Original Article

Oesophageal Atresia - Experience of 7 Years with Data Evaluation

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ABSTRACT

Objective: Study conducted for evaluation of Oesophageal atresia patients to assess the factors influencing the survival and outcome.

Design of Study: Prospective analytical study.

Place & Duration of Study: This study was conducted in dept. of Paediatric surgery at peoples medical college, Nawabshah. from September 2001 to September 2008.

Patients and Methods: A prospective analysis of 37 cases of Oesophageal Atresia (EA) was performed. In this study, 37 neonates were admitted with the diagnosis of EA with or without TEF .The data were collected retrospectively from hospital charts. The preoperative assessment of upper pouch was done with plain X-ray chest with 8 Fr Red Rubber catheter. The associated congenital anomalies were evaluated on the basis of careful examination, radiological and sonological investigations.

Results: The commonest type of Oesophageal Atresia was with distal Tracheoesophageal Fistula (TEF) in 32 cases (86.48 %). Associated anomalies were present in 50% patients, cardiac was commonest followed by gastrointestinal anomalies. Vacterl association was found in 6 (16.21 %) cases. Prematurity, associated congenital anomalies, gap between esophageal ends and preoperative respiratory status were the significant factors affecting the survival. Primary extra pleural repair was the surgical approach in most of the patients except two with difficulty that change to intra pleural approach. Retro pleural drainage was used in 32 classical type 1 cases. Staged procedures were performed in 5 cases of isolated Oesophageal Atresia. Pneumonitis and sepsis were the most common early postoperative complications (30%). Sepsis and cardio respiratory arrest were the most common causes of mortality 11 cases (18.91). Oesophageal leak found in 3 cases, including 2 major and 1 minor leaks. Major leak followed by sepsis caused 1 Deaths. Survival as per Waterston criteria was 85% in group A, 66.6% in group B and 20% in group C.

Conclusion: Factors affecting the survival of patients with Oesophageal Atresia are major or life-threatening associated anomalies, long gap, pneumonia and sepsis at presentation or that acquired during hospitalization and major leaks. The high incidence of low birth weight, delayed diagnosis, poor referral, low-socio economic status and lack of advanced neonatological back up are important contributory factors to poor outcome.

Key Words: Oesophageal Atresia (EA), Tracheoesophageal Fistula (TEF), Pneumonitis and sepsis

INTRODUCTION

The survival of infants born with esophageal atresia (EA), tracheoesophageal fistula (TEF), or both has improved dramatically since Cameron Haight's first successful repair in 1941[1]. Improvements in survival are largely attributable to refinements in neonatal intensive care, anesthetic management, ventilatory support, and surgical techniques. Survival may now be achieved in infants with low birth weight[2], with mortality limited to those patients who have severe life-threatening anomalies

Because of innovative modalities nowadays there has been improvement in management and mortality been reduced coupled with improved anesthesia and good neonatal care. At present, in most of the developed countries, only the presence of associated major congenital anomalies determines the chances of survival[3]. This is not the same in developing countries, where many other preoperative, postoperative and socioeconomic factors continue to contribute to the persisting high mortality. [4],[]In our study we have seen only those factors which has significant importance in the survival rate.

PATIENTS AND METHODS

In this study, 37 neonates were admitted with the diagnosis of EA with or without TEF .The data were collected retrospectively from hospital charts. The preoperative assessment of upper pouch was done with plain X-ray chest with 8 Fr Red Rubber catheter. The associated congenital anomalies were evaluated on the basis of careful examination, radiological and sonological investigations.

Infants were also assigned to risk groups A, B, or C as described by Waterston and associates[6]

- Group A
- 1. Birth weight >2,500 g and well
- Group B
- 1. Birth weight 1,800 to 2,500 g and well, or
- 2. Birth weight >2,500 g but moderate pneumonia and other congenital anomaly
- Group C
- 1. Birth weight <1,800 g or
- 2. Birth weight >1,800 g with severe anomaly or pneumonia

Data collected included age at the time of admission, gestational age, birth weight, sex, site of delivery, history of feeding, associated congenital anomalies, respiratory status, presence of pneumonitis, type of anomaly, operative technique, gap (measurement done intraoperatively), complications and esophageal anastomotic leak and their impact on survival. Waterston prognostic criteria were used for survival.

All the patients operated retropleurally with classical steps ligation of fistula and end to end anastomosis of esophagus and per esophageal drainage along with stent placed. Esophageal anastomosis was performed by 5-0-vicryl single layer interrupted sutures. Feeding started after 48 h of surgery and gradually increased. Because of presence of stent. Contrast esophagogram was done on the sixth day of surgery. Anastomotic leaks after the primary repair were detected either by observing the saliva in the retro pleural drain or by contrast esophagogram. Minor leaks were identified by appearance of frothy saliva in the retro pleural drain with no accompanying deterioration in the general condition. Major leaks were clinically suspected by the contents draining with the accompanying deterioration in the general condition of the patient either due to mediastinitis and septicemia. The finding of contrast in stomach without any clinical deterioration was considered normal.

The term minor leak was used for a small amount of extra pleural leakage and/or a small radiological leak and major leak referred to a large amount of drainage or a leak that caused respiratory symptoms associated with a large defect in anastomosis

RESULTS

Esophageal atresia with distal TEF was the commonest type present in 32(86.48 %) cases; .isolated esophageal atersia without fistula found in 5. Associated congenital anomalies were present in 18 (49.54%) patients, including cardiac diseases in 17 patients, gastrointestinal in 7 cases, vertebral and nervous system anomalies in 8 cases, musculoskeletal anomalies in 4 cases, head and neck problems in 1 cases, genitourinary anomalies in 2 cases and respiratory system anomalies in 2 cases with cleft lip in 1 case. Vacterl association was present in 5 (13.51 %) cases.

Preoperative details and their impact on survival are given in Table 1.

Table 1: Pre. Operative Details With Survival

		Total Cases %	Survival %
Age	< 24 hrs	09 (24.3)	05 (13.5)
	24-48 hrs	13 (35.1)	07 (18.9)
	> 48 hrs	15 (40.5)	14 (37.8)
Maturity	Full Term	27 (72.9)	22 (59.4)
	Pre Term	10 (27.7)	04 (10.8)
Weight	>2.5 kg	20 (54.0)	13 (35.1)
	1.8-2.5 kg	14 (37.8)	10 (27.0)
	< 1.8 kg	03 (8.10)	03 (8.1)
Sex	Male	20 (54.0)	17(45.9)
	Female	17 (45.9)	09(24.3)
Place of Delivery	Home	16 (43.2)	09 (24.3)
	Hospital	21 (56.7)	17 (45.4)
Feeding History	Present	24 (64.8)	20 (54.0)
	Absent	13 (35.1)	06 (16.2)

Only 8(21.62 %) were having no respiratory distress at the time of admission; 12 (32.43 %) were having mild, 8 (21.62 %) were having moderate and 12 (32.43 %) were having severe respiratory distress. Clinically and/or radiologically chest was normal only in 8 (21.62 %) cases, with mild pneumonitis in 8 (21.62 %), moderate pneumonitis in 5 (13.51 %) and severe pneumonitis in 5 (13.51 %) cases. Short gap (<1 cm or one vertebral body) was found in 15 cases (40.54 %), intermediate gap (1-3 cm or 1-3 vertebral bodies) in 12 (32.43 %) and long gap (>3 cm or 3 vertebral bodies) in 5 (13.51 %) cases. Survival rates were 89%, 60% and 23% in cases of mild, moderate and severe respiratory distress, respectively. Survival was 82% in patients with no preoperative pneumonitis falling down to 75%, 72% and 23% with mild pneumonitis, moderate pneumonitis and severe pneumonitis, respectively. Gap was a highly significant factor affecting the survival with 91% survival in short gap and dropping down to 69% and 53% in the intermediate gap and long gap, respectively.

Table 2: Survival according to Waterston

Waterston Classification	Total Cases	Survival
A	20 (54.05%)	17 (85%)
B	12 (32.43%)	08 (66.66)
C	05 (13.51%)	01 (20%)

Our primary approach in all the patients of EA with TEF was extra pleural; however, in 2 cases of EA with

TEF, extra pleural approach was converted to transpleural because of the severe inflammation of parietal pleura in patients with severe pneumonitis or inadvertent breeches in the pleura during the surgery. In the primary repair of EA, azygos vein was ligated in all cases and retro pleural drainage was not performed in 32 cases. Transanastomotic stenting for early feeding after 24 h of surgery was carried out in all 32 (86.4 %) cases.

Table 3: Survival According To Procedure

	Primary Procedure	Survived	Staged Procedure	Survived
Oesophageal atresia with TEF	32	22		
Isolated OA			05	4

Staged procedures were done in 5 cases of EA still under follow-up as given in Table 3

All 4 among 5 cases of isolated esophageal atresia surviving cases are in follow-ups waiting for esophageal replacement.

Out of 32 cases of EA, 10 patients faced early postoperative complications; commonest complication was sepsis with pneumonitis, followed by sepsis alone. The commonest cause for mortality in cases with early postoperative complications was cardio respiratory arrest secondary to hypoxia and pneumonitis in 7 cases. Delayed postoperative complications such as pneumonitis, sepsis, major anastomotic leak, aspiration and tracheomalacia were present in 8 (25%) cases that were responsible for mortality in 3 cases. Major anastomotic leak in patients of EA after primary repair was seen in 2/31 (5.40 %).

DISCUSSION

We have observed few important things to improve outcome in esophageal atresia patients .most of the patient presented late in our territory because of non availability of concerned surgeon nearby so age is bad prognostic marker in our study . Although none of the previous studies from abroad has considered age as a probable risk factor. Prematurity is still a major problem for developing countries due to the additional physiological handicaps in these babies and the increased susceptibility to sepsis. [7

Weight at the time of presentation again a high risk factor and less than 1.8 kg weight has bad prognosis Spitz *et al.* [8] . place of delivery affects the survival as in our study because or referral from rural areas so sepsis is pronounced in such cases as compared to study conducted in developed countries where deliveries conducted in hospitals. Fewer number of female patients is because of male dominant society in our coutries and not presenting female babies .Our findings are similar to that of Bindi *et al* . [9]

Associated anomalies are not so different from those of Hassab *et al.* [10] who reported 60% associated anomalies with VACTERL association in 6%. Spitz *et al.* [8] reported 47%, Saing *et al.* [11] reported 59% and Rokitansky *et al.* [12] reported 52.4% associated congenital anomalies. The survival rate among these cases was low (43%) as compared with 78% in those free from any other congenital anomaly. This shows that association of other congenital anomalies plays a major role in the survival of patients with EA ($P < 0.001$). The survival rate among the patients with EA with congenital heart disease (CHD) was 33% ($P < 0.001$), while in the series of Ein *et al.* , [13] 64% of the neonates of EA with CHD survived. Similar findings had been reported by Choudhury *et al* . [14] The presence of long gap is significantly associated with the poor survival rate ($P < 0.001$). This is because the long gap is associated with high incidence of anastomotic complication and other congenital malformations. Brown *et al.* [16] and Sharma *et al.* , [17] in their study of the measurement of the gap length and mortality in EA, also reached the same conclusions.

The history of feeding that is present in several patients in our study is because of illiteracy and a tradition of giving ghutee after the birth of a neonate, The survival rate was statistically not significant; the reason may be that the neonates who were fed did not have life-threatening anomalies.. Feeding through naso gastric tube makes no significant difference in survival. Kevin *et al,* [18] but we have seen early feeding in stable patients make early recovery. Peri Anastomotic drain placed all patients with classical type esophageal atresia which is veri necessary in all cases as early recognition of complications but Gangopadhyay *et al.* [19] recommended that retro pleural drainage is not necessary in all the cases of EA.

In our study, 32/37 (86.48 %) patients of EA with TEF had primary repair in which

25 (67.56%) patients survived as compared with 36% survival rate in the series of gangopadhayay et al [19] .

In isolated esophageal atresia 5 cases staged procedures (cervical esophagostomy and abdominal esophagostomy or cervical esophagostomy and ligation of the distal esophageal end with gastrostomy) done. Bhatnagar *et al* . [20]studied the exteriorization of the distal esophagus in the abdomen in EA patients with indications of long-gap atresia or isolated esophageal atresia without TEF.

Out of 32 cases of EA, 10 faced early postoperative complications that were responsible for mortality in 7 cases. The most common complication was sepsis , however, the most common cause for mortality was cardio respiratory arrest

Most of these early postoperative complications are not related with surgical procedures. Delayed postoperative complications such as pneumonitis, sepsis, major

anastomotic leak, aspiration and tracheomalacia were present in 8 (15.62 %) cases, and these complications were responsible for mortality in 4 cases. Factors predicting mortality were pneumonia and sepsis at presentation or that acquired during hospitalization, major or life-threatening anomalies, long gaps and major leaks. Similar postoperative complications are also reported by Bindi *et al.* [9] and Hassab *et al.* [10] The incidence of anastomotic leak in patients of EA after primary repair was observed in 3 patients with major leak in 2 (6.25 %) and minor leak in 1 (3.12 %) patients. Spitz *et al.* [8]and McKinnon and Kosloske *et al.* [21] also reported anastomotic leak in 21% cases. Amongst patients with major leak, seven patients associated with pneumonitis or septicemia expired. On comparing the survival rate among patients with major leak and patients without major leak the difference was found to be statistically significant . Statistically significant difference was observed between the survival rates among different classes of Waterston. Spitz *et al.* [8]had earlier reported that survival rates according to Waterston classification was 100% for class A, 86% for class B and 73% for class C cases. In the series of Bhatnagar *et al.* , [7] the survival was maximum in group A (67.6%) and it dropped down to 28.8% in group C. At present, the survival rate has improved in Group A and B; however, it has remained almost the same in group C. On comparing the data of the present series with that of Hassab *et al.*, [10] it was established that although the distribution of cases as per Waterston classification in both these setups were different, the survival rates were almost similar. Our results for survival in class C are lower as compared with other studies, for which the reason might be the higher incidence of low birth weight, delayed diagnosis, poor unsupervised transport, low socioeconomic status and lack of advanced neonatological back up. Waterston classification was statistically the best application in our study. We also propose that the survival in EA can be used as an index for the status of neonatal surgical care because EA had the highest mortality rate amongst all the surgical conditions because of the problems in respiratory care and surgical technical failure. [22]

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