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Original Research Article

Esophageal atresia and Tracheoesophageal fistula with associated anomalies in a tertiary care hospital of north India

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ABSTRACT

Background: Esophageal atresia (EA) with or without Tracheoesophageal fistula (TEF) has been described as the epitome of pediatric surgery and the management of these neonates require high degree of skill, manpower and tertiary care. This clinical study was undertaken to identify the EA and TEF cases along with the pattern of associated anomalies causing mortality in Varanasi .
Methods: This prospective study included 53 patients of EA with TEF who underwent primary esophageal anastomosis. Their clinical profile, time of presentation, various associated anomalies, sex distribution, and their effect on mortality and morbidity were observed.
Results: Mortality was high (41.64%) among patients who reached the tertiary centre late i.e. >24 hours. The incidence of low birth weight babies is approximately 75% and mortality rate increased with low birth weight. Almost 2/3 of patients had clinical or radiological evidence of pneumonia. The mortality was very high in severe pneumonia 66.7% as compared to 15% patients without pneumonia. Septicaemia and leak were most common complications. The mortality in septicemic patients is high (77.8%). The overall incidence of associated anomalies was 47%. Early surgical intervention improved the survival of EA with TEF with major GIT associated anomalies. The overall survival rate was 60.37% and mortality was 39.67%.
Conclusions: Although we have improved a lot in managing these cases of EA with TEF, but we have to do much more to achieve our goal of near 100% survival.

Keywords: Esophageal atresia, Mortality, Tracheoesophageal fistula

INTRODUCTION

Esophageal atresia (EA) with or without Tracheoesophageal fistula (TEF) has been described as the epitome of pediatric surgery and the management of these neonates require high degree of skill, manpower and tertiary care.

The survival of these newborns depends not only in excellent preoperative, intra operative and post-operative care but also on adequate weight of the patient, early diagnosis, prompt referral, good care in transport, and condition of the lungs (pneumonia).

Another major factor contributing to the mortality is presence of associated anomalies. The overall incidence has been reported to be as high as 50% to 70%. Cardiovascular anomalies occur most frequently (11-49%) followed by genitourinary 24%, gastrointestinal 24% and skeletal 13%.[1-3] Neural tube defects 2.3%, hydrocephalus 5.2%, abdominal wall defects 4.3% and Diaphragmatic hernia 2.9% have been reported Harris et al.[4]

Some associated anomalies are major while some are life threatening. Due to the tremendous advancement in the care of newborns viz, neonatal ICU, highly trained

nursing, neonatal ventilators, newer antibiotics, advancement in neonatal anaesthesia, the survival in EA with TEF is approaching about 100% especially in Western countries. But in our country mortality is still very high, due to many factors.

Hence this clinical study was undertaken to identify the cases of EA and TEF in our hospital, along with the pattern of associated anomalies causing mortality.

METHODS

This prospective study was carried out in the Department of Paediatric Surgery, University Hospital, Sir Sunderial Hospital, Banaras Hindu University from March 2003 to February 2005. A total of 68 patients of EA-TEF and pure EA without TEF were admitted. 53 patients who underwent end to end anastomosis of proximal and distal pouch of esophagus with fistula closure were included in this study. Five patients of esophageal atresia and tracheo esophageal fistula died before any surgical intervention due to septicemia and 10 patients were of pure esophageal atresia in whom cervical esophagostomy and gastrostomy were performed, these 15 patients were excluded.

A detailed history was taken, clinical examination and necessary investigation were carried out and recorded in a printed proforma. Birth weight, presence of pneumonia, other associated anomalies and post-operative complications were noted.

Pneumonia was graded into no pneumonia (normal chest x-ray, no respiratory distress), moderate (pneumonia restricted to one lobe or moderate respiratory distress) and severe (pneumonia in both lobe or lung, or presence of severe respiratory distress) category.

Cardiac anomalies were assessed clinically, by presence of murmur or central cyanosis, 2D-echocardiography was performed when feasible. Other congenital anomalies were assessed clinically, radiologically or on exploration. Surgical intervention was done for some anomalies.

Primary repair of EA-TEF was done using standard operational procedure. Cervical esophagostomy and gastrostomy (in cases of EA without fistula) was done.

Post-operative ventilatory support was provided whenever required. Nebulization chest physiotherapy and oropharyngeal suction was done intermittently.

Postoperative complications

Leak was identified by presence of saliva in chest tube and septicemia was diagnosed clinically supported by investigation. Survival was defined as discharge of patients accepting feed orally after surgery. The survived cases were followed up to the end of the study for late post-operative complication and morbidity.

Statistical analysis

The data have been expressed as mean±standard deviation. Chi-square test and Z-test were applied.

RESULTS

There were 36 males (67.9%) and 17 females (32.1%) in the present study. The male:female ratio being >2:1.

Table 1: Mortality in major and minor associated anomalies in different system.

Systems	Anomalies	Survival	Survival (%)	Expired	Mortality (%)
CVS	Minor-5	4	80	1	20
	Major-4	0	0	4	100
GIT	Minor-1	1	100	-	-
	Major-9	6	66.66	3	33.33
Genitourinary	Minor-3	2	66.6	1	33.33
	Major-1	-	-	1	100
Musculoskeletal	Minor-3	2	66.6	1	33.33
	Major-2	1	50	1	50
CNS	Minor-2	1	50	1	50
	Major-1	-	-	1	100
Other (Ear tag, bilateral Pinna agenesis and Down's syndrome)	Minor-2	-	-	2	100
	Major-1	1	100	-	-

Only 17 (32.07%) patients arrived within 24 hours, 24 (45.27%) came within 24-72 hours and 12 (22.64%) cases came after 72 hours. Only 15% of cases were <2 kg birth weight and only 25% patients with normal birth weight. 40 cases (75.5%) had pneumonia clinically

and/or radiologically rendering the survival poor and increasing the morbidity. Septicemia was seen in 18 cases (34%), 7 (21.22%) patients developed esophageal stricture and 6 patients (18.18%) had gastroesophageal dysmotility and 4 (12.12%) had recurrent chest infection due to GER. 18 (33.96%) had major anomalies and 16 had minor anomalies.

Table 2: Sex wise mortality among patients.

Sex	No.	Survival	Percentage	Expired	Percentage
Male	36	21	58.3	15	41.7
Female	17	11	64.70	6	35.3
Total	53	32	60.37	21	39.63

x2=0.196, df=1, p=NS; Survival was higher in female patients

Table 3: Distribution of mortality according to the time of presentation.

Age	Total	Survival	Percentage	Expired	Percentage
<24 hours	17	11	64.7	6	35.29
27-72 hours	24	13	54.16	11	45.84
>72 hours	12	8	66.66	4	33.34
Total	53	32	60.37	21	39.63

X^2=0.42, df=1, p=NS; Survival rate was good in patients arriving within 24 hours

Table 4: Mortality according to the birth weight.

Weight (Kg)	No.	Survival	Percentage	Expired	Percentage
<2	8	4	50	4	50
2-2.5	32	19	59.37	13	40.6
>2.5	13	9	69.24	4	30.76
Total	53	32	60.37	21	39.63

X^2=1.85, df=2, p=NS; The mortality was higher in low weight babies than normal birth weight, having no statistical correlation

Table 5: Mortality according to the severity of pneumonia.

Pneumonia	No.	Survival	Percentage	Expired	Percentage
No	13	11	84.6	2	15.39
Moderate	25	16	64.0	7	36.0
Severe	15	5	33.3	10	66.7
Total	53	32	60.37	21	39.63

X^2=6.42, df=2, p<0.05 (significant); Approximately 75% patients had radiological or clinical evidence of pneumonia and the mortality was statistically significant in this groups

Table 6: Mortality in minor and major anomalies.

Anomalies	No.	Survival	%	Expired	Percentage
EA with TEF with only minor AA	10	7	70	3	30
EA with TEF with only major AA or major associated with minor	18	9	50	9	50
EA with TEF without AA	28	18	64.18	10	35.82
Total	53	32	60.37	21	39.63

x2=0.52, df=2, p=NS

Ten patients had only minor anomalies with 70% survival. The mortality in the group of major associated anomalies was higher. Total number of patients was 18 in this group and the survival rate was 50%. Out of 53 patients, 21 patients (39.63%) expired. The survival rate was 60.37% (32 patients). The mortality in major associated anomalies was higher (50%) than mortality among those with minor associated anomalies and those without associated anomalies but it was not statistically significant.

Table 7: Mortality in relation to complications.

Complication	No.	Survival	%	Expired	%
EA with TEF septicemia	18	4	22.2	14	77.8
EA with TEF anastomotic leak	5	2	40	3	60
EA with TEF without complication	30	26	86.7	4	13.3
Total	53	32	60.37	21	39.63

The mortality was very high in the septicemia group (77.8%) as compared anastomotic leak (60%) ($p<0.001$).

In follow up only 17 patients turned up during the period of study out of 33 patients and approximately 21% patients developed esophageal stricture later managed by repeated esophageal dilation. 18.0% patients were developed regurgitation of feed and 12% patients readmitted with severe chest infection due to GER. These patients were managed conservatively.

DISCUSSION

Remarkable success has been achieved in management of Esophageal atresia in modern times. This has been achieved based on improvements in surgical technique, neonatal anesthesia, ventilatory support; effective suctioning of upper pouch to eliminate the risk of aspiration pneumonia and modern sophisticated neonatal intensive care. Technical advances too have a role to play in the success story, with a better choice of suture material, use of elective mechanical ventilation postoperatively; early recognition and treatment of anastomotic complications. Survival of low birth babies are being increasingly reported, with mortality being limited to neonates with chromosomal defects and/or severe anomalies that are incompatible with life.

Survival of esophageal atresia patients, however, has not shown such a remarkable improvement in developing countries. Various reasons have been cited for this higher mortality rate. These include late presentation of patients at centers, managing this problem, poor transportation facilities for sick neonates, concentration of trained manpower in the major metropolitan cities and lack of the adequate infrastructure at most of the centers in India.

In the present study, among the 53 patients of esophageal atresia and tracheo-esophageal fistula, the sex distribution showed that male patients were more than the double of female patients. Similar male predominance has been reported by Kronemer et al and Arora M et al.[5,6] This may be due to the reason that in our society male child's health is given preference over the female child's health.

Only 32% of our patients reached the tertiary centre within 24 hours. Arora et al, reported similar kind of data i.e. 40.7% cases reached within first 24 hours in spite of 75% deliveries at hospital.[6] Agarwal et al in AIIMS hospital reported that the patients presenting within 24 hours were 44% and similar data (46%) was also obtained by Sharma AK et al.[7,8] Traveling long distance, poor transport system and poverty are also the contributory factors.

Mortality was high among patients who reached the tertiary centre late i.e. >24 hours. This is similar to the observations reported by Arora M et al.[6] In our setup, there is no facility of ultrasonography in primary health centre, sub-center or in antenatal clinic and lack of awareness of this anomaly in medical and paramedical workers. The early diagnosis of EA with TEF is possible if obstetrician suspect the anomaly in antenatal period in the pregnancy associated with polyhydramnios (incidence is 0.5-1% in normal pregnancies). These pregnancies should be taken as high-risk pregnancies and should be kept under observation and regular follow up throughout the pregnancy to facilitate early diagnosis and early referral of EA with TEF patient to the better centre. The role of ultrasonography in prenatal diagnosis of EA with TEF is limited because of less accuracy. Sparey C et al reported 50% of accuracy and 42% accuracy was observed by Robert K et al.[9,10]

In our setup, the breast feeding of baby is routinely recommended by the pediatrician after 30 minutes in normal deliveries and after 3 hours in caesarean section. Thus, the EA with TEF present within few hours following the birth as they received breast feeding with symptoms of cyanotic spells, cough, drooling of saliva or milk etc.

In the present study, the incidence of low birth weight babies was approximately 75% according to the WHO definition but in normal Indian population, the incidence of low birth rate (<2.5 kg) is 33% of all live births (World health report, 1995).[11] The incidence of <2 kg babies was 5.5% in normal population and 15% in the present study which is comparable with Arora M et al they reported the incidence of less than 1.8 kg as 15%.[6]

The reasons for the high incidence of 75% of low birth weight found in the present study are many, such as rural background population, adolescent pregnancies, higher incidence of severe anemia, maternal malnutrition, multiple pregnancies and antenatal infections which are mostly related to poverty. The mortality in the low birth weight patients is quite high than the babies weighted more than 2.5 kg but it was statistically not significant. The results are comparable with those reported by Arora M et al.[6] Almost 2/3 of patients had clinical or

radiological evidence of pneumonia (75.5%). The presence of pneumonia grossly affects the survival. When we compared our data with that of study conducted by Agarwal S et al we observed that survival has improved over the past few years probably because of better pre-operative and post-operative care and potent newer antibiotics.[7] Pneumonia in EA with TEF patients is mainly due to aspiration of saliva, preoperative breast feeding, and reflux from fistulous communication with trachea. So, these can be prevented with vigorous upper pouch section during transportation and early diagnosis before feeding may decrease the incidence of pneumonia and increase the survival of EA with TEF patients.

In the present study, the patient having septicemia at the time of admission or developed post operatively contributes 34% of patients and anastomotic leak was present in only 40% of cases. 56.6% of these cases remain without above complications. In comparison to the Arora M et al where they reported the leak rate of 27% on the basis of presence of salvia in chest drain in 59% of patients and respiratory distress in 41% patients but in the present study the leak was considered when the presence of saliva in chest drain.[6] Moreover, in our setup the autopsy was not performed to look for the cause of death or confirmation of leak.

The total number of major associated anomalies was 34% and minor associated anomalies were 30% in the present study. In comparison to Arora M et al, they found 21% of major associated major anomalies and 33% of minor associated anomalies and the overall incidence of associated anomalies patients in the present study was 47% as of 35% incidence reported by Arora M et al, 49% by Rejjal A et al and 50% by Dwayne C et al.[6,12,13] These data are comparable with the present study. The system wise distribution of associated anomalies is comparable with Rejjal A et al.[12] In comparison to Rejjal A et al, we have higher incidence of GI tract anomalies lower incidence of genitourinary tract anomalies.[12] Survival rate was 60.37% and mortality rate was 39.63% which is comparable with Arora M et al.[6] There are various reasons for higher mortality e.g. late presentation of patients at the centre managing these problems, poor transportation facilities for sick neonates, concentration of trained man power only in the major metropolitan cities, lack of adequate infrastructure at most of the centers in India.

CONCLUSION

The mortality is still very high in cases of EA with TEF (39.63%) with survival being 60.37% in our setup. Among the babies who managed to reach us, pneumonia and septicemia had highest mortality and were found to be highly statistically significant. Presence of other associated anomalies also contributed to high mortality in the present study. Although we have improved a lot in managing these cases of EA with TEF, but we have to do much more to achieve our goal of near 100% survival.

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