

Original Research Article

Survival outcomes of trachea esophageal fistula in infant

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ABSTRACT

Background: Tracheo-esophageal fistula (TEF) is a rare congenital abnormality often associated with several other anomalies including renal, vertebral column, gastrointestinal or cardiovascular defects. This study was carried out to evaluate the outcome of trachea esophageal fistula among patients who underwent various surgeries for the anastomosis of trachea esophageal fistula.

Methods: This study was conducted as a record based cross sectional study among 88 patients who were diagnosed and treated for trachea-esophageal fistula in tertiary care hospital between 2015 and 2018. Data regarding the type of anomaly, presence of associated anomalies, type of surgery and outcomes were documented. Findings of echocardiography and ultrasonography were also documented. Data was analyzed using SPSS software. Chi square test was used to evaluate the outcome of the surgical procedures for management of TEF.

Results: Majority of the participants in our study belong to <1 month of age and were males (56.8%). Type 3 tracheo esophageal fistula (80.7%) was the most common type. Associated cardiovascular anomalies were present in 50% of the participants. Thoracotomy with TEF repair was most preferred surgery (76%). Present study demonstrated that surgical techniques improve the physical and physiological outcome of the patients ($p < 0.05$).

Conclusions: Trachea esophageal fistula needs to be corrected with surgical procedure. Modern techniques like thoroscopic anastomosis, thoroscopic techniques to achieve an anastomosis can also be explored. Future studies may be directed in detecting congenital anomalies during the pre-natal period with the help of genetic techniques.

Keywords: Anastomotic leak, Esophageal atresia, Tracheo esophageal fistula, Thoracotomy

INTRODUCTION

Tracheo esophageal fistula (TEF) is a common congenital deformity identified by abnormal connection between esophagus and trachea and is the most common type of airway fistula. The common symptoms of trachea esophageal fistula (TEF) include feeding difficulties, choking, frothing, following food intake, severe coughing and unmanageable respiratory infections.¹ TEF commonly occurs in approximately 1:3,500 live born infants. There are five sub types attributed to the occurrence of TEF based on the location of the atresia and types of connection between trachea and esophagus.

In majority of the patients there are other associated anomalies which involve the vertebral column, gastrointestinal tract, cardiovascular system, renal and limb abnormalities. There is an increased need for improved surgical techniques and both pre and postoperative care for recovery from TEF.^{2,3} Trachea esophageal fistula is a multifactorial complex disease involving several genetic and environmental factors.

In 6%-10% of the patients there is a clear underlying genetic defect which has been diagnosed, however the remaining 90% of the patients the etiology is often unknown.⁴ Some of the proposed risk factors include

environmental factors like maternal smoking and alcohol, infection diseases during pregnancy, maternal exposures including drugs like methimazole and exogenous sex hormones. Certain studies have shown that maternal and in utero exposure to diethyl stilbesterol could also be a risk factor for TEF.

However, some of the recent trials in Europe have found that increase in the maternal age could increase the risk for TEF.⁵ Since majority of the TEF also present with several other congenital anomalies there could be genetic defect linked to the incidence of TEF. Some of the genetic disorders which are linked to TEF include single gene disorders like feingold syndrome, Charge syndrome, AEG syndrome, Pallister hall syndrome, to name a few. Most of the genetic abnormalities are linked to mutations in MYCN gene, CHD-7 gene, GLI-3 gene, etc.⁵

METHODS

This study was carried out to determine the survival outcome of patients diagnosed with trachea esophageal fistula.

Methodology

This was a cross-sectional record based study was carried out in the Department of Pediatric Surgery of our tertiary care hospital for a period of four years between 2015 and 2018.

All the patients who were admitted in our tertiary care hospital with a diagnosis of TEF during the study period were taken up for the study. A total of 88 patients were included in the study. The participants were selected by convenient sampling.

Ethical approval and informed consent

Approval was obtained from the Institutional Ethics Committee prior to the commencement of the study.

Data collection

Data was elicited from the hospital records. Data pertaining to the antenatal period, ultrasound findings, echocardiography findings and the type of TEF were recorded. Tracheo esophageal fistula was graded as type 1, type 2, type 3 based on the level and type of the fistula. Particulars related to the type of surgery and requirement of ventilator support were also documented. Data pertaining to the outcome of surgery and hospitalization was also recorded.

Data was entered in SPSS version 15 software. The survival outcomes of TEF repair was expressed in percentages. Chi square test was used to compare the association between the outcome of the surgery and the type of surgery. A p value <0.05 was considered as the statistically significant.

RESULTS

The cross sectional record based study was carried out among 88 patients who were diagnosed with TEF. Majority of the participants were newborns within five days of age (93.2%). Also, females were about 38(43.2%) and males were 50(56.8%) (Table 1).

Table 1: Background characteristics of the study participants.

Characteristics	Frequency (N=88)	%
Age (in days)		
<5	82	93.2
5-10	5	5.7
>10	1	1.1
Sex		
Female	38	43.2
Male	50	56.8

Table 2: Anomaly profile of the participants.

Characteristics	Frequency (N=88)	%
Type of TOF		
Type 1	12	13.6
Type 2	3	3.4
Type 3	71	80.7
Others	2	2.3
Echocardiogram findings		
Normal	43	48.9
Atrial septal defect	8	9.1
Ventricular septal defect	25	28.4
Patent Ductus arteriosus	8	9.1
Tetrology of fallot	2	2.3
Mitral Stenosis	1	1.1
Echocardiogram not done	1	1.1
Ultrasound findings		
Normal	47	53.4
Anomalies		
Renal	27	30.7
Liver	3	3.4
Pelvic	11	12.5
Associated anomalies		
Present	44	50.0
Absent	44	50.0
Antenatal ultrasound		
Done	43	48.9
Not done	45	51.1

The anomaly profile of the study participants showed that majority of the participants belonged to Type 3 TEF (80.7%). Echocardiography findings showed that 28.4% of the participants had ventricular septal defect. Moreover, 30.7% of the participants had congenital renal anomalies detected by ultrasoundography, in addition to TEF (Table 2). Majority of the participants underwent surgery for TEF (90.9%) of which thoracotomy with TEF repair was the most common type of surgery (71.6%) of the study participants.

Table 3: Particulars related to the management of TOF.

Characteristics	Frequency (N=88)	%
Procedure		
Thoracotomy+TEF repair	63	71.6
TEF ligation	4	4.5
Esphagostomy+gastrostomy	7	8.0
Moris procedure	4	4.5
Anterolateral stent	2	2.4
Not done	8	9.0
Ventilation		
Yes	51	58.0
No	37	42.0
If yes ventilation (51)*		
<24hrs	17	33.3
24-72hrs	21	41.2
>72hrs	13	25.5
Pre op ventilation		
Yes	32	36.4
No	56	63.6
If yes Preopventilation (32)*		
≤ 24hrs	19	59.4
24-72hrs	11	34.3
>72 hrs	2	6.3
Day of surgery		
≤ 24hrs	51	58.0
24-72hrs	32	36.4
>72 hrs	5	5.6

Ventilatory support was required for 58% of the participants, and pre-operative ventilation was required in 36.4% of the patients. Majority of the participants

(58.0%) underwent surgery ≤48hrs of birth (Table 3). The outcome of surgery and hospitalization was documented. It was observed that majority of the participants improved in terms of respiration and feeding (79.5%).

About 2.3% of the participants had anastomotic leak which was managed conservatively while 15.9% of the participants expired during the hospital stay (Table 4). The association between surgery and outcome of hospitalization showed that surgery significantly improved the outcomes in the patients with TEF.

Patients' condition improved significantly (82.9%) when they underwent surgery compared to those who did not undergo surgery. The observed difference was statistically significant (p <0.049) (Table 5).

Table 4: Outcome of TEF management.

Characteristics	Frequency (N=88)	%
Improved	70	79.5
Not improved	2	2.3
Anastomotic leak	2	2.3
Expired	14	15.9

DISCUSSION

Present study shows that majority of the participants were diagnosed with type 3 trachea esophageal fistula (80.7%). Echocardiography findings were abnormal for 50% of the participants and presence of coexisting anomalies including renal, liver and pelvic anomalies were present in 46.6% of the participants.

Table 5: Association between risk factor and outcome.

Parameters	Condition		N (88)	Chi sq	P value
	Improved	Not improved			
Procedure					
Surgery done	68 (82.9)	14 (17.1)	82	3.8	0.049
Surgery not done	3 (50.0)	3 (50.0)	6		

Studies have shown that >50% of TEF patients have associated anomalies. Majority of the anomalies include renal agenesis, microcephaly, duodenal atresia, limb reduction defects polycystic kidneys.^{6,7} In a study done by Green Wood RD et al, cardiovascular manifestation were coexisting with trachea esophageal fistula in about (14.7%) of patients in their study.

The observed finding was lower than seen in present study. These differences could be due to the regional differences between the studies. Several other studies have shown that hospital mortality in infants with both

trachea esophageal fistula and cardiovascular anomalies was as high as 79% in contrast to 23% among the patients who did not have cardiovascular anomalies.⁸

The classification of TEF is based on the embryological aspect of the defect. The primary type of congenital esophageal atresia with TEF is the majority type of TEF which is seen in 85% of the cases followed by isolated esophageal atresia without TEF which is around 8% and H type of atresia is seen in 4% of the cases. Proximal and distal TEF associated with esophageal atresia is seen in (3%) of the cases.⁹

In present study about 58% of the participants required ventilation in both pre-operative and post-operative period of which 36.4% required pre-operative ventilation. Surgery is the mainstay in the management of TEF. In majority of the participants (58%) surgery was performed within 24 hours of birth. The most common surgery performed for the study participants in present study was thoracotomy with TEF repair (71.6%). Following surgery about 79.5% of the participants had improvement in the respiratory status and did not require ventilation in the postoperative period. About 14 participants (15.9%) expired during course in the hospital. It was observed that performance of thoracotomy or any surgical intervention had significant improvement in the respiratory outcomes among the patients with TEF. The observed association was found to be statistically significant ($p < 0.05$).

There are several theories proposed regarding the embryology and formation of tracheoesophageal fistula. The primary digestive tube emerged from the primitive endoderm subsequently giving rise to esophagus and trachea. There are three theories which have been put forward to explain the phenomenon of esophageal atresia and TEF.

According to the first theory there is evagination of tracheal diverticulum which grows rapidly in caudal direction resulting in separation of trachea and esophagus. In this context the trachea esophageal fistula results from the growth failure which happens in the tracheal diverticulum. In another theory the formation mesenchymal septum in coronal plane of the primitive digestive tube separates trachea ventrally and esophagus dorsally. The failure of this process results in tracheoesophageal malformation. In another theory both first and second theory are combined wherein the rapid growth in tracheal diverticulum does not occur in concert with mesenchymal separation of the primitive digestive tube resulting in the malformation.⁹

The gold standard in the management of TEF is surgery. Several surgeries have proven that standard right posterolateral extrapleural thoracotomy below the tip of scapula is extremely useful in correcting the defect.¹⁰ There has been significant debate in determining the age at which surgery is ideally taken up by the patient. While one faction suggest that delayed surgical intervention helps in the extension and growth of the esophagus thereby facilitating adequate anastomosis, studies have proven that in approximately three months of age, there is adequate growth of the esophagus along with the gastric end which helps in achieving primary anastomosis. However other faction of researches have also attempted repair in the early neonatal period in order to facilitate better survival rates and minimize the complication due to the defect.¹¹ The postoperative period following the TEF repair depends on various factors. If the surgeon has used transanastomotic feeding tube, feeding through the tube is always done slowly beginning 48 hours following surgery. So, the primary complication of post-operative

period includes anastomotic leak, gastro-esophageal reflex, esophageal dysmotility, fistula recurrence, scoliosis, deformities of the thoracic wall and respiratory disorders.^{12,13}

Present study did not have long term complications in the patients who underwent the surgical repair. However, the analysis of the outcome shows that two patients has anastomotic leak (2.3%) while 14 (15.9%) patients expired during the postoperative period. Anastomotic leaks are considered as a minor complication and usually present in 15-20% of the patients.¹⁴ The leak usually results from small friable lower segment ischemia of esophageal ends due to extensive anastomotic tension or sepsis or in some cases poor suturing techniques and excessive mobilization of the distal pouch and increased gap length.¹⁵

CONCLUSION

Present study has shown that majority of the tracheoesophageal fistula is type 3 and is associated with one or other form of cardiovascular anomalies. Majority of the patients with TEF require ventilatory support both during pre and postoperative period. Of the several surgical techniques available for management of TEF thoracotomy with TEF repair is said to be one of the gold standard procedures which has in recent times replaced by thoroscopies techniques of achieving anastomosis. Irrespective of the surgical procedures there are very few complications which occur following surgery which include anastomotic leak, gastrointestinal reflex, strictures in the esophageal tract etc. Implementation of any one of the surgical procedures associated with an improved outcome in the respiratory and gastrointestinal physiology and this is found to be statistically significant. Present study has shown that there is an ongoing need for early surgical technique in order to prevent the complication which results from respiratory failure and feeding difficulties. There is an increase need for detecting TEF early during the antenatal period which helps in prenatal diagnosis of TEF repair and other congenital anomalies.

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