

Esophageal atresia with or without tracheoesophageal fistula: success and failure in 94 cases

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BACKGROUND: The management of newborns with esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) has evolved considerably over the years. Currently an overall survival of 85% to 90% has been reported from developed countries. In developing countries, several factors contribute to higher mortality rates. We describe our experience with 94 consecutive cases of EA with or without TEF.

PATIENTS AND METHODS: We retrospectively studied 94 patients with EA with or without TEF treated at our hospital over a period of 15 years. Medical records were reviewed for age at diagnosis, sex, birth weight, associated anomalies, aspiration pneumonia, method of diagnosis, treatment, postoperative complications and outcome.

RESULTS: Ninety-four newborns (55 males and 39 females) with EA/TEF were treated at our hospital. Their mean birth weight was 2.2 kg (700 g to 3800 g). Age at diagnosis ranged from birth to 7 days. At the time of admission 37 (39.4%) had aspiration pneumonia. Associated anomalies were seen in 46 (49%) patients. Thirteen patients had major associated anomalies that contributed to mortality. Postoperative complications were similar to those from developed countries but overall operative mortality (30.8%) was high.

CONCLUSIONS: The overall mortality was high but excluding major congenital malformations, sepsis was the most frequent cause of death. Factors contributing to mortality included prematurity, delay in diagnosis with an increased incidence of aspiration pneumonia and a shortage of qualified nurses. To improve overall outcome, factors contributing to sepsis should be evaluated and efforts should be made to overcome them.

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Accepted for publication
March 2006

Ann Saudi Med 2006;26(2):116-119

Thomas Gibson first described esophageal atresia (EA) with tracheoesophageal fistula (TEF) in 1697.¹ The first primary repair of this anomaly was not done, however, until 244 years later by Cameron Height in 1941.² Since then, the management of EA with or without TEF has evolved considerably over the years. Improvements in management are attributed to several factors, including early diagnosis, improved neonatal intensive care, neonatal anaesthesia and surgical techniques. Currently, an overall survival rate of 85% to 90% and a survival rate of over 95% in those without major anomalies has been reported from developed countries.^{3,4,5} Improved survival may not be the case in developing countries where several factors continue to contribute to lower survival rates, including a delay in diagnosis, low birth weight and prematurity, increased frequency of aspiration pneumonia, associated major congenital anomalies and increased perioperative morbidity.^{6,7} This study is an analysis of our experience with 94 new-

borns with EA with or without TEF with emphasis on factors pertinent to the final outcome.

Patients and Methods

Over a period of 15 years (1989 to 2004), 94 patients with EA with or without TEF were treated at our hospital. Their medical records were retrospectively reviewed for age at diagnosis, sex, birth weight, gestation, associated anomalies, method of diagnosis, associated aspiration pneumonia, type of anomaly, and morbidity and mortality. The outcome was classified according to the Spitz and Waterston classification.^{8,9}

Results

Ninety-four newborns with EA with or without TEF were treated at our hospital. There were 55 males and 39 females. Their mean birth weight was 2.2 kg (700 g to 3800 g). Twelve (12.8%) had a birth weight less than 1500 g. Their gestational age ranged from 23 weeks to 41 weeks and in 36 (38.3%) the gestational age was less than 35 weeks. In 30 (32%), there was a positive history of polyhydramnios. Four of our patients were one of a twin. Their age at diagnosis ranged from birth to 7 days and a contrast study to confirm the diagnosis was done in 44% of patients prior to referral to our hospital. At the time of admission to our hospital, 37 (39.4%) had aspiration pneumonia. Associated anomalies were seen in 46 (49%) (Table 1). Among these, congenital heart disease was the commonest, seen in 42 (44.7%), including a right-sided aorta in 9 (9.6%). Other associated anomalies included vertebral anomalies in 9 (9.6%), chromosomal abnormalities in 9 (9.6%), including trisomy 18 (Edward’s syndrome) in 6 and Down’s syndrome in 2, VACTERL association in 2, agenesis of the left hemidiaphragm in 2, intestinal atresia in 2, left pelviureteric junction obstruction in 1, associated distal congenital esophageal stenosis in 1 and anorectal agenesis in 6 (6.4%).

The different types of EA/TEF seen in our patients are shown in Table 2. EA with distal TEF was the commonest, seen in 79 (84%) patients, followed by pure EA in 8 (8.5%) patients. EA with both proximal and distal fistula was seen in 3 (3.2%), EA with proximal fistula in 2 (2.1%) and H-fistula in 2 (2.1%). The patient’s classification according to Spitz and Waterston is shown in Table 3.

Twenty-four (25.5%) of our patients had a long gap, which is arbitrarily defined as anastomosis under severe tension or a gap greater than the height of three vertebrae between the two esophageal ends. Eight newborns had pure EA, 15 had EA with distal

Table 1. Associated congenital malformations in 94 newborns with esophageal atresia with or without tracheoesophageal fistula.

Associated anomaly	Number of patients	%
Congenital heart disease	42	44.7
Right-sided aorta	9	9.6
Vertebral anomalies	9	9.6
Chromosomal abnormalities	9	9.6
Edward’s syndrome (trisomy 18)	6	6.4
Down’s syndrome (trisomy 21)	3	3.2
VACTERL association	2	2.1
Agenesis of left hemidiaphragm	2	2.1
Intestinal atresia	2	2.1
Left pelviureteric junction obstruction	1	1.1
Congenital esophageal stenosis	1	1.1
Anorectal agenesis	6	6.4
Others	39	41.4

Table 2. Classification of different types of esophageal atresia.

Type of esophageal atresia	Number of patients	%
Esophageal atresia with distal tracheoesophageal fistula	79	84.0
Pure esophageal atresia	8	8.5
Esophageal atresia with both proximal and distal fistula	3	3.2
Esophageal atresia with proximal fistula	2	2.1
H-type tracheoesophageal fistula	2	2.1

TEF and 1 had EA with proximal TEF. The 8 patients with long gap EA were managed as follows: 2 had gastrostomy only and died, 2 had gastrostomy and delayed primary anastomosis, 3 had gastrostomy and esophagostomy and died and 1 had a gastrostomy and gastric transposition. The 15 patients with EA and distal TEF and a long gap were managed as follows: 6 had primary repair under severe tension, 2 had delayed primary repair, 1 had gastrostomy and delayed primary repair, 3 had fistula ligation plus gastrostomy and esophagostomy and 2 subsequently had colonic transposition, 2 had fistula ligation only and died, and 1 had fistula ligation plus gastrostomy and subsequently had gastric transposition. One patient with EA and proximal TEF had a long gap. He underwent gastrostomy plus esophagostomy and subsequently gastric transposition. Of 94 patients, 70 had

Table 3. Distribution and mortality according to Spitz and Waterston classifications

Waterston classification		
Group	Number of patients	Mortality (%)
A	21	0
B	40	20
C	33	63.6
Spitz classification		
Group	Number of patients	Mortality (%)
I	65	18.5
II	20	50
III	9	77.8

primary repair and 11 had delayed primary repair or primary repair under tension. Twelve of the 70 (17%) had a leak, 9 (12.9%) developed esophageal stricture, 9 (12.9%) had gastroesophageal reflux and 1 (1.4%) developed a recurrent fistula (Table 4). Twenty-nine patients died for an overall mortality of 30.8%. Six patients had Edward’s syndrome, 2 had associated agenesis of the left hemidiaphragm and pulmonary hypoplasia, 3 had associated severe congenital heart disease, 1 had associated severe hyaline membrane disease, and 1 had associated epidermolysis bullosa. If these causes are excluded, mortality was 17.2%. The mortality according to Spitz and Waterston classification is shown in Table 3 and the different causes of mortality are shown in Table 5.

Discussion

Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) is one of the commonest gastrointestinal malformations, second only to anorectal malformations.¹⁰ The exact incidence of EA with or without TEF is not known, but an incidence of 1 in 3000 to 1 in 4500 live births have been reported worldwide.^{11,12} In the kingdom of Saudi Arabia, the incidence of EA with or without TEF is not known, but an incidence of 0.3 per 1000 live births has been reported from the Asir region in the southwest of Saudi Arabia.¹⁰ In our hospital, it was difficult to estimate the incidence as a large number of our patients were referrals from other hospitals in the region.

Associated anomalies are frequent with EA, occurring in more than 50% of patients, and among these, congenital heart disease is the commonest.³ This was the case in our series as 49% of our patients

had associated congenital anomalies and 44.7% had congenital heart disease. Of interest was the finding of right-sided aorta in 9 (9.6%) of our patients. In 4, this was diagnosed preoperatively, while in the other 5, it was detected intraoperatively. In general about 5% of infants with EA have a right-sided aortic arch and if this is detected preoperatively, a left thoracotomy is advocated to achieve repair.^{13,14} A preoperative echocardiogram is valuable in this regard. The finding of a right-sided aortic arch did not however alter our surgical approach, and we found it possible to do the anastomosis from the right side.

The incidence of twinning, monozygotic or dizygotic, appears to be higher in families with EA.¹⁵ Four of our patients were one of monozygotic twins.

Several chromosomal abnormalities are associated with EA/TEF. Among these, trisomy 18 and 21 are the commonest.¹⁶ Nine (9.6%) of our patients had chromosomal abnormalities. The association of EA/TEF with trisomy 18 is of practical importance as in many centres these patients, because of the known outcome, will not be operated on and are left to die. However, this was not the case in our setting, where religious and social factors have to be considered. Add to this the fact that surgical repair makes it easier for the parents to care for them more comfortably until they die. Some of our patients had severe associated anomalies such as severe congenital heart disease, congenital diaphragmatic hernia with pulmonary hypoplasia, trisomy 18 and epidermolysis bullosa, which contributed to our overall high mortality. The association of EA/TEF with congenital diaphragmatic hernia is very rare and the majority died because of associated severe pulmonary hypoplasia.¹⁷

In 1962, Waterston proposed the risk classification of patients with EA.⁹ In recent years, however, and as a result of advances in neonatal intensive care, neonatal anesthesia, refinement in surgical techniques, and pre- and postoperative care, the outcome of these patients has improved markedly. In a large series of 148 patients with EA/TEF, the complication rates were anastomotic leak in 21%, anastomotic stricture in 18% and recurrent TEF in 12%.¹⁸ Our complication rates were anastomotic leak in 17%, anastomotic stricture in 13% and only one (1.4%) of our patients developed a recurrent fistula. These results are comparable with those reported from Western centres, but our overall mortality of 30.8% was high when compared with the less than 10% mortality reported from developed countries.^{4,18,19} Several factors contributed to this high mortality. Associated anomalies, especially major congenital heart disease, have

become the most important predictor of survival in those with EA/TEF.⁸ This was the case in our series where 13 of our patients had associated anomalies that were severe enough to contribute to mortality.

Currently, the diagnosis of neonates with EA/TEF is made within the first few hours of life and arrangements are made to transfer the patient immediately to specialized centres where pediatric surgeons are available. A delay in diagnosis and transfer is reflected in the high incidence of aspiration pneumonia. In our series and since the majority of our patients were referrals from other hospitals, 39.4% had aspiration pneumonia on admission. Pediatricians and nursing staff caring for these patients should be aware of this, and a high degree of clinical suspicion should lead to early diagnosis. This is especially so in newborns with excessive salivation. It is also too late if the diagnosis is made or suspected when the baby chokes on feeds. The diagnosis can easily be confirmed by simply passing a nasogastric tube. The use of a contrast esophagogram to confirm the diagnosis should be abandoned as this will not only delay their transfer but also increases the risk of hypothermia and aspiration pneumonia.

Our overall mortality of 30.8% was high, but if major congenital malformations are excluded, sepsis was the most frequent cause of death. Several factors contribute to this, including prematurity, a delay in diagnosis with an increased incidence of aspiration pneumonia, and a shortage of qualified nurses to care for these patients. Prematurity and associated severe congenital anomalies are two factors that are beyond our control, but to improve

Table 4. Postoperative complications in 70 patients who underwent primary repair.

Type of complication	Number of patients	%
Esophageal leak	12	17
Esophageal stricture	9	12.9
Gastroesophageal reflux	9	12.9
Recurrent tracheoesophageal fistula	1	1.4

Table 5. Causes of mortality.

Cause of death	Number of patients	%
Sepsis	16	55.2
Severe congenital heart disease	3	10.3
Trisomy 18	6	20.7
Hyaline membrane disease	1	3.4
Epidermolysis bullosa	1	3.4
Agenesis of left hemidiaphragm with pulmonary hypoplasia	2	6.9

overall outcome, all other factors contributing to sepsis should be evaluated and every effort should be made to overcome them. These include early diagnosis and early referral of these patients to specialized centres, continuous suction of saliva and the upper esophageal pouch while in hospital and during their transfer, avoidance of contrast media to confirm the diagnosis, and provision of adequate and well-trained nurses to take care of these patients.

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