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Esophageal atresia - Early outcome analysis from a high-volume tertiary care institute in India

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Abstract

EA with or without TEF remains the most common congenital anomaly of the esophagus. Although it is a relative rare condition, this complex anomaly is still a challenging problem in pediatric surgery. In developing countries, many infants that present with EA with TEF exhibit pneumonitis due to late referrals, along with having a low birth weight. The concept of ligation of the TEF and anastomosis of the two ends of the esophagus in EA was proposed by Richter in 1913. First survivors following staged repair with later esophageal replacement were reported by Leven and Ladd in 1939.[5] Cameron Haight is credited with first (1941) successful primary anastomosis of the esophageal segments.[6] With advancement in early diagnoses and neonatal care, there has been a marked improvement in survival. Delayed diagnosis, late presentation, septicemia, preceding pneumonia, congestive heart failure, and overcrowding in ICU were the main causes of poor outcome in our series. Wound-related complications and pneumonia were the major causes of morbidity.

Keywords: Esophageal atresia, tracheoesophageal fistula, anastomotic stricture

Introduction

Esophageal atresia (EA) with or without trachea esophageal fistula (TEF) remains the most common congenital anomaly of the esophagus. Although it is a relatively rare condition, this complex anomaly is still a challenging problem in pediatric surgery. In developing countries, many infants that present with EA with TEF exhibit pneumonia and respiratory distress due to late referrals and having a low birth weight [1]. The overall incidence of EA-TEF ranges from one in every 2500 to 4500 live births [2]. An improvement in survival has been observed in recent decades. This finding is likely multifactorial and largely attributable to advances in

neonatal intensive care, neonatal anesthesia, ventilatory and nutritional support, antibiotics, surgical materials and techniques [3]. Conditions such as prematurity, low birth weight, associated major anomalies compounded with delayed diagnosis of this entity, and scarcity of resources may negatively influence the successful outcome. Herein, we aim to study the early outcome analysis of patients with esophageal atresia (EA) and tracheoesophageal fistula (TEF) at a high-volume tertiary care institute. We are also describing our experience, evaluate the risk factors and measures for improving the outcomes in the future.

Subjects and methods

All neonates with EA and TEF admitted in the Neonatal Intensive Care Unit (NICU) of the department of paediatric surgery over 1 year from January 2021 to December 2021 were retrospectively studied. Patients transferred from paediatric medicine department and other hospitals with suspected EA/TEF and whose diagnosis was later confirmed were also included. The clinical, operative records and other demographic details of patients of EA were analysed along with comorbidities for next 3 months. Charts were reviewed according to age at presentation, sex, antenatal diagnosis, chief complaints, and preoperative investigations undertaken for the establishment of diagnosis, associated anomalies, operative procedure, intra operative findings, pos to perative complications, and treatment outcomes. Classification of EA was done according to Gross's anatomic classification. Various parameters were reviewed to analyse the risk factors associated with the morbidity and mortality. As the resources were restricted (unavailability of bedside echocardiography), all the patients were not subjected to echo for complete cardiac evaluation. The patients who were clinically stable and having either (a) clinical features suggestive of cardiac anomalies (cyanosis/cardiac murmur/overload) or (b) radiographic features, were subjected to echo.

VACTERL association was defined as (a) if the patient has 3 or more anomalies of the vertebral, anorectal, cardiac (excluding patent ductus arteriosus and patent foramen ovale), renal/genitourinary, and limb systems including EA. and (b) If the patient had both the "core" features, that is, anorectal malformation (ARM) and TEF [4]. There should be the absence of any clear evidence for an alternate, overlapping diagnosis. The diagnosis of EA was confirmed by radiographs (both anteroposterior [AP] and lateral views) with red rubber catheter *in situ*. Chest drain was inserted in all patients with EA undergoing thoracotomy. Intravenous fluids, antibiotics, and parenteral nutrition were continued postoperatively. Before starting feeds, a contrast study was undertaken on the 7th postoperative day. Chest drain was removed after a 7th postoperative day when there were no signs of anastomotic leak. Thereafter, gradual feeds were initiated. Routine placement of nasogastric (NG) tube was not performed for feeding purposes (as per institutional protocol). NG tube placement was done in cases with long gap with anastomosis under tension.

Statistical analysis used

Data were coded and recorded in MS Excel spreadsheet program. SPSS v23 (IBM Corp.) was used for data analysis. Descriptive statistics were elaborated in the form of means ± standard deviations and medians or IQRs for continuous variables, and frequencies and percentages for categorical variables.

Results

There were 350 new cases of EA/TEF. 205 (56.74%) males and 145 (41.42%) females with male: female ratio of 1.41:1. Type C was the most common (90.57%), followed by Type A (5.7%) and Type D (0.28%) as shown in Table 1. No case of EA with proximal TEF

(Type B) and type E were identified. Out of 350 admissions, 318 (91.63%) were admitted after being referred from peripheral centres or other institutions, while 32 (9%) neonates were from our institution. Among all the admitted patients nearly two third (234, 67%) cases were admitted in our institution on the 1st day of life [Table 2]. Rest of the patients had delayed diagnoses and presented beyond 24 hours of life ranging from 2 to 10 days.

The weight of the neonates ranged from 900 to 3500 grams and out of which 213 (60.85%) patients were low birth weight. Weight-wise distribution of patients and their survival is summarized in Table 3. The overall survival was 54.86%. Neonate with lowest birth weight who survived in our series weighed 1400 g. Maximum cases (107, 30.57%) were admitted between July and September, with a peak in July (31, 8.85%) patients. Lowest number of cases (50, 14.28%) were seen in February and March.

Associated major anomalies were present in 6 (30 %) cases with Type A and 16 (5%) cases in Type C as shown in Table 1. The most commonly affected organ system associated with EA was gastrointestinal (GI) tract with 15 cases. Anorectal malformation (ARM) was seen in 12 cases (Low - 5 cases, high – 7 cases). Cloacal anomaly was seen in 1 patient and vestibular fistula was seen in 3 patients. ARM was followed by duodenal atresia (DA) which was present in 4 cases. In one of the case, DA was associated with multiple intestinal atresia. Cleft lip/palate anomaly and absent radius were present in three patients each. Other malformations such as severe vertebral abnormalities were seen in two cases.

Surgical procedures were performed in 333 patients (Table 4). In rest of the 17 patients, 4 patients did not undergo definite surgery as the parent's declined further treatment and 13 patients died before any intervention (preoperatively). All procedures were performed between 24 and 48 h following diagnoses. Kimura's duo denoduodenos to my was performed in four patients. Left transverse colostomy for high-type ARM was performed in 8 cases. In all cases with vestibular fistula (3), the colostomy was deferred as primary repair is being performed for these at three months of age. Low-type anomalies were managed by anoplasty in one case. Out of those who survived three patients were reoperated in the same admission for major anastomotic leak and esophagostomy and gastrostomy was performed. In these diverted patients two patients survived.

The overall survival was 55% with a high mortality rate of 45%. The survival was better in Type C than Type A and also, in those without associated malformations (Type C: 31.96% vs. 20.83% and Type A: 28.57% vs. 25%). Out of those 158 neonates who died, 7 patients died before operative intervention. Mortality was highest (112, 70.88%) in 24 to 72 hours postoperatively, followed by 25 (18.98%) patients who died between 4th and 7th post-operative days, while 14 (8.86%) succumbed beyond 1-week postoperatively. The most common etiologic factor was septicaemia (84, 53.16%) followed by respiratory failure (severe pneumonia) in 52 (32.91%) patients, congestive heart failure in 18 (11.39%) and major anastomotic leak was seen in 4(2.53%) patients.

The mean hospital stay for patients with favourable outcome was 12 ±5.2 days. These patients were closely followed up to 3 months. There were 20 readmissions with pneumonia (12) being the major reason for readmission followed by anastomotic stricture (4), septicaemia (2), tracheomalacia (1) and minor leak (1). No case of recurrent TEF was reported.

Out of four patients who demonstrated stricture within 3 months of follow-up post-surgery, three responded to endoscopic esophageal dilatation while in one patient

esophagostomy and gastrostomy was done as it was a tight stricture not able to negotiate even a guidewire. These cases were associated with long-gap (>3 cm) EA. Patients with esophagostomy and gastrostomy are under follow-up for definitive surgery.

Discussion

On reviewing the literature, it is evident that the concept of ligation of the TEF and anastomosis of the two ends of the esophagus in EA was proposed by Richter in 1913. First survivors following staged repair with later esophageal replacement were reported by Leven and Ladd in 1939 [5]. Cameron Haight is credited with first (1941) successful primary anastomosis of the esophageal segments [6]. With advancement in early diagnoses and neonatal care, there has been a marked improvement in survival of these neonates. In our cohort, the presentation was similar to that described in the literature as, drooling of saliva/ frothing from the oral cavity, choking or transient cyanosis shortly after birth, vomiting or regurgitation after attempted feeds, and difficulty in breathing and respiratory distress [6]. With the delay in diagnosis pulmonary symptoms usually develop by the 2nd day and patient may present with pneumonia (attributed to the respiratory infection). Pneumonia was the second most common, but primary preventable cause of death in our study. Pneumonia occurs due to the aspiration of the salivary secretions from the upper esophageal pouch and reflux of the gastric secretions through the distal fistula. EA is confirmed by passing no. 10 sterile, blunt-tipped red rubber catheter into the esophagus. Failure to pass beyond 10 cm (usually) or failure to negotiate into the stomach (occasionally) is considered as diagnostic of EA. In any neonate with respiratory distress, EA should be ruled out as it might have been missed earlier. In addition, it is used to evaluate for the level of upper pouch in radiograph, facilitating identification and dissection of the upper pouch during primary repair. Use of infant feeding tube must be discouraged as it may be associated with false-negative results due to accidental trans tracheal gastric intubation through the TEF or passage up to a significant length in the elongated upper esophageal pouch [6-8].In our series, 29% neonates were not diagnosed at the time of birth or in the first day of life which had pneumonia leading to dismal outcomes and poor survival. In those neonates who have been diagnosed in the first 24 hours of life, low favourable outcomes could be attributed to associated complex congenital malformations. Birth weight has a tremendous impact on the outcome and has been substantiated by different authors from Asia. Low birth weight was reported as 36.1% by Chang et al [9], while higher number of our patients (64.65%) low birth weight. Patients weighing <2000 g at birth had a significantly lower survival rate (26.47%) compared with patients weighing at least 2500 g at birth. The favourable outcome was best (78.57%) with neonates weighing ≥3000 g in our series. Thus, low birth weight was one of the major contributors for poor survival in our study.The incidence of Type A was 5.7% in our series, which is similar to reports presented by various authors. Incidence of Type A is described as 7% according to one study and may range from 2% to 15%. Type C was 90% in our study and this corroborate with the Asian studies [9-10].

Associated major anomalies were present in 35% cases, which correlates with the available literature. The incidence of anomalies associated with EA/TEF range between 30% and 60%. Among all the associations, cardiovascular anomalies are the most common (1/3rd cases), ranging from 30% to 59.7% with the Tetralogy of Fallot and ventricular septal defect being more common. Others are septal defects, pulmonary atresia/pulmonary

stenosis, aortic stenosis, and dextrocardia [11]. VACTERL association refers to the non-random co-occurrence of vertebral anomalies (V), anal atresia (A), congenital heart defects (C), TEF, renal anomalies (R), and limb defects (L). VACTERL association was present in 21% patients in our study which is in accordance with other studies [9].

After the cardiovascular anomalies, the other associations are GI anomalies (24%), followed by genitourinary (20%), orthopaedic, and other miscellaneous anomalies. GI anomalies were reported as 26.67% in one large series, but in our series, the total percentage was lower (13.49%) than reported [11]. ARM was also the most common anomaly (11.86%) among the GI group, as seen in other studies also [11-12].

Surgery in EA should ideally be performed within the first 24 hours, as delay in surgical correction increases the risk of aspiration pneumonia, as discussed earlier. Operative approach for EA with TEF is classical right postero-lateral thoracotomy with the extra pleural approach [5-6]. The gap length of more than 3 cm was associated with high mortality rate. It has been more frequently observed in patients with aberrant vessels and is associated with high morbidity [9-10].Among the postoperative complications mentioned in the literature, anastomotic stricture is the most common and may be present in 30%–40% cases. Other major complications are anastomotic leak (15%–20%), GER (40%), recurrence of TEF (5%–14%), tracheomalacia (10%), recurrent pneumonia, and hiatal hernia [5, 9-12]. Wound-related complications and pneumonia were the major causes of morbidity in our study. Gastro esophageal reflux was seen in 3 patients among 59 cases which were in follow-up; all were treated conservatively. Factors influencing the outcome were birth weight, presence of pneumonia, associated malformations/ anomalies, type of atresia, and long gap [10]. Although the delayed presentation is associated with poor outcome, there are reports of survival with diagnosis as late as 17th day of life [14]. In our series, one patient (Type C) survived who was diagnosed on 15th day of life. In West, the survival of full-term infants with EA without associated anomaly has increased above 95% and birth weight >1500 g with no major cardiac anomaly was 98.5%. The overall mortality rates range from 10% to 30%.[15] Mortality in our series was high (45%) as compared to Western literature and also more than other centres in Asia [5, 9-10]. The most common cause of mortality in our patients was septicaemia, which was also seen in one of the large series available [10]. Among the major congenital anomalies responsible for mortality in EA, almost 6%–11% of have trisomy or complex cardiac defects which are incompatible with life, thereby precluding any active management. Furthermore, infants with major and complex cardiac anomalies have 30% and 70% risk of mortality, respectively. VACTERL association has mortality in 1/5th cases mainly due to complex cardiac anomalies [12]. In our study, mortality was present in 4/5th of patients having VACTERL associations. Respiratory complications were reported as the most common etiologic factor responsible for mortality in one study [16]. In our series, 2.56% patients died before any intervention could be contemplated owing to complex cardiac anomalies and/or pneumonia. Early diagnosis, prevention of pneumonia, adequate preoperative resuscitation, ventilator management, postoperative care in neonates undergoing major surgery is invaluable for the outcome of surgery. We suggest following recommendations for improving the favourable outcomes:

- Improvement of ultrasonography facilities for antenatal diagnoses of EA and associated malformations

for planned institutional delivery and timely diagnosis. EA must be ruled out in all new borns just after the birth by putting no. 10 sterile, blunt-tipped soft red rubber catheter into the esophagus to prevent the undesired complications associated with delayed diagnosis

- Prevention of aspiration of salivary secretions which may lead to wet lungs and thereby pneumonia is paramount. For this purpose, Replogle tube or continuous low-pressure mucus suction apparatus preoperatively and during transport is mandatory [17].
- Evaluation of associated malformations, particularly cardiac anomalies with the help of bedside echo facilities. Urological assessment with bedside ultrasonography facilities for increasing the yield of VACTERL association.
- Round the clock neonatal care under a neonatologist. Thorough improvement in the nursing care and staffing pattern by imparting advanced training and supervision. Patient-to-nurse ratio must be 1:1 from the present situation of 10:1.
- Provisions for genetic studies of patients, especially those with VACTERL association.

Our institution is catering a very large number of EA cases every year. Furthermore, the total cases managed per year are probably one of the largest in the world, to the best of our knowledge. This is a descriptive analysis of all the patients of EA with associated anomalies and the procedures performed along with mortality and morbidity data.

Conclusion

In our study, Type C was the most common and approximately one third cases were LBW. Associated major anomalies were documented in approximately one fifth patients with GI anomalies in 13.49% cases and VACTERL association in 11.63%. Survival was 55%, and it was better in EA without associated anomalies and in Type C than Type A. VACTERL association had a very high (1/5th) mortality rate. Overall survival of neonates with EA is still poor in our high-volume tertiary care centre. Delayed diagnosis, late presentation, septicaemia, preceding pneumonia, congestive heart failure, and overcrowding in ICU were the main causes of poor outcome in our series. Wound-related complications and pneumonia were the major causes of morbidity.

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Legend Tables

Types of esophageal atresia and associated anomalies		
Types of atresia	Frequency	Cases with associated anomalies
Type A (pure EA)	20	6
Type B(proximal tef with distal EA)	0	0
Type C(EA,distal TEF)	317	16
Type D(proximal and distal TEF)	1	0
Type E(TEF)	0	0
Esophageal web	1	0
Esophageal stenosis	1	0
Total	350	

Table 1

Outcomes of esophageal atresia as per the day of admission in institution		
Day of admission in institution	Number of patients	Survival
1	234	141
2	104	46
3	8	4
>4	4	1

Table 2

Outcome of esophageal atresia as per birth weight		
Birth weight (g)	Number of patients	Survival
<1500	12	4
1500 – <2000	56	14
2000 – <2500	145	73
2500 – <3000	109	79
>3000	28	22

Table 3

Summary of procedures undertaken for various types of esophageal		
Type of atresia	Number of patients undertaken for surgery	Types of surgical procedures
Type A (20)	18	Esophagostomy with gastrostomy
Type C (317)	312	Right thoracotomy with fistula ligation and end to end esophageal anastomosis
Type D (1)	1	Right thoracotomy with both fistula ligation and end to end esophageal anastomosis
Esophageal web (1)	1	Right thoracotomy with excision of web and end to end esophageal anastomosis
Esophageal stenosis(1)	1	Right thoracotomy with excision of stenosed segment and end to end esophageal anastomosis

Table 4