admitted to our hospital.

Methods A retrospective study of 65 cases of esophageal atresia with distal tracheoesophageal fistula was studied over a period of 5 years from January 2008 to January 2013. Patients demographic, frequency of associated anomalies, postoperative complications and surgical outcomes were studied.

Results There were 38 male and 27 female with frequency of forty-seven (72%) infants were full term and 18 (28%) were preterm. Survival rate was (66%) for newborn who underwent surgery within the first 24 hours which was higher than those newborn with age above two days. Congenital heart disease was the commonest associated congenital anomalies and accounted for 28% of all our patients. Our patients developed many complications including respiratory tract infection (26%) and anastomatic leak (20%) as the most common complications.

Conclusion The survival rate of the patients with EA/TEF is influenced mainly by associated life -threatening congenital anomalies, prematurity of newborn and the age at the time of surgical repair.

Keywords Esophageal atresia, tracheoesophageal fistula, congenital anomalies.

List of abbreviations: EA = Esophageal atresia, TEF = TracheoEsophageal fisula

Introduction

Esophageal atresia (EA) with distal fistula is the most common subtype of EA, accounting for about 85% of this anomaly. The much-dilated proximal esophagus has a thickened wall and descends into the superior mediastanium. The distal esophagus is slender and has a thin wall. It enters the trachea posteriorly either at the level the level of the carina or 1 to 2 cm above. The survival of infants born with EA, tracheoesophageal fistula (TEF), or both has improved dramatically since Cameron Haight's first successful repair in 1941 [(1,2)]. Since then, the management of EA and TEF has evolved considerably over the years. At present, an overall survival rate of 85% -90% and survival rate of over 95% in those without major anomalies have been reported from developed countries. This is not the same in developing countries, where many preoperative, postoperative and socioeconomic factors continue to contribute to the persisting high mortality [(3)].

The incidence of the various forms of EA in the general population is approximately 1 in 4000 live births. Although nearly all esophageal atresia variants seem to be sporadic, familial esophageal atresia has been reported. The incidence of recognizable congenital defects associate with EA is about 55%. The presence of cardiac malformation is particularly important and is often the major determinant of mortality [3,4].

The infant with EA is unable to swallow, drools saliva and spits up indigestive formula. As liquid pooling in the blind proximal esophageal pouch spills into the airway, the infant may cough or choke.

The diagnosis of EA can be made by chest radiography after placement of a soft nasogastric tube as far as possible in the esophagus. The chest radiograph shows the tube coiled in the upper mediatanium and the presence of gas filled intestinal loops establishes the presence of distal TEF [4,5]. The ideal management of AE With TEF is division of fistula and primary esophageal repair performed in single operation during the newborn period of life. This approach is successful in most patients born with EA and distal TEF, today. The premature infants with significant respiratory distress syndrome or newborn with associated congenital anomalies, specifically cardiac lesions, for whom it is difficult to provide effective support with mechanical ventilation, may not tolerate the lung retraction or operative time necessary for complete repair during single setting. Early surgical repair is done for those babies with adequate arterial blood gases, adequate weight and no significant associated anomalies and delayed repair (gastrostomy first) is used for all other patients.

The objectives of this study was to evaluate the surgical outcomes and postoperative complications in patient with esophageal atresia and tracheoesophageal fistula who were admitted to our hospital.

Methods

This is a retrospective study of 65 EA with distal TEF cases divided into 38 boys (58%) and 27 girls (41%) who were admitted to the pediatric surgery center at Central Teaching Hospital of Pediatrics and Al-Kadhymia Pediatric Hospital (Baghdad) over a period of 5 years from January 2008 to January 2013. Their medical records were reviewed for the age, sex, maturity, birth weight, associated congenital anomalies, operative technique, morbidity and mortality.

The diagnosis was depending on clinical presentation (frothy secretions due to excessive salivation, tachypnea, cyanosis …) and on plain x-ray of the chest with insertion of 10 Fr radio-opaque rubber catheters to the upper blind esophageal pouch. The diagnosis of the associated congenital anomalies was made on the basis of echocardiography (for congenital heart diseases), abdominal ultrasound for renal anomalies and careful systemic examination.

A standard surgical approach for the repair of EA with distal TEF was through right posterolateral thoracotomy, retorpleural approach with ligation of azygus vein directed toward primary repair with insertion of nasogastric transanastomatic tube for 7 days. Regular follow up of the patients for any late complications especially for anastomatic stricture and for exclusion of any other associated congenital anomalies.

Results

Forty-seven (72%) infants were full term and 18 (28%) were preterm. The average birth weight was 2500±500 grams. Early repair of EA with distal TEF within first 24 hours of life was performed in 15 (23%) newborn with survival rate (66%) which is high in comparison to late presentation and late repair for newborn with age more than 24 hours. Survival rate for newborns who operated at age between 1-3 day were (46%) while for those more than three days were (31%) as shown in table 1.

All our patients underwent surgical repair through the right posterolateral thoracotomy via retorpleural approach whenever possible. A

single layer end to end esophageal anastomosis was performed by using interrupted 5-0 silk with ligation of fistula by 3-0 silk.

Table 1. Demographic details and their effect on survival rate

Variables		Cases		Survival rate	
		No.	%	No.	%
Age at the time of admission	< 1day	15	23	10	67
	1-3 day	37	57	17	46
	>3 day	13	20	4	31
Maturity	Full term	47	72	37	79
	Preterm	18	28	4	22
Sex	Male	38	58	25	65
	Female	27	41	16	59

Associated congenital anomalies occurred in 37 patients (57%) and the congenital heart diseases were the most common associated anomalies and occurred in 18 patients (28%). Other associated congenital anomalies were less common like multiple congenital anomalies (12%), genitourinary anomalies in 6 patients (9%) and imperforate anus In 2 patients to whom were divided descending colostomy done in added for thoracotomy.

Table 2. Associated congenital anomalies with esophageal atresia

Type of congenital anomaly	No.	%
Congenital heart disease	18	28
Multiple congenital anomalies	8	12
Genitourinary	6	9
Imperforate anus	2	3
Musculoskeletal	2	3
Chromosomal (Down's syndrome)	1	1.5
Total	37	57

Table 2 shows associated congenital anomalies with esophageal atresia. The mortality rate was 37% (24 patients) and the most common cause for death was due to associated congenital anomalies and pneumonia, while the survival rate was 63 % (41 patients) which occurred mostly in those patients with early presentation within the first day of life as shown in table 1.

The most common postoperative complication was pneumonia (17 patients -26%) which is a result of preoperative aspiration while anastomatic leak occurred in (13 patients -20%), only three of them needed re-thoracotomy and reanastomosis (two of them died postoperatively) while other 10 patients resolved spontaneously on conservative treatment. Anastomatic stricture occurred in 7 patients (11%) all of them had initially anastomatic leak. We referred all of those patients to cardiothoracic department for esophageal dilatation (Table 3).

Table 3. Postoperative complications

Complication	No.	%
Postoperative RTI	17	26
Anastomatic leak	13	20
Anastomatic stricture	7	11
Recurrent tracheoesophageal fistula	2	3

RTI = respiratory tract infection

During long follow up, two of our patients developed recurrent respiratory tract infection with chocking on feeding and failure to thrive. Ba-swallow was done for them and revealed esophageal stricture. Dilatation done for them by thoracic surgeon for many times but without benefit so we did for them a new thoracotomy one of them at 3 year and another one at 4 year of their age and recurrent tracheoesophageal

fistula recognized so surgical repair was done successfully. According to waterstone classification, the survival rate in group A (birth weight more than 2500 gm and otherwise healthy) was 87% while in group B (birth weight 2000-2500 gm with moderate pneumonia and congenital anomaly) was 64% and in group C (birth weight less than 2000 gm with severe pneumonia and congenital anomaly) was 21% (Table 4).

Table 4. Survival rate in patients with esophageal atresia based on Waterston classification

Group	No. of cases	Survival Rate	
		No.	%
A	23	20	87
B	18	18	100
C	14	3	21

Discussion

Esophageal atresia with or without tracheoesophageal fistula is one of commonest gastrointestinal malformations, second only to anorectal malformations. The exact incidence of EA with or without TEF is not known but incidence of 1 in 3000 to 1 in 4500 live births have been reported [7].

The Waterston *et al* devised one of the first classification systems for EA - TEF. They compared the results based on preoperative stratification of cases by severity and expected outcome. Risk factors that Waterston included were pneumonia, birth weight, and associated congenital anomalies [8]. In 1962, Waterstone proposed the risk classification of patients with EA and he reported that survival rate was 100% for group A, 86% for group B and 73% for group C which is higher in comparison to our study (group A: 86.9%, group B: 46.2% and group c: 21.4%) this low survival rate in our hospital is attributed to bad post-operative respiratory care in compare to well-developed countries.

In our study, the survival rate was 66% for newborn who underwent to the surgery within the first 24 hours of life which is high in compare to other newborn older than this age because in early presentation there is low risk for aspiration pneumonia. The prematurity is considered as risk factor for survival for our patient not due to the surgical technique but due to lack of respiratory support preoperative and postoperative (intensive respiratory care unit) and also due to physiological problems in premature neonates. The overall survival rate was 63% which similar to the Maj Daud et al study which was done in Bangladesh [9] but it is lower than Chia-Feng Yang *et al* [10] study where survival rate (83%).

In our study 57% of patients had associated congenital and the commonest being congenital heart diseases, which occurred in 28%. Maj Daud *et al* reported 42.85% associated anomalies with congenital heart disease 29%. Spitz et al [11] reported 47% and Rokitansky *et al* [12] reported 52.4% associated congenital anomalies. Thirty two patients (49.2%) developed different complications with postoperative respiratory tract infection due to aspiration was the commonest complication (26%) followed by anastomatic leak (20%). similar postoperative complications are reported by Hassab *et al* [13] and Okada *et al* [14].

In conclusion, the surgical outcome in a neonate with EA and TEF influenced by many factors including the age associated congenital anomalies and maturity of neonate. The survival rate in our study was 66% which was low as compared with developed countries due to lack of advanced respiratory care and late presentation.

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Author Contribution

The authors are responsible for preparing for all steps of this article.

Declaration of Interest

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