

# Esophageal atresia: our experiences in a university hospital

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**Introduction and aim** Esophageal atresia is a relatively common congenital malformation occurring one in 2500–3000 live births. The aim of this study was to determine the frequency, type of anomaly, and mortality and to detect the associated anomaly in patients with esophageal atresia.

**Patients and methods** All neonates with diagnosis of esophageal atresia who were referred to pediatric surgery ward of Imam Khomeini Hospital were included in this study. Duration of this study was 10 years from 20 March 1997 to 20 March 2006. For comparison, duration of the study was divided into two periods (1997–2001 and 2002–2006). Sex, mortality rate, associated anomalies, type of atresia, mortality, performing thoracostomy or gastrostomy, and packed cell infusion were studied. Gross classification was used for typing of anomaly. Analysis was performed using the Pearson  $\chi^2$ -test and analysis of variance using SPSS.

**Results** In this study, 198 (male = 100, female = 98) neonates were included. The most frequent type of atresia was type C (93.4%). Overall mortality rate was 50%. Mortality during the first period was 54.43% and during the second period was 47.05% ( $P=0.384$ ). The mean age

at the second surgery was significantly higher in type D patients compared with others. The rate of gastrostomy was significantly higher during the first period (89.87%) compared with the second period of study (79.27%) ( $P=0.002$ ). The rate of cervical esophagostomy was decreased from 8.86 to 4.23% ( $P=0.228$ ).

**Conclusion** Mortality rate has decreased in our hospitals. The rate of gastrostomy decreased during the second period of study. The age at the first surgery was significantly higher in type D classification patients. *Ann Pediatr Surg* 10:31–34 © 2014 Annals of Pediatric Surgery.

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**Keywords:** esophageal atresia, gastrostomy, morbidity, mortality, thoracostomy, tracheoesophageal fistula, VACTERL syndrome

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## Introduction and aim

Esophageal atresia is a relatively common congenital malformation occurring one in 2500–3000 live births [1,2]. The most common type of anomaly is esophageal atresia with distal tracheoesophageal fistula, which represents in 86% of patients [1]. Waterson *et al.* [3] reported a survival rate of 57.6% in the early 1950s. In a 25-year study from India, mortality has decreased from 95.4 to 41% [4]. The aim of this study was to determine the frequency and type of anomaly and to detect the associated anomaly in patients with esophageal atresia.

## Patients and methods

All neonates with diagnosis of esophageal atresia who were referred to Imam Khomeini hospital were included in this study. Duration of this study was 10 years from 20 March 1997 to 20 March 2006. For comparison, duration of the study was divided into two periods (1997–2001 and 2002–2006). Sex, mortality rate, associated anomalies, type of atresia, performing thoracostomy or gastrostomy, and packed cell infusion were studied. Gross classification was used for typing of anomaly. Analysis was performed using the Pearson  $\chi^2$ -test and analysis of variance using SPSS (ver. 13.0; SPSS Inc., Chicago, Illinois, USA).

## Results

In the current study, 198 (male = 100, female = 98) neonates were included. Although there was slight

difference in male/female ratio between the first and second period of this study, this difference was statistically not significant. During the first period of study, 60.75% (48 of 79) patients were admitted in neonatal intensive care unit (NICU). During the second period of study, 86.55% (103 of 119) patients were admitted in NICU (Table 1). This difference was statistically significant ( $P=0.00002$ ). The frequency of associated anomalies had no significant changes over two periods of study ( $P=0.471$ ) (Table 1). Congenital heart disease and imperforate anus were detected in 6.09 and 5.05 of patients, respectively. Mortality was decreased over two periods of study, but it was not statistically significant (Table 1). The results of comparisons between four types of anomalies are shown in Table 2. Age at the second surgery was significantly higher in type D patients ( $P < 0.05$ ) (Table 2).

## Discussion

In our study, the mortality rate was 54.4 and 47.5% during the first and second periods of study. In another retrospective study from Iran, the mortality rate was 56% [5]. Mortality rate was similar in two studies. In a study by Snajdauf *et al.* [6], the mortality rate was 20.2%. In the early 1950s, the survival rate was about 57.6% in the report of Waterson *et al.* [7]. In the study by Beasley *et al.* [8], the mortality rate was decreased to 1% in the second decade of the study. In the study by

Seo *et al.* [9] on 97 patients between 1990 and 2007, the mortality rate was 24%. In the study by Peyvasteh

*et al.* [10], from April 1999 to March 2000, the mortality rate was 28.38%. In another study by the authors, the mortality rate was decreased from 75% (1993–1996) to 58.82% (2002–2005) [11]. In study by Orford *et al.* [12], the survival rate was increased from 70% (1970s) to 78% (1990s). O'Neill *et al.* [13] reported 93% surveillance among 53 neonates with esophageal atresia and tracheo-esophageal fistula. In the study from Sweden, the total mortality rate decreased from 33% (1969–1977) to 9% (1978–1984) [14]. Most of the studies showed increased survival rate following surgical repair of atresia [11,12,14]. In the study from China, 12.5% mortality rate was observed in 48 neonates with esophageal atresia [15]. In our study, decreased mortality rate following surgery was observed as seen in other studies [14], but the mortality rate in our hospitals was higher than that in other studies [12,13]. However, decreasing mortality rate in our study may be because of increasing number of patients admitted to ICU for better care. This high rate of mortality in our study may be due to limited equipments of NICU. In addition, different mortality rates may be the results of different birth weight and associated anomalies.

In our study, 50.5% of patients were male and 49.5% were female. There was a slight male predominance in the literature [16]. In the study by David and O'Callaghan [16], 53.9% of patients were male and 46.1% were female. In the study by Engum *et al.* [17], of the 227 infants 127 (55.94%) were male and 100 (44.06%) were female. In the study from northeast of China [15], of the 48 infants 33 were boys and male/female ratio was 2.2, which is significantly different from previous studies. In most studies, male is the predominant sex.

Type C atresia was the most common type of anomaly. Our findings were similar to those of Little *et al.* [18]. In the study by Yang *et al.* [19], 80% of patients had

**Table 1 Sex, type of atresia, and associated anomalies over two periods of study**

	n (%)			P-value
	Total	First period	Second period	
Sex				
Male		44 (55.7)	56 (47.1)	0.249
Female		35 (44.3)	63 (52.9)	
Type of atresia				
A	7 (3.53)	3 (3.80)	4 (3.35)	0.524
B	2 (1.01)	1 (1.26)	1 (0.85)	
C	185 (93.44)	72 (91.14)	113 (94.95)	
D	4 (2.02)	3 (3.80)	1 (0.85)	
Detected anomalies	109 (55.05)	46 (58.22)	63 (52.94)	0.471
Number of surgery				
0	11 (5.55)	2 (2.53)	9 (7.56)	0.188
1	103 (52.02)	39 (49.36)	64 (53.79)	
2	84 (42.42)	38 (48.10)	46 (38.65)	
Gastrostomy				
Yes	157 (79.29)	71 (89.87)	86 (72.27)	0.002
No	41 (20.71)	8 (10.13)	33 (27.73)	
Gastrostomy + TEF repair + esophagoesophagostomy surgery				
Yes	112 (57.14)	42 (53.84)	70 (59.32)	0.464
No	84 (42.86)	36 (46.16)	48 (40.68)	
Cervical esophagostomy				
Yes	12 (6.09)	7 (8.86)	5 (4.23)	0.228
No	185 (93.91)	72 (91.14)	113 (95.76)	
Mortality				
Yes	99 (50)	43 (54.43)	56 (47.05)	0.384
No	99 (50)	36 (45.57)	63 (52.95)	
Packed cell infusion				
0	97 (49.74)	42 (53.85)	55 (47.41)	0.694
1	75 (38.47)	28 (35.89)	47 (40.52)	
2	22 (11.28)	8 (10.26)	14 (12.07)	
3	1 (0.51)	0	0	
Number of NICU admission				
Yes		48 (60.75)	103 (86.55)	0.00002
No		31 (39.25)	16 (13.45)	
Duration of NICU admission	5.47 ± 5.38	5.31 ± 6.16	5.54 ± 5.00	0.807
Duration of hospital admission	12.92 ± 10.24	13.04 ± 11.70	12.84 ± 9.19	0.895

NICU, neonatal intensive care unit; TEF, tracheoesophageal fistula.

**Table 2 Sex distribution, associated anomalies, and demographic features of different types of atresia**

	A	B	C	D	P
Sex					
Male	4 (57.14)	1 (50)	94 (50.81)	1 (25)	0.760
Female	3 (42.86)	1 (50)	91 (49.19)	3 (75)	
Detected anomalies	3 (42.86)	2 (100)	103 (55.68)	1 (25)	0.315
Number of surgery					
0	0	0	11 (5.95)	0	0.748
1	5 (28.58)	2 (100)	94 (50.81)	2 (50)	
2	2 (71.42)	0	80 (43.24)	2 (50)	
Gastrostomy					
Yes	6 (85.71)	2 (100)	147 (79.46)	2 (50)	0.425
No	1 (14.29)	0	38 (20.54)	2 (50)	
Gastrostomy + closure of TEF + esophagostomy surgery					
Yes	1 (14.29)	0	108 (59.01)	3 (75)	0.034
No	6 (85.71)	2 (100)	75 (40.98)	1 (25)	
Cervical esophagostomy					
Yes	4 (57.14)	0	8 (4.35)	0	NA
No	3 (42.86)	2 (100)	176 (95.65)	4 (100)	
Packed cell transfusion					
0	4 (57.14)	2 (100)	90 (49.18)	1 (33.33)	0.877
1	3 (42.86)	0	71 (38.8)	1 (33.33)	
2	0	0	21 (11.47)	1 (33.34)	
3	0	0	1 (0.55)	0	
Age at 1st surgery	2.57 ± 787	2.00 ± 1.414	3.41 ± 2.941	7.00 ± 6.245	0.628
Age at 2nd surgery	7.50 ± 0.707	–	9.69 ± 5.658	14.00 ± 16.513	0.000
Duration of NICU stay	4.67 ± 2.88	3	5.45 ± 5.41	9.5 ± 7.77	0.709
Duration of hospital stay	9.57 ± 5.94	3.00 ± 1.41	13.13 ± 10.44	14.00 ± 4.89	0.435

NA, not applicable; NICU, neonatal intensive care unit; TEF, tracheoesophageal fistula.

type D atresia. In the Deurloo *et al.*'s [20] study, between 1947 and 2000, 88.7% of the 371 patients had type C atresia. In Zhang *et al.*'s [15] study, most of the patients were Gross type C. The results of studies using the Gross classification were similar [1,15].

The age at the first surgery in type D esophageal atresia was significantly higher than that in other types. It is expected because there is usual occurrence of respiratory infection with this type [1].

In our study, there were seven patients with type A esophageal atresia. Of the patients, six (85.71%) and four (57.14%) patients underwent gastrostomy and cervical esophagostomy, respectively. In our study, 50% mortality rate was observed over both periods of study. In the study by Burjonrappa *et al.* [21], 15 patients were with type A esophageal atresia, 13 patients underwent gastrostomy as the initial procedure, and two underwent the Foker procedure. All patients were alive following treatment. Outcome of our patients with type A esophageal atresia was less than that in the study by Burjonrappa *et al.* [21].

In our study, the rate of using gastrostomy was significantly decreased during the second period of study. Technically, gastrostomy and cervical esophagostomy were used for long gap patients [22]. In addition, we used it for ill neonates when anastomosis was technically impossible. Yen *et al.* [23] concluded that routine gastrostomy is contraindicated when primary anastomosis is feasible. Early diagnosis and treatment may be the reason for decreasing use of gastrostomy in our patients. Another reason for this decline in the number of gastrostomy procedures may be the decreasing number of newborn with long gap atresia. However, in this study, we had no reliable data regarding newborn with long gap atresia to draw a conclusion. The number of patients who underwent cervical esophagostomy was decreased in our study.

In our study, cervical esophagostomy was performed for 6.09% (12/197) of patients. In the study by Peyvasteht *et al.* [24], 6.75% of patients underwent cervical esophagostomy. In our study, gastrostomy was used for 79.29% of patients but in Peyvasteht *et al.*'s [24] study, gastrostomy was performed for 17.56% (13 of 74) patients. In addition, the mortality rate (50%) in our study was higher than that in Peyvasteht *et al.*'s [24] study. This difference may be due to difference in the condition of patients. It appears that our patients had more severe illness than their patients.

In our study, of the 198 patients 109 (55.05%) patients had associated congenital anomalies. Congenital heart disease and gastrointestinal anomalies were the most frequent associated anomalies. The frequency of cardiac anomalies in our study was less than that in the Chittmitrapap *et al.*'s study [25]. Associated congenital anomalies were detected in 47.8% of patients in the study from Saudi Arabia [26]. In the Manning *et al.*'s [27] study, cardiovascular and gastrointestinal anomalies were the most common associated major anomalies. The most common minor anomalies were limb defect and skeletal anomalies [27]. Cardiac anomalies, imperforate anus, limb anomalies, and chromosomal anomalies were the

most common anomalies in the study by Zhang *et al.* [15]. Overall rate of anomalies in our study was similar to other studies [28]; however, there was difference between studies regarding the rate of each anomaly.

## Conclusion

Mortality rate has decreased in our hospitals, but it remains higher than other studies. The rate of gastrostomy decreased during the second period of study. The rate of NICU admission was significantly higher during the second period of study. Overall rate of anomalies in our study was similar to other study. However, mortality has decreased in our hospitals, but the mortality rate in our country is higher than in other countries. Our experiences and equipments should be improved to lessen the mortality.

## Limitation

This study is retrospective and we had no reliable data regarding anomalies.

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## Conflicts of interest

There are no conflicts of interest.

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