Esophageal Atresia with Tracheoesophageal Fistula: Ten Years of Experience in an Institute

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Background: Esophageal atresia (EA), tracheoesophageal fistula (TEF), or both is a complicated problem. The purpose of this study was to evaluate the outcomes and postoperative complications in patients with EA/TEF who were admitted to our hospital.

Methods: In total, 15 patients were enrolled from 1994 to 2003, including 8 males and 7 females. Patient demographics, associated anomalies, and outcomes were analyzed.

Results: The most common variant was EA with a distal TEF (type C), which occurred in 12 patients (80%). The latter had associated congenital anomalies, and cardiac anomalies were the most frequent, occurring in 8 patients (53.3%). Of the 6 cases who had life-threatening anomalies, 4 (66.7%) died, and of the 9 cases who had no life-threatening anomalies, 2 (22.2%) died. Tracheomalacia and/or stenosis were diagnosed in 8 patients (66.7%) postoperatively. Though 3 of the 4 cases who suffered from dying spell received intratracheal stent implantation, 2 cases still died.

Conclusion: The survival rate of the patients with EA/TEF is influenced mainly by associated life-threatening anomalies. TMS combined with a history of dying spell may be the major fatal complication. [*J Chin Med Assoc* 2006;69(7):317–321]

Key Words: congenital anomalies, esophageal atresia, intratracheal stent, tracheoesophageal fistula

Introduction

Congenital esophageal atresia (EA) and tracheoesophageal fistula (TEF) are well-known congenital anomalies which affect 1 in 2,400–4,500 live births.[1] The survival of infants born with both EA and TEF (EA/TEF) has dramatically improved since Cameron Haight's first report of successful surgical correction in 1941.[2] Improvement of survival is multifactorial and largely attributable to the advances in neonatal intensive care, anesthetic management, ventilatory support, and surgical techniques over the past decades. Survival can even be achieved in low birth weight infants,[3] and mortality is currently limited to those with coexistent severe life-threatening anomalies.

Despite the improvement in survival, the morbidity associated with surgical repair remains high.[4] In this study, we review our previous 10-year experience of patients with EA/TEF. The results may be beneficial for further clinical management of these patients in future.

Methods

Patients who had been admitted to Taipei Veterans General Hospital with a diagnosis of EA/TEF between 1994 and 2003 were enrolled. We retrospectively analyzed their medical record charts. Their anatomical classification was based on the Gross classification (type A–D, plus H-type TEF).[5]

These infants were also classified into the following risk groups with Waterston and associates' method: group A, birth weight >2,500 g and well; group B, birth weight 1,800–2,500 g and well or birth weight >2,500 g but with pneumonia and other congenital

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anomalies; and group C, birth weight <1,800 g or birth weight >1,800 g with severe anomalies or pneumonia.

Patient demographics, associated anomalies, postoperative complications, and outcomes were studied. The follow-up period ranged from 14 months to 10 years.

Results

In total, 15 patients, 8 males (53.3%) and 7 females (46.7%), were enrolled. The mean gestational age was 37 ± 3.2 weeks (range, 27–41 weeks) and the mean birth weight was 2,491 ± 708 g (range, 770–3,825 g). There were 3 preterm infants (gestational age <37 weeks, 20%) and 5 babies were small for gestational age (2 standard deviations below the mean weight for gestational age, 33.3%).

Patient characteristics and associated anomalies are shown in Table 1. By the Gross classification, the most common type was EA with a distal TEF (type C), which occurred in 12 of the 15 cases (80%). Among the 3 remaining patients, the first was pure EA (type A), the other was H-type TEF without EA, and the third was tracheal atresia with distal fistula (non-type). By risk groups with Waterston and associates' method: group A,

Table 1. Patients demographics and associated anomalies

Case	Sex	GA (wk)	BBW (g)	Type*	Group†	Operation	Anomalies	Survival
1	F	36	1,910	C	B	Yes		Yes
2	M	38	2,430	C	B	Yes		Yes
3	M	40	3,030	C	A	Yes	Microtia	Yes
4	F	35	2,500	A	B	Yes	Dextrocardia Right lung hypoplasia	Yes
5	F	37	2,100	C	C	Yes	TOF, MAPCA Bronchial stenosis	Yes
6	F	39	3,080	C	B	Yes	Type II VSD Conductive hearing loss	Yes
7	F	39	2,730	C	B	Yes	Hypothyroidism	Yes
8	M	39	3,825	C	A	Yes		Yes
9	M	39	2,250	C	C	No	VACTERL syndrome: TOF Imperforate anus Scoliosis Polydactyl	No
10	M	39	2,700	Tracheal atresia/distal TEF	C	Yes	VACTERL syndrome: DORV Type II VSD, PS, PDA Subglottic stenosis Rectourethral fistula Imperforate anus	No
11	F	41	2,680	C	B	Yes	Without left thumb, PDA	No
12	M	39	1,680	C	C	No	Trisomy 18	No
13	F	27	770	C	C	No	Cataract	No
14	M	40	2,680	C	C	Yes	Laryngeal cleft Bronchial stenosis Subglottic stenosis	No
15	M	37	3,000	H	C	Yes	Double aortic arch Type II VSD, ASD, PDA, bilateral PPS Tracheomalacia Bronchial stenosis Cleft palate	No

**Gross classification; †Waterston and associates' method. GA = gestational age; BBW = birth body weight; TOF = tetralogy of Fallot; MAPCA = major aortopulmonary collateral arteries; VSD = ventricular septal defects; VACTERL = vertebral anomalies, anal atresia, cardiac defect, tracheoesophageal fistula, renal abnormalities, and limb abnormalities; DORV = double outlet of right ventricle; PS = pulmonic stenosis; PDA = patent ductus arteriosus; ASD = atrial septal defects; PPS = peripheral pulmonic stenosis.*

2 patients (13.3%); group B, 6 patients (40%); and group C, 7 patients (46.7%). Most patients in our study were in higher risk groups, especially groups B and C.

As shown in Table 2, 12 patients (80%) had associated congenital anomalies. Cardiac anomalies were the most frequent, occurring in 8 patients (53.3%). Airway and lung anomalies occurred in 5 patients (33.3%), which included right lung hypoplasia, bronchial stenosis, laryngeal cleft, subglottic stenosis, and tracheomalacia/stenosis (TMS). Head and neck anomalies occurred in 3 patients (25.0%), which included microtia, cleft palate, and conductive hearing loss. Gastrointestinal anomalies occurred in 2 patients (13.3%) and both were imperforate anus. Skeletal deformities occurred in 2 patients (16.7%): 1 was polydactyly and scoliosis, and the other was absence of left thumb. Major genitourinary tract anomaly occurred in 1 patient (6.7%), diagnosed as rectourethral fistula. Metabolic disease occurred in 1 patient (6.7%) who had hypothyroidism. Ophthalmologic anomaly of cataract occurred in 1 patient (6.7%). Three cases were also diagnosed with chromosomal anomalies: 1 trisomy 18 and 2 VACTERL syndromes. Severe life-threatening anomalies, which mean fatal anomalies of vital organs, especially the cardiac system, were noted in 6 patients (40%: cases 4, 5, 9, 10, 12, 15).

Table 2. Associated anomalies in 15 EA/TEF patients

Anomaly	*n*
Cardiac (*n* = 8)	
PDA	3
VSD	3
TOF	2
PS/PPS	2
ASD	1
Dextrocardia	1
Double aortic arch	1
DORV	1
MAPCA	1
Airway and lung (*n* = 5)	
Bronchostenosis	3
Subglottic stenosis	2
Right lung hypoplasia	1
Laryngeal cleft	1
Tracheomalacia	1
Syndromes (*n* = 3)	
VACTERL	2
Trisomy 18	1
Head and neck (*n* = 3)	
Microtia	1
Cleft palate	1
Conductive hearing loss	1
Skeletal (*n* = 2)	
Polydactyly	1
Left thumb absence	1
Scoliosis	1
GI system (*n* = 2)	
Imperforate anus	2
GU system (*n* = 1)	
Rectourethral fistula	1
Metabolic disease (*n* = 1)	
Hypothyroidism	1
Ophthalmologic (*n* = 1)	
Cataract	1

PDA = patent ductus arteriosus; VSD = ventricular septal defects; TOF = tetralogy of Fallot; PS = pulmonic stenosis; PPS = peripheral pulmonic stenosis; ASD = atrial septal defects; DORV = double outlet of right ventricle; MAPCA = major aortopulmonary collateral arteries; VACTERL = vertebral anomalies, anal atresia, cardiac defect, tracheoesophageal fistula, renal abnormalities, and limb abnormalities; GI = gastrointestinal; GU = genitourinary.

Three patients died before definitive surgical repair operation. Two had chromosomal anomalies (cases 9, 12) and were given up by the family. The third patient died from complications of extreme prematurity (case 13). In total, 12 patients had received surgical correction in our hospital. Five of them (cases 1, 2, 4, 7, 14) were transferred from other hospitals. Of these 5 patients, 3 (cases 1, 2, 14) suffered anastomotic leaks with recurrent TEF after primary surgery and the repair procedures were done at our hospital. One patient (case 4, type A), referred after preliminary gastrostomy and esophagostomy, received reconstruction with right colon later in our hospital. One patient (case 7) got severe stricture at the anastomosis of esophagus after the first surgery and received repair procedure in our hospital. Of the remaining 7 patients, 6 underwent primary esophageal anastomoses within the first 48 hours after birth, and 1 underwent repair of H-type fistula 39 days after birth.

Complications after surgery are shown in Table 3. Postoperative TMS was found in 8 patients diagnosed

Table 3. Complications after surgery in 12 EA/TEF patients

Complications	*n* (%)
Tracheomalacia/stenosis	8 (66.7)
Recurrent pneumonia	6 (50.0)
Gastroesophageal reflux	6 (50.0)
Anastomotic leak	4 (33.3)
Dying spell	4 (33.3)
Anastomotic stricture	2 (16.7)
Recurrent TEF	0 (0.0)

by flexible airway endoscopy. Four patients with TMS manifested dying spell or feeding spell (cases 2, 5, 14, 15), and 1 of them also showed the coexistence of esophageal stricture (case 5). Tracheal stents were deployed in 3 dying spell patients, and 2 of them still suffered recurrent desaturation, respiratory distress and failure after the stenting, and finally died (cases 14, 15). One patient survived tracheal balloon dilatations done twice (case 5). Recurrent pneumonia (≥3 infections per year) occurred in 6 patients (50%) and 1 received lobectomy. Gastroesophageal reflux (GER) was found in 6 patients (50%), 2 of them received antireflux procedures. Anastomotic leakage occurred in 4 patients (33.3%). Anastomotic strictures, defined as an anastomotic narrowing that required dilatations or reoperation, occurred in 2 patients (16.7%) at esophagoesophageal (EE) and esophagocolonic (EC) anastomoses. Balloon dilatation was done in 1 patient and reoperation in the other.

During the follow-up period, 7 patients (46.7%) died. Four patients died after surgical repair, 2 of whom had experienced dying spell (cases 14, 15). Four of the 6 patients with life-threatening anomalies (66.7%) died. In 9 cases who had no life-threatening anomalies, only 2 (22.2%) died. The survival rate in each risk group of A, B and C were 100%, 83.3% and 14.3%, respectively.

Discussion

Several studies have reported the variety of postoperative complications and outcomes in EA/TEF patients.[4–8] Respiratory complications were associated with TMS (11–62%), dying spell (2.7%), and recurrent pneumonia (90%).[4,7,9] Gastrointestinal complications included GER (35–95%) and esophageal stenosis (17– 40%).[5,10] Others were anastomotic leakage (8.5–36%), recurrence of fistula (4–10%), and anastomotic stricture (8–37%).[5–8]

The association of TMS with EA/TEF has been well recognized. It is present in pathologic specimens in 75% of EA/TEF patients.[7,11] In our series, it was diagnosed in 8 patients (66.7%). The rate was higher than in the earlier studies,[4,7] but a bit higher than the recent study[10] because dynamic flexible airway endoscopy for the routine check of the whole airway in these patients was recently used in our hospital. Thus, we could detect more dynamic changes in TMS cases, which may otherwise be missed by computed tomography study. "Dying spell" means patients with TMS getting apnea and cyanosis, especially when they feed, cry, or cough, and can be the most serious problem associated with TMS. When TMS is severe enough to induce obvious clinical symptoms (dyspnea, cyanosis, or recurrent pneumonia), aggressive intervention should not be delayed.[12,13] Patients who have esophageal stenosis can cause more serious problem because the food bolus may stack at the proximal site of the stenosis. This bulging pouch may anteriorly compress the membranous portion of the trachea, resulting in tracheal lumen block, then inducing apnea and hypoxia. In our series, 4 cases (33.3%) were diagnosed as having dying spell. The incidence was higher compared with the other report[7] as we treated the transferred cases (cases 2, 14) with recurrent TEF and anastomotic strictures who had a higher possibility of getting dying spell after reoperation, and case 15, another dying spell case, noted to have TMS before surgery, had the condition worsen after the total correction. Three of the 8 TMS cases with dying spell had received tracheal stent placement, and 2 of them died. One survived and the stent was removed successfully 1 year later. Another TMS patient with dying spell, no stent deployed, received tracheal balloon dilatations twice and had normal respiratory status in the follow-up period.

Esophageal dysmotility and GER are common sequelae in patients who have undergone repair of EA/TEF. In our series, 6 cases (50%) were diagnosed with GER. The incidence was higher than some, but not all reports[4,7,10] because we had more complicated transferred cases (cases 2, 7, 14) who required reoperation that might have worsened the intrinsic motor function of the esophagus. Chittmittrapap et al[4] showed that GER could significantly increase the esophageal stricture rate. Two of the GER cases had strictures over the site of anastomosis. We advocate early detection and treatment of GER to reduce postoperative stricture rate.

The reported incidence of recurrent TEF ranges from 3 to 12%.[4–5] The pathogenesis of recurrent TEF can be an anastomotic suture leakage with further erosion through the previous repair site. All the 3 cases with recurrent TEF in our series were transferred from other hospitals after the primary surgery, and came for further surgical repair.

In 1962, Waterston et al[14,15] devised the first classification systems for EA/TEF. Risk factors included low birth weight, pneumonia, and associated anomalies. Our study discloses different survival rates among groups A, B and C patients (100%, 83.3% and 14.3%, respectively). We demonstrated that this classification may predict outcomes, but the number of our patients is not large enough to make a statistically significant differentiation.

The management of infants with isolated EA (type A) is often complex and presents a challenge for

pediatric surgeons. The operative morbidity associated with esophageal replacement procedure is high.[5,16,17] In addition to the anastomotic leakages, strictures, and graft ischemia, the other late complications include GER, recurrent aspiration, esophageal dysmotility, swallowing problems, ulceration, bleeding, respiratory tract infections, and failure to thrive.[5,17–19] One case of isolated EA in our series had received reconstruction with right colon, which was complicated with esophageal stricture, and who subsequently had balloon dilatations done 4 times, suture site leakage, recurrent pneumonia, and pneumonectomy. She is 11-years-old and has normal respiratory status without any other complications. The mortality rate in our series is higher than in other reports[7,8] because of the more complicated nature of the transferred cases.

We conclude that the survival rate of patients with EA/TEF was influenced mainly by associated life-threatening anomalies and postoperative TMS with dying spell. Therefore, we suggest that EA/TEF patients with clinical respiratory distress should be evaluated aggressively by flexible airway endoscopy that can be used not only for early diagnosis but also for intervention.

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