Esophageal atresia: our experiences in a university hospital

Shahnam Askarpour[a], Mohammad-Hassan Hafezi[a], Hazhir Javaherizadeh[b] and Mostafa Alavi[a]

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 χ^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.**

Annals of Pediatric Surgery 2014, **10**:31–34

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

[a]Department of Surgery, Imam Khomeini Hospital and [b]Department of Pediatrics, Abuzar Children's Hospital, Ahvaz Jundishapur University of Medical Sciences, Ahvaz, Iran

Correspondence to Shahnam Askarpour, MD, Department of Surgery, Imam Khomeini Hospital, Ahvaz Jundishapur University of Medical Sciences, 6193672166 Ahvaz, Iran
Tel/fax: +98 611 221 6504; e-mail: shahnam_askarpour@yahoo.com

Received 23 September 2012 **accepted** 21 February 2014

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 χ^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

DOI: 10.1097/01.XPS.0000445132.59981.8e

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.

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

	n (%)			
	Total	First period	Second period	*P*-value
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.

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 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 Peyvasteh *et al.* [24], 6.75% of patients underwent cervical esophagostomy. In our study, gastrostomy was used for 79.29% of patients but in Peyvasteh *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 Peyvasteh *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 Chittmittrapap *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.

Acknowledgements

This paper is issued from the general practitioner thesis of Mostafa Alavi (No. 269); and financial support was provided by Ahvaz Jundishapur University of Medical Sciences.

Conflicts of interest

There are no conflicts of interest.

References

1 Spitz L. Oesophageal atresia. *Orphanet J Rare Dis* 2007; **2**:24.
2 Shaw-Smith C. Oesophageal atresia, tracheo-oesophageal fistula, and the VACTERL association: review of genetics and epidemiology. *J Med Genet* 2006; **43**:545–554.
3 Waterston DJ, Carter RE, Aberdeen E. Oesophageal atresia: tracheo-oesophageal fistula. A study of survival in 218 infants. *Lancet* 1962; **1**:819–822.
4 Sharma AK, Shekhawat NS, Agrawal LD, Chaturvedi V, Kothari SK, Goel D. Esophageal atresia and tracheoesophageal fistula: a review of 25 years' experience. *Pediatr Surg Int* 2000; **16**:478–482.
5 Davari HA, Hosseinpour M, Nasiri GM, Kiani G. Mortality in esophageal atresia: assessment of probable risk factors (10 years' experience). *J Res Med Sci* 2012; **17**:540–542.
6 Snajdauf J, Kalousova J, Styblova J, Fryc R, Pycha K, Petru O, *et al.* Results of treatment of esophageal atresia. *Cas Lek Cesk* 2004; **143**:614–617.
7 Waterston DJ, Carter RE, Aberdeen E. Oesophageal atresia: tracheo-oesophageal fistula. A study of survival in 218 infants. *Lancet* 1962; **1**:819–822.
8 Beasley SW, Shann FA, Myers NA, Auldist AW. Developments in the management of oesophageal atresia and tracheo-oesophageal fistulas. *Med J Aust* 1989; **150**:501–503.
9 Seo J, Kim do Y, Kim AR, Kim DY, Kim SC, Kim IK, *et al.* An 18-year experience of tracheoesophageal fistula and esophageal atresia. *Korean J Pediatr* 2010; **53**:705–710.
10 Peyvasteh M, Askarpour S, Shoushtari MHS. A study of esophageal strictures after surgical repair of esophageal atresia. *Pak J Med Sci* 2006; **22**:269–272.
11 Peyvasteh M, Askarpour S, Javaherizadeh H, Fatahian T. Evaluation of epidemiologic indices of neonate's diseases in the Pediatric Surgery Ward of the Ahvaz Jundishapur University hospitals during the period 1993–1996 and 2002–2005. *Ann Pediatr Surg* 2011; **7**:7–9.
12 Orford J, Cass DT, Glasson MJ. Advances in the treatment of oesophageal atresia over three decades: the 1970s and the 1990s. *Pediatr Surg Int* 2004; **20**:402–407.
13 O'Neill JA Jr, Holcomb GW Jr, Neblett WW III. Recent experience with esophageal atresia. *Ann Surg* 1982; **195**:739–745.
14 Sillen U, Hagberg S, Rubenson A, Werkmaster K. Management of esophageal atresia: review of 16 years' experience. *J Pediatr Surg* 1988; **23**:805–809.

15 Zhang Z, Huang Y, Su P, Wang D, Wang L. Experience in treating congenital esophageal atresia in China. *J Pediatr Surg* 2010; **45**:2009–2014.
16 David TJ, O'Callaghan SE. Oesophageal atresia in the South West of England. *J Med Genet* 1975; **12**:1–11.
17 Engum SA, Grosfeld JL, West KW, Rescorla FJ, Scherer LR 3rd. Analysis of morbidity and mortality in 227 cases of esophageal atresia and/or tracheoesophageal fistula over two decades. *Arch Surg* 1995; **130**: 502–508, discussion 508–509.
18 Little DC, Rescorla FJ, Grosfeld JL, West KW, Scherer LR, Engum SA. Long-term analysis of children with esophageal atresia and tracheoesophageal fistula. *J Pediatr Surg* 2003; **38**:852–856.
19 Yang CF, Soong WJ, Jeng MJ, Chen SJ, Lee YS, Tsao PC, *et al*. Esophageal atresia with tracheoesophageal fistula: ten years of experience in an institute. *J Chin Med Assoc* 2006; **69**:317–321.
20 Deurloo JA, Ekkelkamp S, Schoorl M, Heij HA, Aronson DC. Esophageal atresia: historical evolution of management and results in 371 patients. *Ann Thorac Surg* 2002; **73**:267–272.
21 Burjonrappa S, Thiboutot E, Castilloux J, St-Vil D. Type A esophageal atresia: a critical review of management strategies at a single center. *J Pediatr Surg* 2010; **45**:865–871.
22 Louhimo I, Lindahl H. Esophageal atresia: primary results of 500 consecutively treated patients. *J Pediatr Surg* 1983; **18**:217–229.
23 Yen M-L, Wang N-L, Chang P-Y, Sheu J-C, Huang F-Y, Chen C-C. Routine gastrostomy is contraindicated in esophageal atresia. *Pediatr Surg Int* 1995; **10**:90–92.
24 Peyvasteh M, Askarpour S, Sarmast MH, Javaherizadeh H, Mehrabi V, Ahmadi J, *et al*. Esophageal atresia: comparison between survivors and mortality cases who underwent surgery over a 2-year period in two referral hospitals, Tehran, Iran. *Ann Pediatr Surg* 2012; **8**:42–44.
25 Chittmittrapap S, Spitz L, Kiely EM, Brereton RJ. Oesophageal atresia and associated anomalies. *Arch Dis Child* 1989; **64**:364–368.
26 Al-Salem AH, Qaisruddin S, Varma KK, Abusrair H, Al-Dabbous I, Al-Hayek R. Esophageal atresia and tracheoesophageal fistula. *Ann Saudi Med* 1997; **17**:481–484.
27 Manning PB, Morgan RA, Coran AG, Wesley JR, Polley TZ Jr, Behrendt DM, *et al*. Fifty years' experience with esophageal atresia and tracheoesophageal fistula. Beginning with Cameron Haight's first operation in 1935. *Ann Surg* 1986; **204**:446–453.
28 Myers N, Beasley S, Auldist A. Oesophageal atresia and associated anomalies: a plea for uniform documentation. *Pediatr Surg Int* 1992; **7**:97–100.