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Effect of number of associated anomalies on outcome in oesophageal atresia with or without tracheoesophageal fistula patient

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ABSTRACT

Background: The objective of this study was to assess effect of number of associated anomalies on outcome in oesophageal atresia with or without tracheoesophageal fistula patients. **Materials and Methods:** Retrospective analysis of records of neonates admitted with a diagnosis of oesophageal atresia (EA) with or without tracheoesophageal fistula during January 2005 to May 2011. Preoperative investigation included chest X-Ray, ultrasonography of abdomen and echocardiography. Associated anomalies were grouped as minor or major depending on whether life threatening or not. Major anomalies were further sub grouped according to the involvement of single, two or > 2 organ systems. Survival was correlated with the presence of anomalies and the number of systems involved. **Results:** Out of 301 patients with EA, 240 survived (79.7%). Of these 301, 117 (38.9%) had no associated anomalies. Of the total 61 deaths, 59% (36/61) were in patients with cardiac anomalies and 44% (27/61) were in those with >2 associated anomalies. The mortality rate was highest in those with >2 anomalies 27/34 (79.4%), whereas survival was best in those without any associated anomalies 104/117 (88.9%). **Conclusions:** Apart from other factors, an association of more than two system anomalies influence the mortality in oesophageal atresia.

Key words: Associated anomalies, mortality ratee oesophageal atresia

INTRODUCTION

Oesophageal atresia (OA), with or without tracheoesophageal fistula (TEF), is the commonest oesophageal malformation of the esophagus with an incidence of 1 in 3,500 births.[1] In the majority of cases, the aetiology is still unknown. Ample reports are available in English literature describing associated anomalies with EA, with cardiac malformation being the most common. Association can occur either as part of VATER (Vertebral defects, Anal atresia, Tracheo-oesophageal fistula (TEF) with esophageal atresia, and Radial and Renal dysplasia) or VACTERL [abnormalities of vertebrae (V), anus (A), cardiovascular tree (C), trachea (T), oesophagus (E), renal system (R), and limb buds (L)] syndrome as described by Quan and Smith in 1973 or as nonsyndromic oesophageal atresia.[2] Apart from other factors, these associated major anomalies play a significant role in the final outcome in term of survival especially in developing countries like India.

In the present study, we have tried to correlate survival with the presence of major associated anomalies and the number of organ systems involved.

MATERIALS AND METHODS

We retrospectively analysed our records of neonates admitted with a diagnosis of EA with or without TEF from January 2005 to May 2011. All cases were admitted through emergency services. Preoperative investigations include X-ray chest with red rubber catheter per orally, ultrasonography of the abdomen and echocardiography. Associated anomalies were classified as minor or major. Minor anomalies were defined as those anomalies which are not life threatening, whereas major anomalies were defined as those anomalies which were life threatening. Major associated anomalies were further sub-classified as having involvement of either single organ system or two or more than two organ systems. All children underwent thoracotomy and primary repair except pure esophageal atresia, where a primary diversion in the form of cervical oesophagostomy and abdominal oesophagostomy was

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performed. All cases were electively ventilated for 48 hours in the postoperative period. Records were maintained regarding the demographic profile of the cases, therapeutic intervention, ventilatory requirement and final outcome in term of survival. Institute's ethical committee approval for the study was taken.

RESULTS

A total of 301 cases of oesophageal atresia was admitted during this period. Out of 301, 189 were males while 112 were females with M:F ratio of 1.6:1. Mean age at presentation was 26.7 hours (range 3-96 hrs). Mean weight at presentation was 2.34 kg (range 1.78-3.2 kg). Of 301 patients with EA, 240 survived (79.7%). Of these 301, 117 (38.9%) had no associated anomalies and of them 104 (88.9%) survived. Out of 184 (61.1%) with various anomalies, 136 (73.9%) survived. Of 301patients, 130 (43.2%) had cardiac anomalies, 30 (9.9%) had anorectal malformation, 12 (3.9%) had duodenal atresia [Table 1]. Other non-lethal anomalies were radial anomalies (14), vertebral (12) and hypospadias (1). Out of 301, 33 (10.9%) patients had >2 associated anomalies of whom only six (18%) survived. Of the total 61 deaths, 44 % (27/61) were in those with >2 associated anomalies. Survival was better in those with no associated anomalies 104/117 (88.9%) or single associated anomaly 97/107 (90.6%).

There was no statistical significant value comparing survival of EA patients with no associated anomalies with patients associated with either single or two anomalies but survival was significant when compared with EA patients with >2 anomalies ($P < 0.001$) [Table 2]. There was statistical significant value comparing survival of OA patients with ≤2 associated anomalies with >2 associated anomalies ($P < 0.001$).

Table 1: Esophageal atresia patients with associated anomalies

Esophageal atresia patients	Total (*n*=301)	Percentage (%)
Cardiac anomalies	130	43.2
Anorectal malformation	30	9.9
Radial ray defects	14	4.6
Duodenal atresia	12	3.9
Vertebral defects	12	3.9
Hypospadias	1	0.3

Table 2: Survival of oesophageal atresia patients with number of associated anomalies

Associated anomaly	Total	Survival	Percentage	*P* value
EA without anomaly	117	104	88.9	0.29
Single associated anomaly	107	97	90.6	0.92
Two associated anomalies	44	33	75	0.52
≤2 associated anomalies	151	130	86.1	0.86
>2 Associated anomalies	33	6	18	<0.001
Total	301	240	79.7	

EA- Esophageal atresia

DISCUSSION

The early disturbance in organogenesis which results in esophageal atresia also leads to other associated anomalies, the incidence of which varies from 40% to 55%.[3-6] These associated anomalies plays pivotal role in the final survival outcome.

A total of 184 (61.1%) cases of EA had other associated anomalies, an incidence that confirms other large reported series.[7,8] Most commonly found association in OA is VACTERL or VATER. In 20-67% cases of VACTERL anomalies, EA was associated while 5-15% of EA cases are reported to have VACTERL anomalies.[9-12] Other associated anomalies include trisomy 21, trisomy 18, Potter syndrome, Goldenhar syndrome, CHARGE and duodenal atresia.[1,9,11] Hasaab *et al.*, has reported associated anomaly in 60% of cases with VACTERL seen in 6% of them.[13] Associated congenital anomalies in other series were Spitz *et al.*, (47%), Saing *et al.*, (59%), Rokitansky *et al.* (52.4%).[14-16] Ein SH *et al.*, in 1989 tried to establish the prognosis with presence of associated anomalies.[3] In a series of 97 newborns they concluded that newborn who have a common type of EA – TEF without any associated anomalies have a survival rate of 100% against 64% in cases with associated anomalies. Most commonly associated congenital defects are cardiovascular (28%), gastrointestinal (17%), skeletal (12%) and genitourinary (8%).[3] Choudhaury SR *et al.*, in a series of 240 cases reviewed the influence of various factors on the outcome and concluded that cardiac and chromosomal anomalies are significant causes of death, particularly for an early demise.[17]

Through our study we have not only tried to establish the effect of associated anomalies on final outcome but also tried to emphasise that the severity and number of associated anomalies are inversely related to the survival outcome. Studies have shown that the mortality rate is double in cases which have associated anomalies.[3,6] Iuchtman M *et al.*, also showed 24% more mortality rate in cases with associated

anomalies, than without any anomalies.[18] In our study group, survival rate was 88.9%, 90%, 75% and 18%, respectively, for no associated anomalies, single associated anomaly, two associated anomalies and >2 associated anomalies, with an overall survival of 79.7%. Most common anomaly seen in our study was cardiac (43.2%) followed by anorectal malformation (9.9%), radial ray defect (4.6%), duodenal atresia (3.9%), vertebral defect (3.9%) and hypospadias (0.3%) [Table 2]. Only six cases out of 33 having >2 associated anomalies survived.

Successfully treating infants with combinations of severe congenital anomalies is one of the most challenging aspects of paediatric surgery. Apart from other prognostic factors as described by Waterson and Spitz *et al.*, presence of a number of associated anomalies and the severity of associated anomaly determine the final outcome. Our study supports the findings of Saing H *et al.*, that apart from other factors, an association of two or more system anomalies and the severity of associated anomalies influence the mortality in esophageal atresia.[15]

CONCLUSION

Apart from other prognostic factors, an association of more than two system anomalies influences the mortality in esophageal atresia.

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