

ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA

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Thomas Gibson is credited with the first description of esophageal atresia/tracheoesophageal fistula (EA/TEF) in 1697.¹ It took more than two hundred years for the first two patients to survive a multiple-staged procedure by Ladd² and Leven.³ In 1943, Haight and Towsley⁴ reported the first survivor following a primary anastomosis. After this there was an improvement in the outcome of patients with EA/TEF, but the survival rate continued to be poor. This was attributed to several factors, including delay in diagnosis, prematurity, preoperative morbidity and associated congenital anomalies. Recently, significant advances have been made in the management of neonates with EA/TEF.^{6,7} This has resulted in a progressive decrease in mortality as a result of early diagnosis and improved neonatal intensive care and neonatal anesthesia, to such an extent that the validity of Waterston's 1962 classification⁸ has been questioned, and at present in most developed countries the presence of associated major congenital anomalies determines survival.⁹ This, however, is not the case in developing countries, where many other factors continue to contribute to the persisting high mortality.^{10,11} This report documents our experience with a series of patients with EA/TEF, and highlights the management of the condition, complications encountered and the survival factors pertinent to each case.

Materials and Methods

During the period between 1989 to 1995, a total of 23 cases of EA and/or TEF were admitted to Qatif Central Hospital in Saudi Arabia. The records of all these patients were reviewed. The data abstracted included age, sex, clinical condition on admission, gestational age, birthweight, mode and place of delivery, method of diagnosis and transfer, the presence of associated anomalies, preoperative management, operative details, postoperative care and outcome. The infants were classi-

fied according to Waterston's classification. All x-ray and laboratory data were also reviewed.

Results

A total of 23 cases (16 males and 7 females) of EA and/or TEF were admitted to our hospital. The distribution of esophageal anomalies seen is shown in Table 1. None of our patients had isolated TEF. The mean birthweight was 2.3 kg (range 1.3-3.08 kg), and 14 out of 23 (60.9%) had a birthweight of less than 2.5 kg. The majority of our patients were referrals from nearby hospitals. One of the patients was a twin. Twelve infants (52%) had pneumonitis, with the right upper lobe being the most common site. They were treated with antibiotics for two to three days—longer if necessary—prior to surgery, and were intubated and ventilated if necessary.

The diagnosis of EA was suspected because of excessive mucous secretions. In 10 of our patients, choking on first feeding was the single most important symptom for suspecting EA. The diagnosis was confirmed by resistance to the passage of a stiff radio-opaque nasogastric tube and a babygram with the nasogastric tube in place. Two patients had had additional contrast esophagogram done before referral.

Associated congenital anomalies were detected in 11 of the patients (47.8%) (Table 2). The distribution of patients according to Waterston's classification is as shown in Table 3.

Only six of our patients were operated on within 24 hours of admission. The rest were operated on as late as six days after admission. They were given intensive therapy, including the use of antibiotics, and were operated on when they were deemed fit to withstand anesthesia. All of our patients were operated on via an extrapleural approach. Postoperatively, the patients were fed intravenously until extubated, and then via the nasogastric tube until the anastomosis was checked radiologically by a barium swallow on the 8th-10th postoperative day. If found intact, the retropleural drain was removed and gradual oral intake was started.

Fifteen of the 16 patients with EA and distal TEF had primary repair performed. The other patient had a long gap

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between the two ends, and had ligation of the fistula and gastrotomy. When there was no significant growth of the two esophageal ends after four months of waiting, gastric pull-up replacement was done. This patient died two months after the operation because of septicemia. One patient with EA, distal TEF and associated anorectal agenesis, sacral agenesis, patent ductus arteriosus and multiple rib anomalies, had primary esophageal anastomosis on the first day and a temporary colostomy on the third day. This patient died after seven days because of septicemia and associated congenital anomalies. Two of the three patients with EA and both proximal and distal TEF had primary esophageal repair, but the third one had a long gap. This patient was treated by ligation of both fistulae, followed by gastrotomy and by delayed primary anastomosis at the age of four weeks. He developed severe esophageal stricture, which was not dilatable. This stricture was excised and esophageal anastomosis was done at the age of five months. One of the two patients with EA and proximal TEF had primary esophageal anastomosis, while the other one had a long gap between the two ends. This was treated by ligation of the fistula and gastrotomy, but there was no significant growth between the two esophageal ends, so esophagostomy was performed, followed by gastric replacement at the age of one year. This patient is now 3½ years old and doing well. One of the two cases of esophageal atresia without TEF was treated by gastrotomy and delayed esophageal anastomosis at the age of three months, while the other one had associated agenesis of the left diaphragm. He was treated by gastrotomy and gortex graft of the hemidiaphragm, but the patient died at the age of 31 days because of severe pulmonary hypoplasia.

All the patients were ventilated postoperatively, except one who was extubated accidentally immediately after surgery. This patient developed minor leak clinically on the third postoperative day and was treated conservatively. Barium swallow on the 10th postoperative day revealed a distally placed associated congenital esophageal stenosis which we think contributed to the leak. There was no evidence of gastroesophageal reflux. The patient was treated with dilatation and is now 2½ years old and doing well. The average length of intubation for those with primary anastomosis was 4.8 days (range 2-12 days).

On follow-up ranging from nine months to six years (mean 3.6 years), 5 of our 23 patients developed complications. Three developed minor leaks (13%) which were treated conservatively with no sequelae. Two patients had a long gap EA and TEF, which was repaired under tension. Both of them developed a very tight stricture (8.7%), which was not dilatable, and so they were treated with excision of the stricture and esophageal anastomosis. Although incompletely assessed, gastroesophageal reflux was seen in three patients (13%).

TABLE 1. *Distribution of esophageal anomalies.*

Type of anomaly	Number
Esophageal atresia with distal TEF	16
Esophageal atresia with both proximal and distal TEF	3
Esophageal atresia with proximal TEF	2
Esophageal atresia without proximal TEF	2
Total	23

TEF=tracheoesophageal fistula.

TABLE 2. *Associated anomalies.*

Case no.	Associated anomalies
1	Cleft palate, right-sided aorta
2	Hydronephrosis due to PUJ obstruction, right-sided aorta
3	High imperforate anus, sacral agenesis, PDA, multiple rib anomalies
4	VSD, hydrocephalus
5	Congenital esophageal stenosis
6	Vertebral anomalies
7	Down syndrome, VSD
8	Right-sided aorta
9	Edward's syndrome, PDA, ASD, cleft palate
10	Agenesis left hemidiaphragm
11	Right-sided aorta

VSD=ventricular septal defect; ASD=atrial septal defect; PUJ=pelvi-ureteric junction; PDA=patent ductus arteriosus.

Two of these were treated conservatively and settled down, while the third is awaiting fundoplication. None of our patients developed recurrent TEF.

Four of our patients (17.4%) died at the ages of 1 week, 55 days, 31 days and 6 months, respectively, giving an overall survival rate of 82.6%. In two of the cases, sepsis was the main cause of death, while in the other two, severe associated anomalies were the main cause of death. All those who died were in Group C, according to Waterston's classification (Table 3).

Discussion

As a result of recent advances in the management of neonates with EA/TEF, including early diagnosis and improvement in neonatal intensive care, there has been a dramatic fall in morbidity and mortality in developed countries.^{5,6,12} Neonates with EA are now diagnosed within a few hours of birth and transferred expeditiously to pediatric surgical centers. Delays in diagnosis in developing countries, however, are reflected in the high

TABLE 3. *Classification of patients according to risk groups (Waterston's classification).*

Group	Number	%	Survival	%
A	4	17.4	4	100
B	11	47.8	11	100
C	8	34.8	4	50

incidence of aspiration pneumonitis. In our series, 52% of the patients had pneumonitis on admission. Many physicians are unaware that excessive mucous secretion or salivation is an early sign of EA, which can lead to delay in diagnosis. The fact that in 43% of our patients the diagnosis was made when the baby choked on feeding appears to confirm this impression. The outcome in the condition of these patients can be optimized by diligent measures directed at early diagnosis. A high degree of clinical suspicion among pediatricians and nursing staff should lead to confirmation of diagnosis by the inability to pass a size 6 or 8 nasogastric tube into the stomach. Contrast esophagogram yields no further information, but only delays the transfer of these neonates and exposes them to the risks of hypothermia and aspiration pneumonia.

Our present series provides a unique variation from others, in that the proportion of infants with the most common anomaly (i.e., EA with distal TEF) make up 69.6% of the total number. This is different from other series, where it accounts for over 85% of the different types of anomalies.^{12,13} This apparent discrepancy in our study is explained by the presence of three infants with EA and both distal and proximal fistula. The combination of EA with proximal fistula is infrequent, occurring in only 0-3% of patients, while EA with both proximal and distal fistula is seen in only 0.25-7.7% of patients.^{13,14} In our series we saw three patients with EA and both proximal and distal TEF (13%) and two patients with EA and proximal TEF (8.7%). One explanation for this variation is the small number of patients in our series. The presence of proximal fistula increases the incidence of aspiration pneumonia and thereby adds to morbidity. We found that mobilization of the upper pouch to the thoracic inlet is of great importance, as this enables us to gain length to attain a tension-free anastomosis. We also found this maneuver helpful in defining associated pouch fistula which can be missed if not properly looked for. This maneuver eliminated the need for circular myotomy in our patients. We also think this makes the need for preliminary endoscopy or preoperative contrast study unnecessary for diagnosis of an associated upper pouch fistula.

In our series, among infants with EA and TEF (21 infants), 18 were taken through a primary anastomosis. Two of these infants had long gaps between the two esophageal ends and anastomosis was made under tension. Both of them strictured, and secondary operations became necessary. In the remaining three patients, it was decided at

primary assessment to delay and stage surgery. Two of them required gastric replacement, while the third had a successful delayed primary esophageal anastomosis. There are several methods of esophageal replacement, and having a stomach for the anastomosis is as effective as any other method.¹⁵⁻¹⁸ In the two cases of esophageal atresia without fistula, one was successfully treated by delayed esophageal anastomosis, while the second one died as a result of associated agenesis of hemidiaphragm and severe pulmonary hypoplasia. Delayed primary esophageal anastomosis is feasible in some of those with long gap between the two esophageal ends, although a high percentage of them develop esophageal stricture.^{19,20} Like others, we do not routinely perform gastrostomy in our patients,¹² which has contributed to a low incidence of gastroesophageal reflux. We routinely pass a nasogastric tube before the anastomosis is completed and use this for feeding prior to oral feeding. Four of our patients (17.4%) had a right-sided aortic arch, although in general about 5% of infants with EA have a right-sided aortic arch.^{21,22} Although a left thoracotomy approach is advocated in all cases in which a right aortic arch is detected preoperatively, we found it possible to perform the anastomosis from the right side.

In a large series of 148 patients with EA/TEF, the survival rate for group A was 100%, group B 86%, and group C 73%.¹² The complication rates were anastomotic leak in 21% of the cases, anastomotic stricture in 18% and recurrent TEF in 12%. Our complication rates were anastomotic leak in 13%, and anastomotic stricture in 8.7%. None of our patients developed recurrent TEF or tracheomalacia. The low incidence of anastomotic complications in our series is attributable to several factors. These include the use of one-layer monofilament absorbable sutures for anastomosis, and electively ventilating our patients. Silk was shown to be associated with a high incidence of anastomotic complications. We found that elective mechanical ventilation is beneficial in reducing tension on the anastomosis in the early stage, by eliminating swallowing and maintaining the neck in a flexed position.²⁴

Our overall survival rate of 82.6% is by any standard quite acceptable and comparable to that of developed countries.^{12,13,25} Among those who died, two in the group C category had associated life-threatening anomalies and could not be saved in any case. One of them had an associated agenesis of the left hemidiaphragm and in our experience, this anomaly is associated with a severe degree of pulmonary hypoplasia. The other two died of severe sepsis, and arguably could have been saved. However, these two belonged to Waterston's Group C category.

References

1. Gibson T. The anatomy of human bodies epitomized. 6th edition. London: Awnsham and Churchill, 1697.
2. Ladd WE. The surgical treatment of esophageal atresia and tracheoesophageal fistula. *N Engl J Med* 1941;230:625-37.
3. Leven NL. Congenital atresia of the esophagus with tracheoesophageal fistula: report of successful extrapleural ligation of fistulous communication and cervical esophagostomy. *J Thorac Surg* 1941;10:648-57
4. Haight C, Towsleg HA. Congenital atresia of the esophagus with tracheoesophageal fistula: extra-pleural ligation of fistula and end to end anastomosis of esophageal segment. *Surg Gynecol Obstet* 1943;76:672-5.
5. Holder TM, Cloud DT, Lewis JE Jr, et al. Esophageal atresia and tracheoesophageal fistula: a survey of its members by the surgical section of the American Academy of Pediatrics. *Pediatrics* 1964;34:542-9.
6. O'Neil JA Jr, Holcomb GW Jr, Neblett WW III. Recent experience with esophageal atresia. *Ann Surg* 1982;195:739-45.
7. Randolph JG, Newman KD, Anderson KD. Current results in repair of esophageal atresia with tracheoesophageal fistula using physiologic status as a guide to therapy. *Ann Surg* 1989;209:526-31.
8. Waterston DJ, Carter REB, Aberdeen E. Esophageal atresia: tracheoesophageal fistula: a study of survival in 218 infants. *Lancet* 1962;1:819-22.
9. Spitz L, Kiely EM, Morecroft JA, Drake DP. Esophageal atresia: at risk groups in the 1990's. *J Pediatr Surg* 1994;29:723-5.
10. Agarwal, Bhatnagar V, Bajpai M, Gupta DK, Mitra DK. Factors contributing to poor results of esophageal atresia in developing countries. *Pediatr Surg Int* 1989;4:76-9.
11. Debo Adeyemi S. Management of Nigerian neonates with high risk esophageal atresia: early versus delayed repair. *Pediatr Surg Int* 1989;4:76-9
12. Spitz L, Kiely E, Brereton RJ. Esophageal atresia: five years experience with 148 cases. *J Pediatr Surg* 1987;22:103-8.
13. Goh DW, Brereton RJ. Success and failure with neonatal tracheoesophageal anomalies. *Br J Surg* 1991;78:834-7.
14. Van der Zee DC, Van der Staak FHJ, Severinjen R SVM, Festen C. Proximal fistula in esophageal atresia: pitfall in a routine procedure. *Pediatr Surg Int* 1988;23-6.
15. Spitz L. Gastric transposition via the mediastinal route for infants with long gap esophageal atresia. *J Pediatr Surg* 1984;19:149-54.
16. Freeman NV, Cass DT. Colon interposition: a modification of Waterston's technique using the normal esophageal route. *J Pediatr Surg* 1982;17:17-21.
17. Foker JE, Ring WS, Vorco RL. Technique of jejunal interposition for esophageal replacement. *J Thorac Cardiovasc Surg* 1982;83:928-33.
18. Cohen D. Esophageal reconstruction using a gastric tube: a preliminary report. *Austr Pediatr J* 1970;6:22-4.
19. Myers NA, Beasley SW, Auldish AW, Kent M, Wright V, Chetcuti P. Esophageal atresia without fistula, anastomosis or replacement. *Pediatr Surg Int* 1987;2:216-22.
20. Puri P, Ninan GK, Blake NS, et al. Delayed primary anastomosis for esophageal atresia: 18 months to 11 years follow-up. *J Pediatr Surg* 1992;27:1127-30.
21. Harrison MR, Hanson BA, Mahour GH, Takahasi M. The significance of right aortic arch in repair of esophageal atresia and tracheoesophageal fistula. *J Pediatr Surg* 1977;12,:861-9.
22. German JC, Mahour GH, Woolley MM. Esophageal atresia and associated anomalies. *J Pediatr Surg* 1976;11:299-306.
23. Rivosecchi M, Bogolan P, Matarazzo E. Esophageal atresia: critical review of 10 years experience. *Pediatr Surg Int* 1989;4:95-100.
24. Mackinlay GA, Burtles R. Esophageal atresia: paralysis and ventilation in management of the wide gap. *Pediatr Surg Int* 1987;2:10-2.
25. Beasley SW, Myers NA. Trends in mortality in esophageal atresia. *Pediatr Surg Int* 1992;7:86-9.