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Original Article

Esophageal Atresia: Management and Outcome in Resource Limited Settings

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Abstract

Background: Esophageal Atresia (EA) with or without associated tracheo-esophageal fistula (TEF) is one of the common congenital anomaly that can be life threatening if left unattended. In low and middle income countries like Pakistan, the management and outcome of such type of cases depends upon many factors related to resource limitation.
Objective: To prospectively evaluate the management and outcome of esophageal atresia at Children Hospital, Pakistan Institute of Medical Sciences (PIMS), Islamabad, Pakistan.
Study design, settings and duration: An Observational- descriptive study was conducted at the Department of Pediatric Surgery, The Children Hospital, PIMS, Islamabad from October 2017 to August 2018.
Methodology: Consecutive patients diagnosed with esophageal atresia were included in the study. Demographic data, investigations, procedure performed and outcome were collected on a pre designed proforma and results were analysed.
Results: Total 140 consecutive patients of esophageal atresia were enrolled in study. Out of 140 patients, 79 (56.4%) were male and 61 (43.6%) were female. Mean age at presentation of esophageal atresia was 5.5 days (ranged from 1-30 days). Mean weight was 2.43 kg. Regarding type of esophageal atresia, 10 (7.1%) patients had type A, 97 (69.3%) had type C, 1 (0.7%) had type E and 1 (0.7%) had type F esophageal atresia. Nine patients with type A underwent cervical esophagostomy along with feeding gastrostomy. Right thoracotomy was performed in 98 cases. End to end esophageal anastomosis was possible in 76 patients. Twenty one patients had long gap EA for which cervical esophagostomy and feeding gastrostomy was done. Sepsis was the main complication post operatively (29.3%) followed by pneumonia (14.3%), Anastomotic leak (7.9%) and surgical site-infection (2.1%). Overall mortality was 57.9% (81/140) with pre operative mortality of 21.4% (30/140) and post operative mortality of 36.5% (51/140). Low birth weight and post operative sepsis, anastomotic leak and pneumonia had statistically significant relationship with mortality using SPSS version 21.
Conclusion: With the improvement of medical facilities, better survival rates of patients with esophageal atresia can be achieved.
Key words: Esophageal atresia, outcome, management, tracheo-esophageal fistula, VACTERL association, Pakistan.

Introduction

Esophageal Atresia (EA) with or without associated tracheo-esophageal fistula (TEF) is a common congenital anomaly occurring in 1/2500 to 1/4000 live births.[1] This is life threatening if not managed early as it can lead to significant morbidity and mortality.[2] Early diagnosis and referral to pediatric surgeon for surgical intervention is important in reducing morbidity and mortality and plays vital role in short term as well as long term prognosis.[3] Outcome depends upon time of diagnosis, birth weight, respiratory complications, chromosomal abnormality, sepsis, socioeconomic status, prematurity, associated congenital anomalies and anastomotic leak.[2] In developing countries poor outcome is associated with delayed diagnosis, low birth weight, presence of sepsis and respiratory complications, low socioeconomic status

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MUN conceptualized the project. MUN & ARC did the data collection. MUN, SS & ARC did the literature search. Statistical analysis was done by NAK. MUN, NJ & MAC did the drafting, revision & writing of manuscript.

and lack of pre and post operative neonatal intensive care.[4, 5]

In developing countries like Pakistan, management of esophageal atresia is still challenging for pediatric surgeons as mortality is very high. Contributing factors to high mortality rate are prematurity, low birth weight, home deliveries conducted by traditional birth attendants, lack of antenatal diagnosis, poor referral system, lack of pediatric surgical and/or pediatric anaesthetist coverage and delayed presentation.[6]

Keeping in view the importance of subject, this study was conducted with an objective to prospectively evaluate the management and outcome of esopahgeal atresia at Children Hospital, Pakistan Institute of Medical Sciences (PIMS), Islamabad.

Methodology

This is a Observational-descriptive study in which consecutive cases of esophageal atresia presenting to Children Hospital PIMS, from October 2017 to August 2018, were included. All the cases were admitted through emergency.

WHO calculator was used for sample size calculation in which the population proportion was 25% & precision was taken as 5%, with confidence level of 95%.[3]

Patients with esophageal atresia admitted in surgical ward were included and those admitted in Neonatal Intensive Care Unit were excluded.

Parameters studied include age at presentation, gender, gestational age, mode and place of delivery, weight, associated congenital anomalies, type of esophageal atresia, type of operation, post-operative complications and outcome as survival or mortality.

Depending on availability of resources standard management was given to all the patients. Resuscitation was done in those who were sick at presentation. Haematological laboratory tests included total leukocyte count, haemoglobin, platelet count and coagulation profile. Biochemical tests included urea, creatinine, random blood sugar and serum electrolytes. Radiological investigations including X-ray chest and abdomen after passing gastrograffin impregnated nasogastric tube which showed level of curling of NG indirectly indicating level of upper pouch and presence/ absence of TEF (Air in abdomen), X-ray whole spine to rule out any vertebral anomalies, Ultrasound of renal system was done in babies who were stable enough to be sent to radiology suite.

Management of neonates started with initial stabilization by nil per os (NPO), intravenous fluid resuscitation, broad spectrum antibiotics, intermittent deep throat suctioning, nursing in prop up position and nebulisation. Oxygen inhalation was started to those who were not maintaining saturation. Transfusion of blood products was done in those who were having haematological deficiencies.

Neonates who were stable after initial resuscitation underwent surgery. All those having type A esophageal atresia underwent cervical esophagostomy and feeding gastrostomy. All those with type C esophageal atresia had right thoracotomy via standard posterolateral, extra pleural approach. TEF identified, transfixed and divided. Azygos vein was preserved in majority of the case. Azygos vein ligation was done in those cases where there was difficulty in exposure of esophagus. Primary end to end anastomosis was done in those patients who had tension free upper and lower ends of esophagus after fistula division. Trans-anastomotic feeding tube was passed in cases of primary repair. More than 3 vertebra between upper and lower end of espophagus was considered as a long gap and cervical esophagostomy and feeding gastrostomy was done after fistula ligation in such cases. Chest drain was placed closed to primary anastomosis. Neonates were managed in High Dependency Area (HDA) of the ward. Apparently standard postoperative management protocol was followed in all cases. Feeding through trans-anastomotic tube stared on 2nd post operative day. In stable patients tolerating NG feed and no signs of anastomotic leakage (no saliva in chest drain or normal contrast esophagogram), trial for oral feed was given at 6th post operative day. Some children also had contrast esophagogram at 7-8 postoperative days. Rest of management was individualized according to condition of the patient. Patients who were stable, tolerating orally and on demand feed were discharged. Survival was considered as good outcome. Mortality was considered as poor outcome.

Data analysis was done using SPSS version 21. Descriptive statistics like mean and standard deviation for quantitative data and frequency and percentages for qualitative data were calculated. Chi-square test was applied to associate different parameters with surgical outcome in terms of discharge and died. *p*-value <0.05 was considered significant.

A written informed consent was taken from parents/guardians and the study was approved by institutional ethical review board (Ref. no. F.1-I/2015/ERB/SZABMU/181)

Results

Total 140 patients were included in this study. Seventy nine (56.4%) were males and 61 (43.6%) were females. Mean age at presentation was 5.5 days with minimum age of 1 day and maximum age of 30 days details of which are given in Table-1.

Table 1: Age of the neonates at the time of presentation.

Day of Life at Presentation	*Frequency*	*%*
Within 24 hours	23	16.42
24-48 hours	19	13.57
48-72 hours	24	17.14
72-96 hours	20	14.28
After 96-120 hours	11	7.85
Between 6-10 days	16	18.57
Between 11-15 days	13	9.28
Between 16-30 days	4	2.85
Total	140	100

Twenty (14.3%) patients were born preterm and 120 (85.7%) were born at term. Anomaly scan was not done in 36 (25.7%) cases. In 99 (70.7%) cases, anomaly scan was normal or polyhydramnios was the only finding. Only 4 (2.9%) cases had confirmation of esophageal atresia on anomaly scan. Out of 140 cases 118 (84.3%) were delivered in hospital and 22 (15.7%) neonates were delivered at home. Out of 118 hospital deliveries 80 were born by normal vaginal delivery and 38 were born by caesarean section. Minimum weight at presentation was 1 kg and maximum was 4.20±0.464 kg with mean weight of 2.43kg. Thirty (21.4%) neonates died during initial stabilization period, without undergoing any surgery. Out of 30 who died during initial stabilization period, 11 died due to sepsis, 9 had pneumonia, 4 had both sepsis and pneumonia and 7 died due to multiple congenital anomalies. Neonates were screened for associated anomalies details of which are shown in Table-2.

Regarding type of esophageal atresia, 10 (7.1%) patients had type A, 97 (69.3%) had type C, 1 (0.7%) had type E and 1 (0.7%) had type F esophageal atresia. In 30 patients, type could not be assessed as they died during initial stabilization period before surgery and two patients left against medical advice.

Nine patients with type A underwent cervical esophagostomy along with feeding gastrostomy and one died before surgery. Right thoracotomy was performed in 98 cases (97 type C; 1 type E). Out of 97 patients of type C atresia, end to end esophageal anastomosis was possible in 76 patients and in 21 patients end to end anastomosis

Table 2: Associated anomalies with enrolled patients of esophageal atresia.

Vertebral Frequency	*Gastrointestinal Frequency*	*Cardiac Frequency*	*Genitourinary Frequency*	*Limbs Frequency*	*TEF** Frequency*
Butterfly vertebra (2)	ARM* with recto-vasical fistula (1)	ASD* (8)	Renal agenesis (3)	Bilateral radial ray defect (3)	Present (131)
Bifid vertebra (2)	Anteriorly placed anus (2)	VSD* (4)	Hydronephrosis (3)	Unilateral radial ray defect (1)	Absent (9)
Hemivertebra (5)	ARM with bucket handle deformity (1)	ASD=VSD (1)	Small kidney (2)		
Coronal cleft vertebra (16)	ARM with RVF* (8)	Complete AVSD* (1)	Ectopic kidney (1)		
	ARM without Fistula (10)	PDA* (2)	Horseshoe kidney (1)		
	ARM + Duodenal alresia (1)	ASD + PDA* (2)	PPV* (1)		
	ARM + Malrotation of gul (1)	PDA + VSD (5)	Clitoromegaly (1)		
	Common cloaca (1)	Right Aortic arch (1)	Hypospadias (2)		
	Duodenal atresia (2)	Pentalogy of fallot (1)	Posterior urethral valves (1)		
	Meckel's divertivulum (1)		Undescended testis (3)		

* ARM = Anorectal malformation, RVF = Rectovestibular fistula, ASD = Atrial septal defect, VSD = Ventricular septal defect, AVSD = Atrio-ventricular septal defect, PDA = Patent ductus arteriosis, PPV = Patent processus vaginalis, **TEF = Ttacheo-esophageal fistula

was not possible due to long gap between two ends, so cervical esophagostomy and feeding gastrostomy was performed. TEF was ligated in 97 type A and one Type E patients. Azygos vein was preserved in 78 cases and in 22 cases it was ligated to attain better exposure.

Mean duration of nil per os was 4.64 days. Contrast esophagogram was done in 21 cases, between 5 to 12 days post operatively with mean duration of 7.71 days.

Sepsis was the main complication post operatively (44 paitents) followed by pneumonia, Anastomotic leak and surgical site infection (Table-3). Nine out of 11 patients with anastomotic leak were managed conservatively and 2 patients had to undergo cervical esophagostomy and feeding gastrostomy.

Table 3: Post-operative complications in neonates suffering from esophageal atresia.

Post on Complications	*Frequency*	*%*
Surgical site infection	3	2.14
Sepsis	41	29.28
Chest infection	20	14.28
Anastamotic leak	11	7.56

Overall mortality was 57.9% (81/140) out of which 21.4% (30) died before surgery and 36.5% (51) died post operatively.

Table 4: Correlation of various variables with outcome.

Variables	*Survival*	*Mortality*	*p value*
Post of complications	29	46	*0.002
Weight less than 2.5 kg	16	42	*0.004
Pre-term	4	16	0.15
Full term	53	65	0.15
Azygos vein ligation	14	8	0.104

*p value <0.05 is significant

There was statistically significant relationship between post operative complications and poor outcome (p =0.002). Post operative sepsis, chest infection and anastomotic leak being common contributors in post operative mortality, 65.8% (27/41) patients having post operative sepsis, 60% (12/20) patients with post operative chest infection and 63.6% (7/11) patients with post operative anastomotic leak died. No significant relationship was found when outcome is compared to age at presentation (p =0.69), preterm delivery (p =0.15), gender (p =0.25), additional surgery for associated anomalies (p =0.88), birth to operative time (p =0.53), type of atresia (p =0.06), Azygos vein preservation (p =0.10) (Table-4).

Significant relationship was found when outcome was compared to weight at presentation (p =0.004).Outcome was poor in patients with weight less than 2.5 kg at presentation. There was also a significant relationship to good outcome in those patients who primarily underwent cervical esophagostomy and feeding gastrostomy (p =0.019). Results show that 70% of patient survived post operatively who underwent cervical esophagostomy and feeding gastrostomy as primary procedure.

Discussion

Since first primary repair in EA done by Cameron Haight in 1941, there is a remarkable improvement in treatment and outcome. The survival rates of esophageal atresia are increasing over last few decades and in developed countries survival rates are 85% to 95% in cases where esophageal atresia is not associated with severe congenital anomalies. The improvement in survival rates are attributed to early diagnosis, better understanding of the disease and neonatal physiology, better Neonatal Intensive Care, parenteral nutrition, antibiotics, advances in neonatal anaesthesia and improved surgical techniques. In developing countries, however, survival rates of EA with or without TEF are not that much high. Lack of adequate and proper medical facilities in developing countries hamper the standard management of EA patients, especially those with prematurity, low birth weight and severe congenital anomalies, resulting into increased morbidity and poor outcome,[4,5,7] this is what our study also indicates.

EA is difficult to diagnose on antenatal scans and less than half of cases are diagnosed antenatally.[8] But in our study only 4 (2.9%) patients had prenatal diagnosis of EA. This may be related to experience of sonologist/ radiologist. Moreover, most of our patients came from far flung areas where there is no availability of expert and experienced sonologist/ radiologist. In a study.[9] Prenatal diagnosis rate of EA was 18.1% and they concluded that prenatal diagnosis does not modify neonatal management and one year outcome.[9] In our study as well, antenatal diagnosis of EA did not alter the outcome. Although having prenatal diagnosis, all 4 cases were not delivered at tertiary care hospital where pediatric surgery department is available and presented to us late. In order to prevent aspiration pneumonia, prompt diagnosis of esophageal atresia before first feed is necessary.

Incidence of EA is more common in males as compared to females in our study. This male predominance is elicited in various studies.[7,8,10-12]

Surgical management of esophageal atresia varies across the institutions.[13] As many aspects of EA management are still controversial. A survey from 45 countries across the world was conducted which include questions regarding pre, per and post operative management.[5] According to them 81% surgeons would request pre operative echo but only 56% went for left thoracotomy in case of right aortic arch. In our institution echocardiogram is not routinely requested and we always approach via right thoracotomy. Open thoracotomy with extrapleural approach was preferred by most of surgeons which are similar to our institution.[14] Different methods are applied for identification of different types of EA/TEF and fistula location.[15] A reterspective study[11] in order to find out common peri-operative practices in management of EA/TEF, authors collected data from 11 different hospital over 5 years and found wide variations across the centres without any standardized and evidence based best practice. Peri-operative bronchoscopy in order to localize fistula ranged from 0-100%, proximal pouch contrast during surgery ranged from 0-69%, placement of ranged from 21-100% trans-anastomotic tube and thoracoscopic repair ranged from 0-21%.[11] In our institution, it is our practice to place trans-anastomotic tube in all primary repairs but we do not use rest of methods.

Outcome of EA is good in developed countries with survival rate of 85-95% in cases where EA does not have associated severe congenital anomalies.[4] In a study[4] authors analyzed prognostic factors in 60 patients who were treated for EA in Mother and Children Health Institute Serbia over 10 years. In their study mortality was 28.3%. According to them birth weight, mean apgar score value, prematurity, birth-operative time >24 hours and post operative sepsis were significantly associated with mortality.[4] In our study as well low birth weight and post operative complications were significantly associated with poor outcome. They also analyzed type of EA as prognostic factor and concluded that type of EA does not have influence on mortality. In our study type of esophageal atresia has not significant influence on mortality corresponding to various studies in literature.[4,16-18]

In our study, overall mortality was very high when compared with data of developed countries.[11,12] However, several regional studies such as one in Iran[19] showed mortality rates of 56%, which are similar to our study. Authors found that major contributors of mortality in their patients were late operation, prematurity, congenital malformations, low birth weight and sepisis.[19] In our study low birth weight and post operative sepsis were significantly related to mortality. This finding is in line with results of another local study in which overall mortality was comparable to our study i.e. 58%.[3] Several factors contribute to high mortality in our study. Lack of Neonatal Intensive Care Unit (NICU) due to limited space, non availability of dedicated and specialized paramedical staff round the clock, low doctor to patient ratio, non availability of neonatal echocardiography/ pediatric cardiologist for screening and management of associated cardiac anomalies and non availability of ventilator support for most of the neonates are some of the limiting factors which affect overall outcome in patients with EA. Survival can be improved by overcoming these limitations. Another study conducted at National Institute of Child Health, Karachi, showed pre-operative mortality of 13% and post operative mortality was 28% with post-operative sepsis being the major cause of mortaliy.[6]

Low birth weight was associated with poor outcome in our study. Organising Nutritional education programmes for pregnant ladies by public health specialists and leady health workers can help in overcoming low birth weight thus making better outcome possible in case of EA.

Limitation in our study is small sample size. Large sample size may better correlate various variables such as late presentation, prematurity, additional surgery for associated anomalies, birth to operative time and Azygos vein preservation with outcome. Further studies with larger sample size including factors associated with health care delivery system are required to better delineate the correlation of these variables with outcome.

As compared to developed countries, survival of EA is low in developing countries. Survival can be improved by evaluation of factors contributing to sepsis and overcoming them, improving surgical techniques, provision of adequate and well trained nurses, continuous suction of saliva to prevent aspiration pneumonia, prevention of hypothermia and availability of neonatal intensive care. Hence, for overall improvement in survival of EA betterment in entire health care delivery system is the dire need of our country.

Conflict of interest: None declared.

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