FACTORS AFFECTING SURVIVAL IN PATIENTS WITH OESOPHAGEAL ATRESIA AND TRACHEO-OESOPHAGEAL FISTULA

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Objective: To evaluate the various factors affecting survival in babies with oesophageal atresia and tracheo-oesophageal fistula. **Design:** Descriptive study. **Setting:** The study was carried out at the Department of Paediatric Surgery, The Children's Hospital, Pakistan Institute of Medical Sciences (PIMS), Islamabad from March 2004 to March 2005. **Patients and Methods:** All neonates with oesophageal atresia (EA) and tracheo-oesophageal fistula (TEF) during the study period were included in the study. Patients having isolated EA were excluded. A total of 80 patients were included in the study. Patients were received from the emergency department, OPD and Neonatal ICU. Diagnosis was confirmed by passing a radio opaque orogastric tube. Investigations were done to look for other associations. After stabilisation, right thoracotomy was performed, fistula was ligated and divided. An attempt was made to do a primary oesophago-oesopahgostomy. Nasogastric feeding was started on 2nd post-operative day. A contrast oesophagogram was performed on the 7th postoperative day and having ruled out leak, oral feeding was started. **Results:** Out of the total, 33 (41%) survived and 47 (58%) patients died. Out of 47 deaths 20 (25%) died before surgery and 27 (34%) died after surgery. Mean follow up period was 6 months. Sixteen (20%) patients had anastomotic leak, 24 (30%) had anastomotic stricture, and 64 (80%) patients had postoperative pneumonia. **Conclusion:** We conclude that proper antenatal check ups will detect the problem early, avoid home deliveries and hence improve survival. Pneumonitis and septicaemia significantly affect survival. Availability of ICU is one of the main determinants of survival. The likely cause of high mortality rate in pre-operative patients in our series is non-availability of NICU due to limited space in our setup.

Keywords: Oesophageal atresia, tracheo-oesophageal fistula, survival, neonates

INTRODUCTION

Various factors affect survival of babies with Oesophageal atresia (EA) and tracheo-oesophageal fistula (TEF). These are antenatal diagnosis and timely referral to the place where facility for treating such babies is available, birth weight and gestational age, early referral after the diagnosis, pulmonary status at the time of admission, septicaemia, and associated congenital abnormalities especially cardiac. Other factors are availability of ICU care, need for ventilatory support and technical problems like long gap, need for staged repair rather than single stage repair.

The objective of this study was to evaluate the various factors affecting survival in babies with EA and TEF.

PATIENTS AND METHODS

This study was conducted in the department of paediatric surgery, The Children's Hospital, PIMS, Islamabad from March 2004 to March 2005. A total of 80 neonates were included in the study.

All neonates who had EA and distal TEF were included in the study. Those having isolated EA were excluded. Patients were received from the emergency department as well as from the OPD. Some were referred from other hospitals. And yet few patients were taken directly from the NICU. The main symptom was drooling of saliva, frothing from mouth and respiratory difficulty. At the time of first presentation a stiff NG tube of Fr 8 size impregnated with radio opaque contrast solution was inserted in each patient and an x- ray taken to confirm the diagnosis. Special attention was given to look for gas shadows in the abdomen to rule out isolated EA. After confirming the diagnosis, initial resuscitation was done. A continuous pharyngeal suction was instituted and the babies were put on regular chest physiotherapy, broad spectrum antibiotics, intra venous fluids and oxygen therapy with or without assisted ventilation where required. After stabilization of the patient, surgery was performed. A right thoracotomy through the 4th space was performed in all patients with an extra plural approach. The azygos vein was ligated and divided. The fistula was transfixed and upper pouch was searched. In those patients who remained stable during surgery the procedure was continued; the fistula was divided and an attempt was made to do a primary repair with interrupted single layer 5/0 vicryl sutures over a stent (Fr 6 feeding tube). A chest drain was left in the extra pleural space and attached to an underwater seal bottle. Those patients who remained unstable during surgery or those who had a long gap, only fistula was transfixed without dividing it and cervical oesophagostomy and feeding gastrostomy was performed. Post operatively all the babies were kept in the NICU and later shifted to the ward after complete stabilisation. Feeding was started through the same NG tube after 48 hour. Chest tube was removed on the 5th

post operative day if there are no signs of leak. A contrast oesophagogram was performed on the 7th postoperative day and if there is no leak then oral feeding was established gradually reaching to the full requirement as per tolerance of the baby. A weekly follow up was advised for the first month followed by two weekly follow up for next two months and then monthly follow up for 6 months. Contrast study in the follow up was done only in those who had dysphagia or recurrent respiratory infection.

RESULTS

A total 80 patients were received and managed. Out of them 53 were male and 27 were female. Mean follow up was 6 months. Antenatal diagnosis was made in 2 (2.5%) and Polyhydramnios was seen in 11 (14%). Out of the total 18 (21%) babies were delivered in the hospitals and 62 (78%) were home delivered. Pre term babies were 2 (2.5%).

Five (6.2%) patients had syndromic/ dysmorphic appearance, 1 (1.2%) had polycystic kidney and 10 (12.5%) had anorectal malformation. Hydrocephalus was seen in 1 (1.2%), congenital heart disease in 5 (6.2%) and one (1.2%) patient had triple atresia (Oesophageal, duodenal and rectal). Limb defects were seen in 3 (3.8%), un-descended testes in 3 (3.8%), cervical hemivertebra in 1 (1.2%), meningocoele in 5 (6.2%), and pinna deformity, eye deformity, mouth diverticula was seen each in 1 (1.2%).

Primary anastomosis could be done in 52 (65%). Two (2.5%) patients required circular myotomy. In 6 patients (7.5%) the gap was wide; so the fistula was ligated only without dividing and cervical oesophagostomy and feeding gastrostomy was performed.

Table-1: Body weight of babies

Body weight	Number of patients (n=80)	%
<1.5 Kg:	4	(5 %)
2.5 Kg:	42	(52 %)
>2.5 Kg:	34	(42 %)

Table-2: Age at arrival to our hospital

Age at arrival:	Number of patients (n=80)	%
0-48 hours:	35	44%
48 hours-7days:	26	33%
7-12 days:	14	17%
>12 days:	5	6%

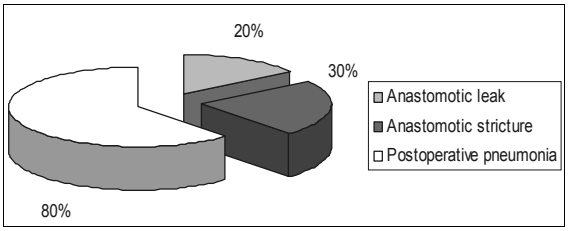

Figure-1: Post operative complications

Out of the total 80 patients 33 (41%) survived and 47 (58%) died. The deaths which occurred before the surgery were 20 (25%) and deaths after surgery were 27 (34%).

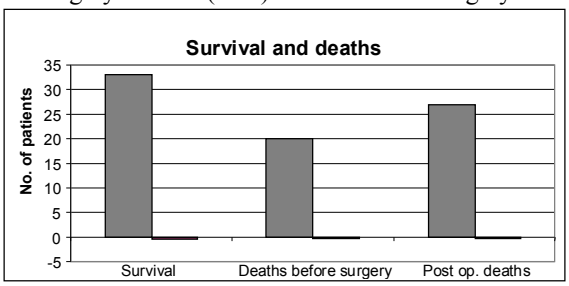

Table-3: Final outcome

Table-4: Stay at other hospitals before referral to us

Stay at other hospitals	No. of patients	%
1–2 days	12	15%
3–4 days	6	7.5%
5–6 days	2	2.5%
10 days	2	2.5%

Table-5: Stay in our ward before the surgery

Pre operative stay at ward	No. of patients	%
1 day	15	18.8%
2 days	10	13%
3 days	11	14%
4 days	6	7.5%
5 days	7	8.8%
6 days	8	10%
7 days	5	6%
8 days	3	3.8%
Directly admitted to NICU	15	18.8%

Table-6: Pre-operative problem adversely affecting survival

Pre operative problems	No. of patients	%
Feeding already given	(n=64)	80%
Pneumonia	(n=64)	80%
Septicemia	(n=48)	60%
Hypothermia	(n=56)	60%

Table-7: Assisted ventilation required

	No. of Patients	%
Pre-Operative	9	11
Post-operative	18	22
None	53	66

DISCUSSION

Oesophageal atresia with TEF is invariably associated with a certain degree of morbidity and mortality. There are different reasons for these complications. The pathology itself is simple and straight forward but may be associated with serious congenital malformations especially congenital heart diseases and acquired lung conditions.

In our set up patients reach the hospital very late and hence have already established acid pneumonitis and septicaemia. Secondly the facility of cardiac assessment in newborn babies is only scarcely

available in limited centres due to which cardiac associations may go unnoticed.

Our set up is the referral centre for the northern Punjab, NWFP and the Northern Areas of Pakistan, and Azad Jammu and Kashmir. Due to such a large catchment area we see this pathology more frequently. In one year time from March 2004 to March 2005 we have received a total of 80 patients of EA and TEF with a male to female ratio about 2:1. Most of the patients came to the hospital after 2–3 days of birth. One patient came even after 22 days of birth.

Only 22 (27.5%) patients in our series had a referral letter with them. All the rest were walk-in patients with no proper referral.

The mean gestational age was 36 weeks, which is comparable to international data.[1,2] The average birth weight was 2.45 Kg. Other studies show more or less similar data.[3,4]

Most of the babies have been managed in the treatment room of surgical ward with no facility for care of the new born babies. Forty of the total patients (50%) had to wait in the surgical ward for more than 48 hours before they could find a place in the NICU. This is due to the limited capacity and workload on the NICU. Fifteen patients (18.7%) were already in NICU.

On the total, 36 (45%) patients had other associations; 10 (12.5%) patients had anorectal malformation and one patient had polycystic kidney, 5 (6.2%) patients had congenital heart disease. One had triple atresia (Oesophageal, duodenal and Rectal). Three patients (3.8%) had limb defects and another 3 (3.8%) had un-descended testes. One had cervical hemivertebrae and 5 (6.2%) had meningocoele. All patients were full term except 2 (2.5%) who were preterm.

Amongst the total, 12 (15%) stayed in other hospitals for 2 days before they were referred to us. Two (2.5%) stayed for ten days before referral. This shows the lack of awareness in the medical community to diagnose the condition in time and subsequently refer the patient to a proper tertiary care facility.

In 64 (80%) patients feeding has already been attempted with history of choking on feed. They had respiratory distress due to established pneumonia. Frank septicaemia was seen in 48 (60%) patients and 70% patients were already hypothermic.

In our study 8 (10%) patients had a long gap between the two pouches so circular myotomy was done in two (2.5%) in order to achieve length and in the rest of 6 (7.5%) patients the upper pouch was so high that primary repair could not be done. So only the fistula was ligated and cervical oesophagostomy and feeding gastrostomy was performed with a plan for delayed primary anastomosis.

In 21 (26%) patients primary anastomosis was performed. According to one observation even in extremely low birth weight and very low birth weight babies, primary repair of EA is technically feasible and offers a good treatment option. Staged repair can be avoided.[5]

Mean follow up was 6 months. In our series 16 (20%) patients had anastomotic leak, 24 (30%) had anastomotic stricture, 64 (80%) had postoperative pneumonia. We have not seen any case with recurrent fistula in our study. The figures can be compared with another study by Okada *et al* who reported anastomotic leakage (26.5%), anastomotic stricture (49.1%), postoperative pneumonia (57.0%) and gastro-oesophageal reflux (52.0%).[6]

Total deaths were 47 (58%) and the survival was 33 (41%) but out of the total of 80 patients 20 (25%) died before surgery. The reason for high mortality is that most of the patients (78%) were home delivered and were definitely attempted feeding which led to pneumonia. These were the ones who came late and already had severe respiratory complications and septicaemia. Amongst the rest of 60 patients who were operated 27 (34%) died. These babies cannot be managed in the general ward. They need ICU care especially those who require assisted ventilation. The high mortality before surgery is due to non-availability of bed in NICU.

To evaluate the outcome and prognosis in patients with EA and TEF, several classification systems and prognostic groupings have been devised. These systems are Waterston classification, Montreal classification, Bremen Classification and Spitz classification depending upon various variable.[7–9]

Since 1962, the Waterston classification has been used to stratify neonates who have EA and/or TEF into prognostic categories based on birth weight, the presence of pneumonia, and the identification of other congenital anomalies. In response to advances in neonatal care, the surgeons from the Montreal Children's Hospital proposed a new categorisation system in 1993 in an attempt to define the current risk factors for patients who have EA and TEF. In the Montreal experience only two characteristics independently affected survival, i.e., preoperative ventilator dependence and associated major anomalies. Studies have been done to determine which system had the greatest validity and it was concluded that Montreal system more accurately identifies children at highest risk than the Waterston classification.[10]

The Spitz classification of EA based on the birth weight and the presence of a major cardiac anomaly was proposed 1994. It may be useful when counselling parents and in comparing outcome among centres. But this requires the facility for cardiac assessment of newborns.[11]

According to some authors it is the bodyweight at birth and the existence of pre-surgical

respiratory complications which have a significant impact on the survival (Waterston and Spitz classification).[12]

In our study 64 (80%) patients had already established respiratory infection and 48 (60%) patients had septicaemia which were the main contributing factors to morbidity and mortality.

Early antenatal diagnosis has also been proved to improve outcome in EA.[13] In our study only 11 mothers (14%) had antenatal ultrasound which showed polyhydramnios but only two patients had an antenatal diagnosis of EA. This shows that in our community most of the mothers do not get regular antenatal check ups. The sensitivity of ultrasound in the diagnosis of EA is extremely variable as this investigation is subjective and depends upon the expertise of the sonologist and the fact that how much a sonologist is familiar with the pathology. Moreover other anomalies are also picked up on antenatal ultrasound.[14]

Anastomotic complications after primary repair are the main complications. Several manoeuvres have been attempted to take care of this problem. Postoperative elective ventilation support (PEVS) under paralysis with neck flexion after primary repair of EA has been reported to reduce anastomotic leakage effectively.[15]

In one study mortality was 30.7% and the causes were divided into avoidable causes and unavoidable causes. Possible avoidable causes of mortality were primary sepsis, technical problems and severe pneumonia. The unavoidable causes, they reported were major congenital heart disease and anomalies incompatible with life and hence they concluded that primary sepsis and sepsis acquired during hospitalisation were the main causes of mortality besides major or life threatening anomalies, long gaps and major leaks.[7]

Low birth weight (<1500 g) and associated cardiac pathology necessitates special anaesthesia considerations.[16]

In another study operative mortality was reported 30.8% with the incidence of aspiration pneumonia 39.4% and other associated anomalies 49%. In that study sepsis was the most frequent cause of death after associated anomalies, prematurity, delay in diagnosis leading to increased incidence of aspiration pneumonia and a shortage of qualified nurses.[4]

In most of the studies the most frequent surgical complication was anastomotic leak and septic complications with an overall mortality of 20%. It is not the long gap between proximal and distal pouch of EA which significantly affect survival but there are some other factors like birth weight below 2000 g, cardiac anomalies, and need for mechanical ventilation before the operation. They recommend that attempts at primary anastomosis even in children in sub optimal general condition could decrease mortality.[17]

Ventilator dependence shows a poor physiologic status of the neonate and is considered to be the most reliable prognostic risk factor.[18]

Similarly cardiac disease and multiple abnormalities carry a substantial increased risk of mortality.[19]

A routine retropleural drain placed near the anastomosis may not be necessary in all cases of EA and TEF. Good prognosis patients (Waterston class A and B) who undergo an uncomplicated extrapleural repair without undue tension do not appear to benefit from having a chest drain in place, and there is potential for complications. In complicated cases, however, retropleural drainage remains a reasonable adjunct.[20]

CONCLUSION

We conclude that proper antenatal check ups will pick the problem early, avoid home deliveries and hence improve survival. It is the pulmonary status and septicaemia which had a significant impact on the survival. Moreover these babies cannot be managed in the general ward. They need ICU care especially those who require assisted ventilation. Availability of ICU is one of the main determinants of survival. The likely cause of high mortality rate in pre-operative patients is non-availability of NICU due to limited space.

REFERENCES

1. 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(1):267–72.
2. Tsai JY, Berkery L, Wesson DE, Redo SF, Spigland NA. Esophageal atresia and tracheoesophageal fistula: surgical experience over two decades. Ann Thorac Surg 1997;64(3):778–83.
3. Seguier-Lipszyc E, Bonnard A, Aizenfisz S, Enezian G, Maintenant J, Aigrain Y, de Lagausie P. The management of long gap esophageal atresia. J Pediatr Surg 2005;40(10):1542–6.
4. Al-Salem AH, Tayeb M, Khogair S, Roy A, Al-Jishi N, Alsenan K, Shaban H, Ahmad M. Esophageal atresia with or without tracheoesophageal fistula: success and failure in 94 cases. Ann Saudi Med 2006;26(2):116–9.
5. Seitz G, Warmann SW, Schaefer J, Poets CF, Fuchs J. Primary repair of esophageal atresia in extremely low birth weight infants: a single-center experience and review of the literature. Biol Neonate 2006;90(4):247–51.
6. Okada A, Usui N, Inoue M, Kawahara H, Kubota A, Imura K, Kamata S. Esophageal atresia in Osaka: a review of 39 years' experience. J Pediatr Surg 1997;32(11):1570–4.
7. Al-Malki TA, Ibrahim AH Esophageal atresia with tracheoesophageal fistula and early postoperative mortality. West Afr J Med 2005;24(4):311–5.
8. Konkin DE, O'hali WA, Webber EM, Blair GK. Outcomes in esophageal atresia and tracheoesophageal fistula. J Pediatr Surg 2003;38(12):1726–9.
9. Grignon A, Dubois J. Ultrasonography of twin pregnancies: J Radiol. 2002;83(12 Pt 2):1899–908.
10. Teich S, Barton DP, Ginn-Pease ME, King DR. Prognostic classification for esophageal atresia and tracheoesophageal

fistula: Waterston versus Montreal. J Pediatr Surg 1997;32(7):1075–9.
11. Lopez PJ, Keys C, Pierro A, Drake DP, Kiely EM, Curry JI, Spitz L. Oesophageal atresia: improved outcome in high-risk groups? J Pediatr Surg 2006;41(2):331–4.
12. Sugito K, Koshinaga T, Hoshino M, Inoue M, Goto H, Ikeda T, Hagiwara N. Study of 24 cases with congenital esophageal atresia: what are the risk factors? Pediatr Int 2006;48(6):616–21.
13. Khorshid EA, Dokhan AL, Turkistani AF, Shadi SM, Hassab MH. Five years experience in prenatal ultrasound diagnosis of esophageal atresia in Saudi Arabia. Ann Saudi Med 2003;23(3–4):132–4.
14. Yang CF, Soong WJ, Jeng MJ, Chen SJ, Lee YS, Tsao PC, Hwang B, Wei CF, Chin TW, Liu C. Esophageal atresia with tracheoesophageal fistula: ten years of experience in an institute. J Chin Med Assoc 2006;69(7):317–21.
15. Uchida K, Inoue M, Otake K, Okita Y, Morimoto Y, Araki T, Miki C, Kusunoki M. Efficacy of postoperative elective ventilatory support for leakage protection in primary anastomosis of congenital esophageal atresia. Pediatr Surg Int 2006;22(6):496–9.
16. Diaz LK, Akpek EA, Dinavahi R, Andropoulos DB. Tracheoesophageal fistula and associated congenital heart disease: implications for anesthetic management and survival. Paediatr Anaesth 2005;15(10):862–9.
17. Snajdauf J, Kalousova J, Styblova J, Fryc R, Pycha K, Petru O, Pachmannova D, Rygl M, Tomasek L. [Results of treatment of esophageal atresia] Cas Lek Cesk 2004;143(9):614–7.
18. Pueyo Gil C, Elias Pollina J, Gonzalez Martinez-Pardo N, Pison Chacon J, Romeo Ulecia M, Esteban Ibarz JA. [Prognosis assessment of esophageal atresia: our experience of 29 years] Cir Pediatr 2001;14(4):145–51.
19. Driver CP, Shankar KR, Jones MO, Lamont GA, Turnock RR, Lloyd DA, Losty PD. Phenotypic presentation and outcome of esophageal atresia in the era of the Spitz classification. J Pediatr Surg 2001;36(9):1419–21.
20. Kay S, Shaw K. Revisiting the role of routine retropleural drainage after repair of esophageal atresia with distal tracheoesophageal fistula. J Pediatr Surg 1999;34(7):1082–5.

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