BAOJ Surgery

Lei Keng Sun, et al., BAOJ Surgery 2018, 4: 1
4: 032

Case Report

# A Retrospective Study of Esophageal Atresia and Tracheoesophageal Fistula in Centro Hospitalar Conde De São Januário, Macao, China During A Period Of 23 Years

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## Introduction

Esophageal atresia (EA) and tracheoesophageal fistula (TEF) is a congenital anomaly occurring in newborns, with the incidence of 1 in 3000-4500 births [1]. The incidence worldwide is reducing in trend for unknown reasons. The highest incidence of this disorder is reported in Finland (1/ 2500 births) [2].

EA is congenitally interrupted esophagus. TEF is a congenital or acquired communication between the trachea and esophagus.

The outcome depends on many factors, mainly the associated congenital anomalies with the TEF. Waterston and Spitz had suggested different classifications that determine the prognosis of the patient [2]. 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. Statistics regarding mortality in EA are constantly changing and improving.

Identifying the relevant risk factors in the local hospital will enable us to stratify the prognosis of the babies based on suitable prognostic criteria.

## Purpose

The objective of this review was verify 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, types of EA/TEF, types of surgery and complication.

## Materials and Methods

Retrospective record review was carried out among babies with EA/TEF in the Neonatal ICU(NICU), Department of Pediatric, CHCSJ from 1st January 1993 to 31st December 2016 (23 years). All babies admitted into NICU in CHCSJ who was diagnosed with EA/TEF throughout the review period were included. Babies whom were diagnosed with EA/TEF but non-operation and those with transferred to Hong Kong Hospital were included in this review. A prepared data collection sheet was used to collect the relevant information.

We inserted ICD-9 code 750.3 system to check the numbers of affected cases. The preoperative assessment of upper pouch was done with plain X-ray chest (posterior –anterior and lateral view) with 5Fr nasogastric tube. The diagnosis of associated congenital anomalies was performed on the basis of care full systemic examination, radiological and sonological investigations. Data collected included age at the time of admission, gestational age, birth weight, sex, which hospital delivery, associated congenital anomalies, respiratory status, presence of pneumonitis, type of anomaly, operative technique, complications and esophageal anastomotic leak (incidence, diagnosis and treatment). We defined survival as an infant who leaves hospital and who is able to effectively take the feeds. Surgery was performed with a retropleural approach with or without azygos ligation and chest tube drainage. In almost all the cases in our hospital records, end to end esophageal anastomosis was performed by monofilament absorbable suture (5-0 Polydioxanone) and changed braided absorbable suture (5-0 Polyglactin 910) recently 3 cases. All surgeries were performed using general anesthesia and patients were extubated

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**Sub Date:** February 9th 2018, **Acc Date:** February 20th 2018, **Pub Date**: February 21st 2018

**Citation**: Lei Keng Sun, Lui Kin Man, Jorge Sales Marques and Pang Heong Keong (2018) A Retrospective Study of Esophageal Atresia and Tracheoesophageal Fistula in Centro Hospitalar Conde De São Januário, Macao, China During A Period Of 23 Years. BAOJ Surgery 4: 032.

72 hours postoperatively unless they had respiratory distress, associated cardiac anomalies or marked tension at the anastomotic site. If the patient was on an endotracheal tube, he or she was shifted to neonatal intensive care unit for ventilatory support. A tube placed through the anastomosis into to the stomach allows decompression of the stomach and eventual enteral feeding; feeding was started after 24 h of surgery and gradually increased. If there was no evidence of any postoperative complications, contrast esophagram was done on the seventh day of surgery. In patients who had a chest drain, anastomotic leaks after the primary repair were detected by contrast study of the esophagus. An alternative method to confirm this was by giving oral methylene blue and then observing its appearance in the chest drain. Major leaks were clinically suspected by the contents draining with the accompanying deterioration in the general condition of the patient either due to mediastinitis or pneumonitis and septicemia. In patients who had no chest drain, leaks were clinically suspected by increased respiratory distress, fever and sepsis or plain X-ray chest showing pneumonitis and confirmed by the contrast study of esophagus. The findings suggestive of leak were extravasation of contrast from esophagus into mediastinum by esophagram. The finding of contrast in stomach without any clinical deterioration was considered normal. Esophageal stricture was defined as presence of symptoms (dysphagia and recurrent respiratory problems from aspiration or foreign body obstruction and narrowing noted on endoscopy or contrast esophagram.

## Results

### Demography

There were 16 patients with EA admitted into Neonatal ICU in CHCSJ from January 1993 to December 2016.

Out of the 16 patients, 9(56%) were males and 7 (44%) females.

The distribution of patients by race were 14(87.5%) Chinese, 2 (12.5%) Filipino. There were no significant racial predispositions of EA/TEF worldwide.

Maternal age ranges between 20 to 40 years old. The gestational issue included 1 with Beta-Thalassemia, 1 with IgA Nephropathy and preeclampsia, 1 with Myoma and Placenta previa, 1 with Gestational Diabetes mellitus and 1 with epilepsy history taking regular oral anti-epilepsy drug. In 8 cases showed history of polyhydramnios.

There were 9 (56%) premature babies out of the 16 babies diagnosed with EA. 3 of them (30%) were death which non-surgical intervention. All died because have increased rate of complications generally as compared to term babies.

## Types of EA/TEF

The types of EA/TEF were described by Gross and Boston in 1953 [3].

There were only 3 different types of EA/TEF seen in this review. Out of the 16 babies, 1(6.25%) was Type A, 11 (68.75%) were Type C (the commonest type) and 1 (6.25%) was Type D. Type B and E, EA/TEF were not found, 3 cases were unknown (Fig 1).

The case survival rates were 1 (100%), 9(82%), 1(100%) and 0% respectively (Tab 1).

Type A Esophageal atresia without a tracheoesophageal fistula. 1 of baby diagnosis was Type A EA/TEF, which upper pouch is high and the distance between 3 thoracic vertebraes confirmed by preoperative measurement of gap length by inserting a bougie into the upper pouch and placing contrast through a previously plasced gastrostomy tube. No more information record in document of above baby (which was referred to other center).

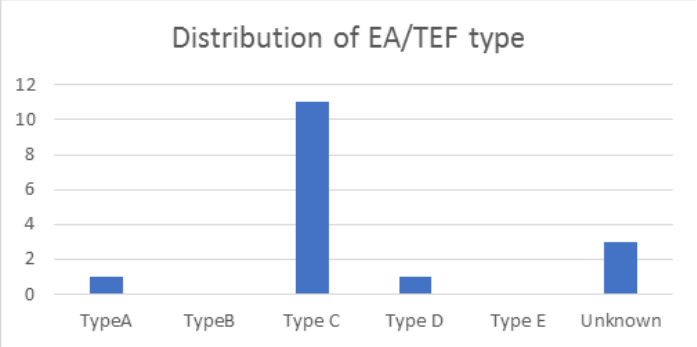


**Fig 1.** Distribution of types of Congenital Malformations in patients with EA/TEF in CHCSJ from 1993 to 2016.

**Table 1:** Survival related to type of TEF in our hospital. (n=16)

| | No. case | Case survived |
|---|---|---|
| Pure EA (Type A) | 1 (6.25%) | 1 (100%) |
| EA with distal TEF (Type C) | 11(68.75%) | 9(82%) |
| EA with proximal and distal TEF (Type D) | 1(6.25%) | 1(100%) |
| Unknown | 3(18.75%) | 0(0%) |

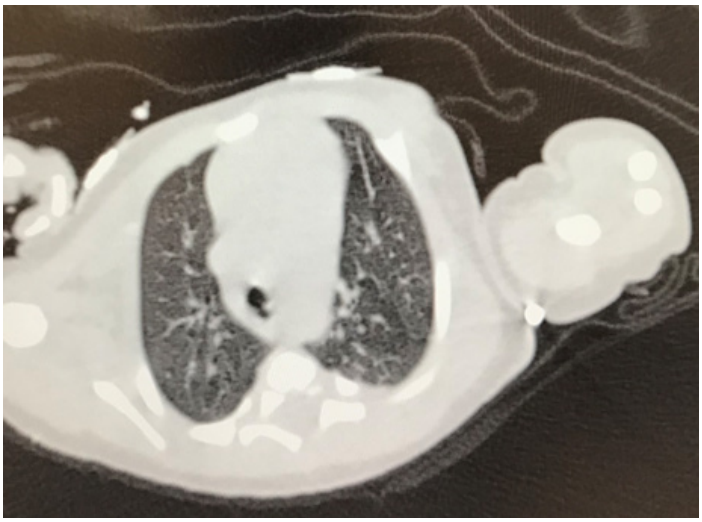

**Fig 2:** The tracheal esophagus fistula was noted before the tracheal carina.

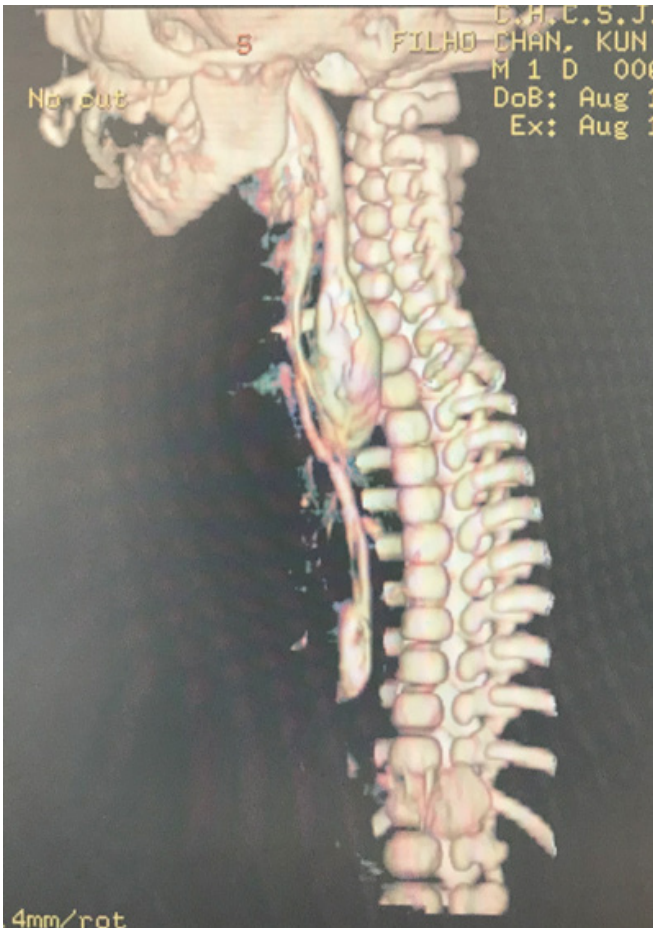


**Fig 3:** CT, administrating of dilated Iopamiol 20cc through gastric tube, showed EA and contrast medium in trachea and stomach after injecting at medium from the tube. A fistula is found from the carina to stomach.

## Birth Weight

The birth weight of the babies ranged from 1.3kg to 3.39kg with the smallest surviving baby was 1.92kg. Four babies were less than 2 kg, 5 babies were between 2.0-2.5kg, 5 babies were more than 2.5kg and 2 cases were unknown.

## Time Taken for Surgery

The time taken for surgical intervention for EA, is divided within 24 hours of birth, between 24 to 48 hours and more than 48 hours of birth. In 1(8%) was operated within 24 hours of birth, 7 (54%) were operated within 48 hours of birth. 5 (38%) of babies were operated in more than 48 hours after birth, 4 of them were referred to another center. Above 8 (89%) of babies were operated in primary surgery, 1 (11%) of them with preformed delayed surgery due to who had pre-operative pneumonia and needed ventilation. Antibiotic policy according the NICU protocol in our hospital.

In the past 23 years, a total 9 cases were responded by the same general surgeon and the neonatology team in our hospital, and antibiotic use was under our hospital NICU policy.

## Congenital Malformations

Out of 16 babies, 7 (44%) of them were born with associated congenital malformation. The types of congenital malformations were referred to the VACTERL associations. Vertebral anomalies were present in 2(12.5%) cases, anorectal malformation in other 2 (12.5%)es, cardiac anomalies in 8 (50%) babies, renal malformation in 4(25%)babies and limb dysplasia in 3 (18.7%). Out of 7 babies with congenital malformations, 3 of them were found to have VACTERL syndrome, which is 43% of the total number of babies with EA/FET. Two of babies were cleft palate. 2 of babies were clinical diagnosed Down Syndrome (1 was confirmed Robertsonian Translocation). 1 was Goldenhar Syndrome (Oculoauriculovertebral Syndrome). 2 of babies were single umbilical vein (1A1V) (Table 2). Out of 7(50%) were anomalies, just 2 (29%) cases are survived (Table 3.)

The review will also provide a preliminary database for the cases performed in CHCSJ in the past 23 years.

**Table 2:** Distribution of types of Congenital Malformation in patients with EA/TEF in CHCSJ from 1993 to 2016:

| Malformation type | Incidence | Percentage |
|---|---|---|
| Vertebral | 2 | 12.5% |
| Multiple thoracic hemivertebrae,<br>(11 pairs of ribs, fusion of right 6th ,7th ribs)<br>Sacrum bone defect | <1><br><1> | |
| Anorectal | 2 | 12.5% |
| Cardiac<br>PDA<br>VSD<br>PDA + PFO<br>PDA + PFO+ VSD<br>PDA + PFO + Mild TR<br>PDA + VSD<br>VSD + ASD + AV valve regurgitation | 8<br><1><br><1><br><2><br><1><br><1><br><1><br><1> | 50% |
| Renal | 4 | 25% |
| Absence Kidney(Right)<br>Renal agenesis<br>Hydronephrosis | <1><br><1><br><2> | |
| Limb | 3 | 18.7% |
| Short limbs<br>thumb defect, fingers camptodactyly<br>polydactyly | <1><br><1><br><1> | |

**Table 3:** Rate of complication post operation in CHCSJ: n (%)

| Anastomotic leak | Anastomotic Stricture | GERD | Tracheomalacia |
|---|---|---|---|
| 1 with recurrent TEF (14%) | 5 (71%) | 5 (1 done fundoplication) (71%) | 2 (29%) |

### Preoperative Diagnosis of Pneumonia and Ventilation

3 of the babies (19%) had preoperative pneumonia and 1 died.

## Types of Surgery and Complications

Out of the 13 babies whom underwent surgery repaired, 2 (15.4%) died (1 was confirmed that he died of non-trauma intracranial hemorrhage), another one was done delayed gastrostomy on Day 9 of birth. 4 babies lost follow up in CHCSJ after referred to Hong Kong Hospital.

7 of babies underwent surgical intervention of babies underwent surgical intervention; 5 of them were performed via right thoracotomy with transverse mid-axillary incision to advocate the extrapleural approach for EA-TEF primary repair for the division and ligation fistula and a single layered anastomosis using absorbable suture which is 5-0 PDS II (Polydioxanone), 2 of them performed transpleural primary repaired due to thoracoscopy primary repaired first unsuccessfully and for the operation time was shorter right using 5-0 Vicryl (Polyglactin 910), they were performed in CHCSJ; 1 was underwent partial artificial esophagus by Right colon in Hong Kong hospital.

The complications seen were noted early anastomotic leak 1 baby who presented recurrent pneumonia and septicemia postoperatively, he had repaired recurrent fistula referred to Hong Kong Hospital when he was 2 months age. Anastomotic stricture 5(71%) babies, 3 of needs balloon dilatation via OGD-endoscopy and 1 (Type A, colon artificial esophagus) needed to insert stent. and gastroesophageal reflux were 5 (71%) babies, needed Proton pump inhibitor treatment, 1 of them had fundoplication. 2 (29%)were tracheomalacia. The complications were diagnosed based on clinical symptoms and radiological imaging. 1 of tracheomalacia had undergone tracheostomy and gastrostomy, another tracheomalacia was lost of follow up.

### Mortality Rate

The number of deaths in this review was 5 of the 16 babies, which gives a mortality rate of 31.25%. The causes of death were multiple congenital malformations with cardiac malformation (3), pneumonia (1), non-intracranial hemorrhage (1).

**Table 4:** Survival in patient with EA in our series based on Waterston Classification (n=14)

| Waterston classification | Total case | Survival |
|---|---|---|
| | Number (%) | Number (%) |
| A | 4 (29%) | 4(100%) |
| B | 3 (21%) | 3(100%) |
| C | 7 (50%) | 3(43%) |

**Table 5:** Comparison of survival rate of patient with EA/TEF based on Waterston Classification

| Waterston Classification | Spitz 2006 (London)[4] | Azim 1999 (HUSM, Kelantan, Malaysia)[5] | HSB 2009 (Alor Star, Kedah, Malaysia)[6] | In CHCSJ (Total 16, Lost F/U 4) |
|---|---|---|---|---|
| A (100%) | 99% | 100% | 100% | (3) 100% |
| B (85%) | 93% | 71.4% | 89% | (3) 100% |
| C (65%) | 71% | 51.9% | 33% | (1) 14% |

## Discussion

Although over 23 years, the review sample size is too small to compare the data statistically, this review demonstrated associated anomalies, Waterston Group, and Type of EA/FET.

The outcomes measured included time from birth to operation, leak rate, recurrence fistula rate, presence of stricture and need for dilatation and gastro-esophageal reflux disease requiring fundoplication and mortality are considered. Further data demonstrated here on above tables.

The mortality rate in this review was 31.25%. According to Waterston classification, Spitz observed the survival rate of 71% in his Group C infants, as shown in Table 5. This is much higher than HUSM, HSB in Malaysia and us.

We noted the congenital malformation significantly affect an outcome of the babies after surgery for EA/TEF.

The improvement survival rate is largely attributable to advance in neonatal intensive care, ventilator, neonatal anesthesia, nutrition support, antibiotic, early surgery intervention and surgery materials and technique, and thank for our neonatologist and intensive care and multidisciplinary team work.

Indeed, mortality is currently limited to that case with coexisting severe life-threatening anomalies.

The diagnosis of EA is most commonly made during the first 24 hours of life but occurs either antenatal or may be delayed.

The primary surgical correction for EA and TEF is the best option in the absence of severe malformations.

The primary complication during the postoperative period are leak, stenosis of anastomosis, gastro-esophageal reflux, esophageal dysmotility, fistula recurrent, respiratory disorders and deformities of the thoracic wall.

Neonatal anesthesia in EA w/TEF, especial in Type C and D, is very difficult. Newborns with prematurity or severe lung disease can be difficult to ventilate; the ventilatory gases can easily flow down the low-resistance fistula, and this is worsened if lung compliance is poor. The result is inadequate ventilation and gastric distention; the latter can further impede ventilation or even cause gastric rupture and pneumoperitoneum.

When surgical procedure, lung retraction can be tolerated poorly, requiring high FiO2 and altering ventilation settings. Direct large airway compression can also occur; for these reasons so many anesthetists prefer to hand ventilate throughout these periods for this altered respiratory physiology, it is difficult to maintain ventilation until the time of tracheoesophageal fistula ligation.

The ventilation problems occur due to gas preferential owing into the fistula rather than lungs. Various anesthetic and surgical maneuvers are possible; the choice depends on the clinical urgency and the plan which made between the anesthesiologist and the surgical team [7].

Overall, the incidence of this disease in Macao is similar to that in the field. The complication rate of surgery is not too high. Should this surgery be maintained in this hospital with a low operating rate?

EA and TEF were successfully managed in the pediatric unit in CHCSJ. Survival and outcomes were similar to those of a specialist children's hospital. Therefore, this demonstrates that pediatric neonatal and surgical service provides effective treatment comparable with that of a world –class facility.

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