

Original Article

Profile and Outcome of Esophageal Atresia in a Rapidly Developing Area

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ABSTRACT

Objective: To study the profile of cases of esophageal atresia (EA) and/or tracheoesophageal fistula (TEF), and the effect of modalities of surgical and postoperative management on their outcome.

Setting: MCH, Riyadh, Saudi Arabia.

Methods: Retrospective analysis of records of 100 cases, over eight years between 1990 and 1998.

Results: The prevalence rate was 4.3 per 10,000 neonates. Associated anomalies were detected in 60% of cases. Of these, 33% were cardiac and 25% were renal. Primary anastomosis was successfully performed on 95% of patients, in which the gap between the two-esophageal pouches was less than 2 inches. Delayed anastomosis was performed in those with longer gap length or extreme prematurity. The majority (68%) of cases (group-I) received postoperative elective ventilation (POEV), and

32% (group-II) did not. Gastroesophageal reflux (GER) affected 26.4% of group-I compared with 56.3% of group-II ($p < 0.001$). Mean length of stay (LOS) in ICU was 12.2 days (SD: 2.0) in group-I and 13.2 days (SD: 3.2) in group-II, ($P < 0.05$). Leakage affected 8.8% of group-I and 25% of group-II ($P < 0.01$). Stricture developed in 7.4% of group-I and 12.5% of group-II ($P < 0.01$). Survival rate was 93%. Severe congenital malformations were present in 6 out of 7 deaths.

Conclusions: Prevalence of EA is high in our area. Both primary and delayed repairs are successful when the patient is properly selected, according to the gap length and the degree of prematurity. Severe congenital malformation represents the major leading cause of mortality. POEV was associated with a lower rate of GER, leakage, stricture and LOS.

KEY WORDS: elective ventilation, esophageal atresia, tracheoesophageal fistula

INTRODUCTION

Over 200 years, and after the first description of EA by YALadd, this condition was universally fatal until successful surgical management was established^[1,2]. The outcome of management varies from one center to another according to facilities and associated anomalies^[1-3]. Furthermore, pre-, intra-, and postoperative management represent the keystone for successful management. In our rapidly developing territory, it reflects the progress of medical services offered to patients.

The aim of this study is to explore the profile of this congenital anomaly and to find out the effect of timing decision making on the associated malformations, type of surgery, and postoperative elective ventilation (POEV) on the outcome of our cases.

PATIENTS AND METHODS

In a retrospective study, we reviewed 100 consecutive patients with EA or TEF, or both, who underwent surgical intervention at MCH, Riyadh, over a period of eight years from January 1990 to December 1997. Cases were either delivered in this centre and detected by the routine placing of a

feeding tube and confirmed by chest radiographs, or referred from other hospitals. Renal ultrasound scan and echocardiography were performed on all patients. Dismorphic babies were subjected to computer search and chromosomal studies.

Primary repair was performed in cases with reasonable gap length of no more than 2 inches between the ends of the two pouches. Delayed anastomosis was done on cases with a distance of more than 2 inches, or with extreme prematurity. Repair was done through extrapleural approach, in two layers, or one layer using mono-filament absorbable suture (polydioxanone 5/0), and stented on nasogastric tube. All cases received their postoperative care at NICU and PICU.

Total parenteral nutrition (TPN) was initially given in all patients. Feeding through a nasogastric tube was started on 4-7th postoperative day. The chest drain was left until esophagogram ruled out leakage on the 7th postoperative day. If no leakage was detected, TPN was stopped, the nasogastric tube was removed, and oral feeding was completely established. Delayed feeding took place after four weeks in cases with major disruption. In minor leakage, feeding was delayed for two weeks

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Table 1

Waterston's classification among patients with esophageal atresia

Group	No.	Mortality	
		No.	%
A	60	0	0.0
B	27	1	3.7
C	13	6	46.2

only. All babies received prophylactic antibiotics, and empirical antibiotics were given whenever infection was suspected until a culture and sensitivity was available. Age at surgical intervention, postoperative ventilation, postoperative complications, length of ICU stay, and the outcome were studied and analyzed.

Statistical Methods: The SPSS-PC version 7.5 program was used for data management and analysis. Student's t-test was used in statistical analysis. All reported P values were two - tailed, value < 0.5 was considered significant.

RESULTS

During the study period, 100 neonates had EA and/or TEF. Among these patients, 55 cases were delivered in our centre representing a prevalence of 4.3 per 10,000 neonates.

A total of 45 cases were referred from other hospitals; 76% were preterm and 24% were term deliveries. The mean gestational age was 34 weeks (SD = 3.2), 52% were males and 48% females. A total of 60% fell into Waterston's risk category A, 27% B and 13% C (Table 1).

Maternal age ranged between 18 and 32 years (mean = 22.6; SD = 3.2). Parents of 65% of cases were consanguineous. Polyhydramnios affected 72 pregnancies, 6 cases were diagnosed by antenatal ultrasonography. A overwhelming majority (94%) of the cases were EA with distal TEF, while 4% had proximal and distal blind pouches without fistulas and 2% had EA with proximal TEF.

A total 60% had associated congenital malformations. Cardiac malformations affected 33% of the cases; renal anomalies affected 25%, genitourinary 18% and skeletal anomalies 10% of cases. Furthermore, 6% had VACTERL (Vertebral, Anal, Cardiac, Tracheo-Esophageal, Renal or Radial, Limb anomalies) associations while 3% had chromosomal aberrations, and one had Potter syndrome. Overall, 8% of cases had severe multiple malformations (Table 2).

A total of 55 cases were operated upon in the first 24 hours and 35 between 24 to 36 hours. The latter group was all referred from other hospitals. A

Table 2

Congenital malformations associated with esophageal atresia

Congenital anomaly	No.	%
Cardiac:	33	33
VSD	12	12
ASD	7	7
PDA	6	6
PS	1	1
Complex	7	7
Renal	25	25
Genitourinary	18	18
Skeletal	10	10
VATER associations	6	6
Chromosomal aberrations:	3	3
Trisomy-18	1	1
Trisomy-13	1	1
Trisomy-21	1	1
Potter syndrome	1	1

VSD: ventricular septal defect ASD: atrial septal defect
PDA: patent ductus arteriosus PS: pulmonary stenosis

total of 95% of cases had primary anastomosis and 5% had delayed repair.

Of the total 100 cases, 68 (group-I) received POEV for 48 hours and 32 (group-II) did not. Mean LOS was 12.2 days (SD = 2.0) in group-I, compared with 13.2 days (SD = 3.2) in group-II, (P<0.05). Similarly, GER developed in 18 (26.4%) out of group-I, compared with 18 (56.3%) of group-II (P<0.001).

GER, leakage and strictures were significantly less in group-I (Table 3). Most cases of GER were grade one and two, and responded well to conservative management. Only one case needed Nissan fundoplication by the age of one year. Stricture has developed in five cases (7.4%) of group-I and four (12.5%) of group-II (P<0.01). It was mild in seven cases, and severe in two cases. Of the latter, one responded to dilatation and the other one needed thoracotomy and revision of anastomosis by age of 9 months. Leakage occurred in 14 cases, 6 (8.8%) from group-I and eight (25%) from group-II (p<0.01). A total of 12 cases had mild leakage, which was improved within 7 to 10 days, and two had major leakage necessitating gastrostomy tube feeding.

Complications included aspiration pneumonia in 19 (42%) out of 45 referred cases and 2 (3.6%) out of 55 cases delivered in our center; one of them had apneas and cyanotic spells, which improved following aortopexy by age of 4 months. Two cases developed tracheomalacia detected by bronchoscope. One recurring fistula was seen and was repaired (Table 4). Out of seven deaths, five were due to complex congenital cardiac lesions, one due to gram negative septicemia and one with severe multiple congenital malformations (Potter syndrome).

Table 3

Comparison between group-I and group-II in LOS and GER

	Group-I (POEV) No. = 68	Group-II (no POEV) No. = 32	t	p
LOS in days:				
Mean (SD)	12.2 (2)	13.2 (3.2)	2.3	<0.05
Range	9 - 18	10 - 24		
GER: Mean (SD)	18 (0.38)	75 (0.44)	6.7	<0.001
Leakage	6	8	4.1	<0.01
Stricture	5	4	3.6	<0.01

LOS: length of stay GER: gastroesophageal reflux

DISCUSSION

The prevalence of 4.3/10,000 EA with or without TEF according to this study is higher in our area in comparison to other reports of 2.6 and 2.86 in Europe^[4,5]. That might be explained by the high rate of consanguinity in Gulf area, which constitutes 65% among parents of our patients. Polyhydramnios was present in 72 of our series. The presence of ultrasonographic evidence for in-utero growth retardation and absence of stomach liquid helps in making the antenatal diagnosis^[6]. Such findings were detected in six cases. This antenatal diagnosis has helped in supporting them by inserting a feeding tube and prohibiting any oral feeding after birth thus giving a chance for safe intervention. Chromosomal aberrations were detected in 3% (3 patients), compared to 10% in Depaeppe et al^[4]. Similarly, patients with VACTERL associations were fewer (6%), in comparison with 37% in other reports^[7].

In agreement with other studies^[8], our study has shown significant low birth weight of affected babies, and low age and parity among their mothers.

Most cases with aspiration pneumonia were referred from peripheral hospitals. This reflects the late diagnosis in peripheral hospitals, following the first feed. On the contrary, and similar to the finding shown by A. Salem et al^[1], our study has shown that the early detection of EA by the routine nasogastric tube insertion and awareness of the attending pediatricians in our centre has resulted in fewer aspirations.

Some authors^[9] preferred the delayed repair in all cases with EA and TEF regardless of the distance between the two esophageal pouches, assuming that such policy preserve their native esophagus. Others^[10] favored the primary repair irrespective of gap length, assuming that esophageal anastomosis can withstand considerable tension and allows a reliable true primary repair for the full EA spectrum. In further support to Rosinha et al^[11], our study has shown the best outcome when the

Table 4

Postoperative complications of esophageal atresia repair

Complication	No. of cases
Septicemia	6
GER	36
Aspiration	21
Leakage:	
Major	2
Minor	12
Tracheomalacia	2
Stricture:	
Major	2
Minor	10
Recurrent fistula	1

GER: gastroesophageal reflux

primary repair is performed for a reasonable distance of less than 2 inches between the two pouches, and the delayed anastomosis is used for those with longer distance or extreme prematurity.

The presence of recurrent fistula in 1% in our series is comparable to other reports^[12].

The value of POEV in reducing anastomotic leaks is still controversial^[13,14]. Like Chittmitrapap et al^[15], our report showed that POEV is associated with lower rate of strictures, leakage and GER. This might be explained by the protection of esophageal anastomosis following the repair of EA. Furthermore, a shorter LOS has been shown among those with POEV. This considerably reduces ICU bed utilization and overall cost^[16].

In agreement with Rogers et al^[3] and Giadaro et al^[17], we have found that mortality is mainly related to the presence of severe multiple associated congenital malformations and severe congenital cardiac malformations. VACTERAL associations were seen in 6% of our cases, such associations are far less than that in other studies^[7,18,19]. A survival rate of 93% in our series is comparable to other neighboring gulf countries^[20].

In conclusion, EA with or without TEF has high prevalence in this part of the world. Low maternal age and low parity are significant risk factors for this anomaly. Early oral feeding and delay in making the diagnosis are significant risk factors for morbidity. Primary anastomosis has an excellent outcome whenever the gap length is reasonable. Alternatively, delayed repair is needed for those with long-gap-anastomosis. POEV is significantly associated with lower rate of leakage, stricture and GER, and the length of ICU stay.

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