ORIGINAL ARTICLE

Review of Oesophageal Atresia and Tracheoesophageal Fistula in Hospital Sultanah Bahiyah, Alor Star. Malaysia from January 2000 to December 2009

S Narasimman, MMed Surg, M Nallusamy, FRCS, S Hassan, MMed Surg

Department of Surgery, Hospital Sultanah Bahiyah, Alor Star, Kedah

SUMMARY

Oesophageal atresia (EA) and tracheoesophageal fistula (TEF) is one of the congenital anomaly occurring in the newborns with the incidence of 1 in 2500 births seen worldwide. A retrospective review of newborns admitted to Hospital Sultanah Bahiyah (HSB) from 1st January 2000 to 31st December 2009 was done. The objective was to look at the influence of birth weight, time of surgical intervention, presence of other congenital anomaly and presence of preoperative pneumonia to the immediate outcome (mortality) of the surgery.

There were 47 patients with oesophageal atresia, out of which 26 (55%) were males and 21 (45%) females. The distribution of patients by race were 34 Malays (72%), 9 Chinese (19%) and 4 Indians (9%). The birth weight of the babies range from 0.8 kg to 4.0 kg and there was a significant association with the outcome of the surgery (p< 0.05). Most of the babies (20) were operated within 24 hours of presentation but there was no significant association to the outcome. 23 (49%) of them were born with congenital malformation and there was a significant association with the outcome of the surgery (p<0.05). Based on the chest roentgenogram, 20 (43%) of them had pneumonia with significant association with the outcome (p<0.05). The mortality rate is 23% and the causes of death were pneumonia (36%), renal failure (18%), cardiac malformation (18%) and multiple congenital malformations (28%). The outcome of EA and TEF is determined mainly by birth weight, congenital malformations and presence of preoperative pneumonia in HSB.

KEY WORDS:
Oesophageal atresia, tracheoesophageal fistula, outcome, congenital anomaly

INTRODUCTION

Oesophageal atresia (EA) is a congenitally interrupted oesophagus. Tracheoesophageal fistula (TEF) is a congenital or acquired communication between the trachea and oesophagus [1]. EA and TEF are among the commonest congenital anomaly occurring in the newborns, with the incidence of 1 in 2500 births [2]. The incidence worldwide is reducing in trend for unknown reasons [3].

The outcome depends on many factors, mainly the associated congenital anomalies with the TEF. Waterston and Spitz have suggested different classifications that will determine the prognosis of the patient [4]. These classifications are based on the birth weight, timing of surgery and associated cardiac anomaly. There is no racial predilection for this condition. EA and TEF are usually diagnosed very early in life. Currently, the survival rate is around 80 - 90% [5].

Identifying the relevant risk factors in the local centre will enable us to stratify the prognosis of the babies based on suitable prognostic criteria. This study will also provide a preliminary database for the cases performed in HSB in the past 10 years and to identify the problems in order to improve the management of EA/TEF in HSB.

OBJECTIVE

To determine the immediate outcome (mortality) and the association with birth weight, time taken for surgical intervention, presence of congenital anomaly and preoperative pneumonia.

MATERIALS AND METHODS

Cross sectional study with retrospective record review was carried out among babies with EA/TEF in the Paediatric Surgical Unit, Department of Surgery, HSB from 1st January 2000 to 31st December 2009 (10 years). All babies registered in HSB who was diagnosed with EA/TEF throughout the study period were included. Babies whom were diagnosed with EA/TEF but did not undergo surgery and those with incomplete or damaged records were excluded from this study. The sample size was calculated based on the single proportion formula with the precision of 0.05. Statistical significance was assigned to a p value < 0.05 with the confidence interval of 95%. Universal sampling technique was applied. A prepared data collection sheet was used to collect the relevant information.

RESULTS

Demography

There were 52 patients with EA admitted to HSB from January 2000 to December 2009. The response rate was 91%, giving the total number of 47 patients with EA in this study.

This article was accepted: 22 November 2012
Corresponding Author: S Narasimman, Department of Surgery, Hospital Sultanah Bahiyah, Alor Star, Kedah
Email: drnara@hotmail.com

Out of the 47 patients, 26 (55%) were males and 21 were females (45%). There was no significant relationship between sex and outcome (X^2 = 1.761, df = 1, p = 0.185). The death rate for males (31%) was higher than females (14%).

The distribution of patients by race were 34 (72%) Malays, 9 (19%) Chinese and 4 (9%) Indians. There are no significant racial predispositions of EA/TEF worldwide [1, 19].

Maternal age ranges between 19 to 50 years with the mean age of 32 years old. There was no significant relationship between maternal age and the outcome of the babies (X^2 = 2.332, df =1, p = 0.112).

There were 16 (34%) premature babies out of the 47 babies diagnosed with EA. The mortality risk for premature babies was 43% as compared to 13% for the term babies and there was a significant relationship between them (X^2 = 5.601, df =1, p = 0.018). This is because premature babies have increased rate of complications generally as compared to term babies [3].

Types of EA/TEF

The types of EA/TEF were described by Gross and Boston in 1953[21]. There were only 3 different types of EA/TEF seen in this study. Out of the 47 babies, 4(9%) were Type A, 41(87%) were Type C (the commonest type) and 2(4%) were Type E. Type B and Type D EA/TEF were not seen in this study. There was no significant relationship between the types of TEF/EA and the outcome of the patient (X2 = 3.130, df =2, p = 0.209)(Figure 1).

Polyhdromnios was noted to be present in 75% of the Type A EA.

Birth weight

The birth weight of the babies range from 0.8 kg to 4.0 kg with the smallest surviving baby weighing 1.1 kg. The mean and median birth weight in these babies was 2.2 kg. Most of the babies, 28 of them were between 1.8 to 2.5 kg. There were 9 of them weighing less than 1.8kg and 10 of them with the weight more than 2.5kg. There was a significant relationship between the birth weight of the babies and the outcome of the surgery (X^2 = 8.306, df = 2, p = 0.016)(Figure 2).

Time taken for surgery

The time taken for surgical intervention for EA, either single stage or staged procedure for the babies were divided to within 24 hours of birth, 24 to 48 hours of birth and more than 48 hours of birth. Twenty (43%) of the babies with EA were operated within 24 hours of birth, 16(34%) babies were operated within 48 hours of birth and 11(23%) babies were operated in more than 48 hours of birth. There was no significant association between the time taken for surgical intervention and the outcome (X^2 = 0.314, df = 2, p = 0.855).

Congenital Malformations

Out of 47 babies, 23 (49%) of them were born with associated congenital malformation. The types of congenital malformations were referred to the VACTERL associations. Vertebral anomalies were present in 5(10%) babies, anorectal malformation in 13(28%) babies, cardiac anomalies in 14(30%) babies, renal malformation in 9 (19%) babies and limb dysplasia in 8(17%). Out of 23 babies with congenital malformation, 9 of them were found to have VACTERL syndrome, which is 19% of the total number of babies with EA/TEF. See Table 1. There was a significant association between the incidence of malformation in the babies and the outcome of the surgery (X^2 = 6.214, df = 1, p = 0.013). There was 39% mortality rate among the babies with TEF and EA and other associated congenital anomalies.

Table I: Distribution of types of congenital malformations in patients with EA/TEF in HSB from Jan 2000 to Dec 2009

Malformation type	Incidence	Percentage
Vertebral	5	28%
Hemivertebra	2	
Hypoplastic	0	
Fused vertebra	2	
Tethered cord	2	
Anorectal	13	28%
High	8	
Low	5	
Cardiac	14	30%
PDA	2	
VSD	7	
PDA + VSD	3	
ASD	2	
Renal	9	19%
Agenesis	2	
Horseshoe	3	
Polycystic	4	
Limb	8	17%
Syndactily	2	
Polydactily	3	
Radial aplasia	3	

Preoperative diagnosis of pneumonia and ventilation

Twenty of the babies (43%) had preoperative pneumonia and 8 died, as compared to 3 deaths among those without pneumonia. There was a significant relationship between preoperative pneumonia and the outcome (X^2 = 5.349, df = 1, p = 0.021).

Types of surgery and complications

Out of the 40 babies whom underwent primary repair, 8 died (20%). Whereas, 7 babies underwent staged repair and 3 died (43%). All the surgeries were performed via right thoracotomy, with transverse inferior axilla incision for the division of the fistula and anastomosis. A single layered anastomosis was performed using absorbable monofilament polydioxanone (PDS II) suture. There was no significant relationship between the outcome of babies in the primary and staged surgery group (X^2 = 1.736, df = 1, p = 0.188).

The complications seen were anastomotic leak (12 babies), anastomotic stricture (7 babies) and gastroesophageal reflux (8 babies). These complications were diagnosed based on clinical symptoms and radiological imaging. All the complications were treated non-operatively with continuous observation and transanastomotic feeding for leak, oesophageal dilatation for stricture and proton pump inhibitor for reflux.

Table II: Comparison of survival rate of patients with EA/TEF based on Waterston Classificaton

Waterston Classification	Spitz 2006 (London)	Azim 1999 (HUSM, Kelantan)	HSB 2009 (Alor Star, Kedah)
Group A	99%	100%	100%
Group B	93%	71.4%	89%
Group C	71%	51.9%	33%

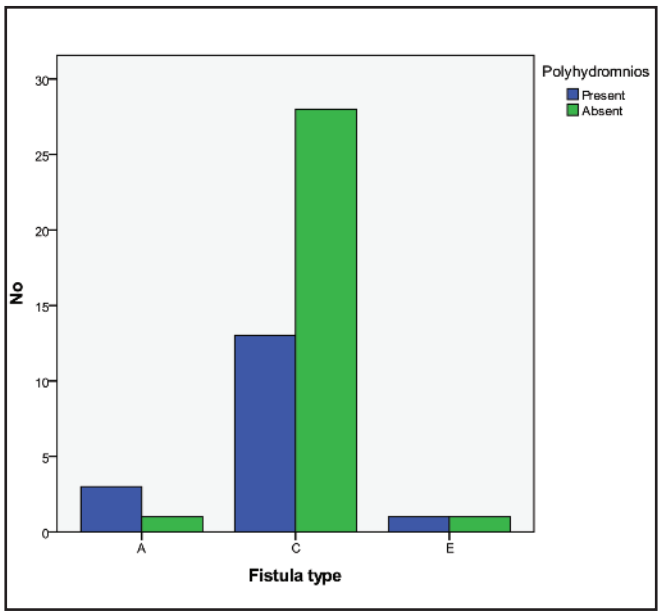

Fig. 1: Distribution of fistula type as compared to polyhydromnios of EA patients in HSB from Jan 2000 to Dec 2009.

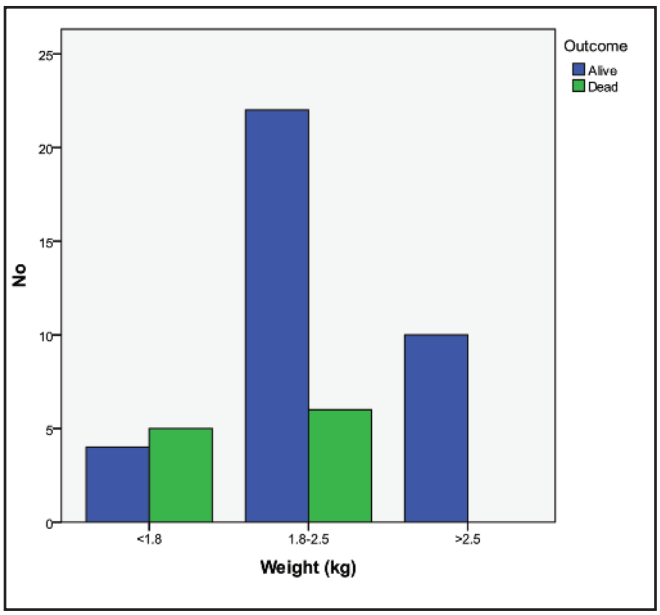

Fig. 2: Comparison of birth weight and outcome of EA/TEF patients in HSB from Jan 2000 to Dec 2009.

Mortality rate

The number of deaths in this study was 11 out of the 47 babies operated, which gives a mortality rate of 23%. The causes of death were pneumonia (4), renal failure (2), cardiac malformation (2) and multiple congenital malformations (3).

DISCUSSION

There was a male preponderance (1.2 : 1) in this study. This was also shown in other larger studies worldwide. Depaepe et al. and Nawaz et al reported similar findings in their study[6,7]. There is no explanation for the male preponderance, but similar pattern is also seen in other congenital anomalies like Hirschsprung's disease and pyloric stenosis[7].

The influence of maternal age on EA/TEF has been variously reported to be U-shaped (higher rates among both older and younger mothers) or increasing with greater maternal age[6].

The birth weight of the babies in this study has got a significant relationship with mortality. This could be the reason for the observation of significant relationship of preterm babies and mortality as preterm babies will generally be low in birth weight[8]. Sharma et al. reported that there was a 50% reduction in the mortality rate as the birth weight of the babies increased in different phases of his studies[9]. However, Poenaru from Montreal suggested that birth weight is not a significant factor affecting mortality as compared to presence of pneumonia[1]. Spitz *et al.* also believes that low birth weight is not a contraindication for primary surgical repair to be carried out[2].

Although there was no significant relationship seen between time of surgical intervention and place of birth, it is seen in developing countries that the most important factor influencing the outcome is the duration taken for the baby to reach the tertiary centre, especially in India[10].

There was a significant relationship between presence of other congenital anomalies and mortality. Keckler et al. observed that the wide spectrum of the VACTERL association of EA/TEF starts early in the embryogenesis[11]. Babies with EA/TEF with associated two or more malformations have a significant drop in the survival rate as compared to those with minor anomalies or none[12].

Presence of preoperative pneumonia was found to be significantly associated with mortality. In a study of 500 consecutive babies treated for TEF and EA, the single most important factor improving survival in those babies is the prevention of pneumonia[13]. Davari *et al.* on the other hand believes that with modern medical care and facilities available, presence of other congenital malformation and birth weight plays more important role in determining the outcome[14].

Agarwala *et al.* reported a mortality rate of 80% in India, which is higher than this study [10]. This could be due to the remote location of health centres in India with delayed identification of the severity in the presentation and referral to the tertiary centre.

The mortality rate in this study was 23%. Based on Waterston classification, Spitz observed in his series of Group C babies had the survival of 71%, as shown in Table II [15]. This is much higher if compared to series by Azim in Kelantan, Malaysia and by us [16]. This could be due to the advancement of paediatric care in the developed country. Based on the observation by Azim in Kelantan from 1990 to 2000 and by us in this study, it is shown that the mortality rate of babies with EA/TEF in Malaysia over the period of 20 years has been static [16]. Although this is not a direct comparison, it generally has some reflection of the paediatric services in terms of management of EA/TEF in our country and a multicentre study needs to be conducted to clarify this.

CONCLUSION

We found the birth weight, preoperative pneumonia and congenital malformation significantly influence the outcome of the babies after surgery for EA/TEF. The mortality rate of 23% is in keeping with worldwide figures.

This study is only observing the patients over a 10-year period. A larger sample size is needed to get better correlation and associations. This study should be continued and reviewed every 5 years. A multicentre study should be carried out to get a better picture of the paediatric care in Malaysia.

REFERENCES

1. Poenaru D. A new prognostic classification for oesophageal atresia. Surgery 1993; 113(4): 426-32.
2. Spitz L, Kiely E, Brereton R.J and Drake D. Management of Esophageal Atresia. World Journal of Surgery 1993; 17: 296-300.
3. Dave S, Bajpai M, Gupta D.K, Agarwala S, Bhatnagar V and Mitra D.K. Oesophageal Atresia and Tracheo-Esophageal Fistula, A Review. Symposium: Neonatal Surgery Indian Journal of Paediatrics 1999; 66: 759-72.
4. Tsai JY, Berkery L, Wesson DE, Redo SF and Spigland NA. Esophageal Atresia and Tracheoesophageal Fistula: Surgical Experience Over Two Decades. Ann Thorac Surg 1997; 64: 778-83.
5. Deurloo JA, Rien de Vos, Ekkelkamp S, Heij HA and Aronson DC. Prognostic factors for mortality of oesophageal atresia patients: Waterston revived. European Journal of Paediatrics 2004; 163: 624-5.
6. Depaepe A, Dolk H and Lechat MF. The Epidemiology of Tracheoesophageal Fistula and Oesophageal Atresia in Europe. Archives of Disease in Childhood 1993; 68: 743-8.
7. Nawaz A, Matta H, Shawiz R, Jazcobsz A, Kassir S and Al Salem H. Oesophageal Atresia and Tracheoesophageal Fistula: Succes and Failure Rates in United Arab Emirates. Paediatric Surgery International 1998; 14: 214-7.
8. Beck S, Wojdyla D, Say L, Betran A P, Merialdi M, Requejo J H, Rubens C, Menon R & Van Look PF. The worldwide incidence of preterm birth: a systematic review of maternal mortality and morbidity. Bulletin of the World Health Organization 2010; 88: 31-8.
9. Sharma AK, Shekhawat NS, Agrawal LD, Chaturvedi V, Kothari S.K, Goel D. Esophageal Atresia and Tracheoesophageal Fistula: A Review of 25 Years Experience. Pediatric Surgery International 2000; 16: 478-82.
10. Agarwala S, Bhatnagar V, Bajpai M, Gupta DK, Mitra D.K. Factors contributing to poor results of treatment of oesophageal atresia in developing countries. Paediatrics Surgery International 1996; 11: 312-5.
11. Keckler SJ, St. Peter SD, Valusek PA, Tsao KJ, Snyder CL, Holcomb GW and Ostlie DJ. VACTERL anomalies in patients with oesophageal atresia: an updated delineation of the spectrum and review of the literature. Pediatr Surg Int 2007; 23: 309-13.
12. Encinas JL, Luis AL, Avila LF, Martinez L, Guereta L, Lassaletta L and Tovar J.A. Impact of preoperative diagnosis of congenital heart disease on the treatment of oesophageal atresia. Paediatrics Surgery International 2006; 22: 150-3.
13. Louhimo I and Lindahl H. Oesophageal atresia: Primary results of 500 consecutively treated patients. Journal of Paediatric Surgery 2006; 18 (3): 217-29.
14. Davari H, Esfandiari R, Talaei M. Surgical Outcomes in Oesophageal Atresia and Tracheoesophageal Fistula: A Comparison between Primary and Delayed Repair. Journal of Research Medical Sciences 2006.; 11(1): 57-62.
15. Spitz L, Kiely E and Brereton R J Esophageal atresia: Five year experience with 148 cases. Journal of Pediatric Surgery 2006; 22 (2): 103-8.
16. Azim A. Management of esophageal atresia in Hospital Universiti Sains Malaysia from January 1991 to July 1999. Universiti Sains Malaysia 1999.