

Is tracheomalacia a postoperative complication of Oesophageal Atresia with Tracheo-oesophageal Fistula ?

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Abstract

Postoperatively survived 24 oesophageal atresia (EA) with tracheo-oesophageal fistula (TEF) infants were studied. Development of postoperative tracheomalacia (TM) is whether due to anomaly of tracheal structure or by surgical manipulation is not yet known well. We analyzed these factors in our infants. The onset of TM and tracheal structural anomalies in relation with sex, types of EA, TEF communicating level and gap between two oesophageal ends were analyzed. Surgical procedures during primary radical operation were evaluated according to the division of azygos vein or without it. A total of 11 infants developed postoperative TM within 98 ± 88 days; 38% postoperative EA with TEF infants developed TM; only 14% of them had severe symptoms and needed aortopexy. Post-aortopexy (83%) improved well and are comfortable, for more than 33.8 ± 35.3 months. However, mild to moderate symptomatic infants, managed conservatively, also improved and are asymptomatic for 88 ± 46 months in the recent follow-up.

Male predominance was observed significant ($p = 0.004$) for the development of postoperative TM. Those who developed postoperative TM had only Gross C type of EA. Statistically Gross C type of EA showed significant relation ($p = 0.02$) with postoperative TM, whereas other types of EA could not show significant relation between them. However, statistical relationship with postoperative TM between TEF communicating level, gap between two oesophageal ends and division of azygos vein or without it, could not show significant differences. These findings suggested that sex of infants and type of EA play a significant role, and surgical procedure has only relative association to develop postoperative TM in EA with TEF corrected infants.

Keywords: Oesophageal atresia; Tracheo-oesophageal fistula; Postoperative Tracheomalacia.

Introduction

Infants born with congenital anomalies, as oesophageal atresia (EA) with tracheo-oesophageal fistula (TEF), is an emergency condition and in recent trend, most of them are managed surgically as early as possible. After definitive surgical procedure occasionally some infant develop tracheomalacia (TM). Whether this condition is congenitally associated with EA and pronounced postoperatively or an acquired condition of surgical procedure are not mentioned enough in the literature. Very limited publications are available on the development of postoperative TM in EA with TEF corrected infants. Some of the infants with congenital TM manifest early symptoms within first few days of their life, but some do not show until two months. Most of these infantile symptoms disappear usually within about 18 months of age, but has been reported remaining longer in some cases.¹⁴ These infants may need treatment as early as possible or could be severe symptomatic even in mild respiratory tract infections.

We reviewed hospital records of our patients, aiming to explore the onset, incidence and possible risk factors for the development of postoperative TM in EA with TEF infants. The available records were analyzed according to the structural anomalies of the trachea in relation with sex, types of EA, TEF communicating level, gap between upper and lower oesophageal ends. The primary radical surgical procedures were evaluated in two groups as division of azygos vein or without it, to identify their association with postoperative tracheomalacia.

Materials and methods

Out of 31 EA with TEF infants in the past 11 years, 21 infants survived in our hospital post operatively. Three postoperated infants of EA with TEF were referred to us for the management of TM. Hospital records of these 24 infants were analyzed for the mentioned purpose.

Mostly dying spells, near miss syndrome or severe cyanotic attack (cyanotic spell) and history of repeated barking cough, persistent stridor and wheeze or difficulty in extubation of endotracheal tube were the main

presenting symptoms among these TM infants. Initial diagnosis was made by clinical symptoms, simple radiological study and later under general anesthesia, confirmed by rigid ventilating bronchoscope.

Mild to severe tracheal collapse was observed on bronchoscopic examination at the lower part of trachea, mostly around the carina level in all infants with TM. Two of them were associated with postoperative TEF site granulation and three were along with also bronchial collapse.

Available data on post operative TM were divided into two main groups;

(a) Symptomatic TM infants and

(b) Asymptomatic TM infants.

Symptomatic TM infants were further divided into two sub groups according to the severity of presenting conditions. (i) Severe symptomatic TM infants, infants with acute symptoms of dying spells or severe cyanotic attacks or history of near miss and confirmed severe tracheal collapse on ventilating bronchoscopy were considered as severe symptomatic TM infants. (ii) Mild to moderate symptomatic TM infants: Infants with clinically mild to moderate symptoms and mild to moderate degree of tracheal collapse on bronchoscopy were grouped in this category. Asymptomatic TM infants were observed in yearly interval even though they were without symptoms.

Severe symptomatic TM infant's airway was evaluated with contrast radiological imaging study (aortography with contrast bronchography), CT scan and Magnetic Resonance Imaging (MRI) to identify the severity of the malacia segment and level. Mild to moderate symptomatic TM group infants were evaluated on regular intervals either clinically or bronchoscopically according to the severity of the presenting problems.

Severe symptomatic TM infants were managed with surgical aortopexy through mid sternal incision approach without using any internal or external tracheal stents. Surgical procedure as aortopexy or aorto-right innominate arteriopexy or pericardial flap aortopexy was decided individually during the surgical procedure by direct lifting of aorta and observing simultaneously elevation of the tracheal wall and widening of tracheal lumen on direct bronchoscopy.

The data were analyzed through Macintosh computer and compared with the two-way Chi-Square test using Stat view package 4.01 version. The data are presented in mean \pm standard deviation and $p = <0.05$ are considered significant.

Result

Out of 24 EA with TEF infants, 11 developed symptoms of postoperative TM (eight in our series and 3 referred cases) within 98 ± 88 (19-318) postoperative days. There were male predominance, which was statistically significant ($p=0.004$) to develop postoperative TM. The incidence of postoperative TM was 38% (8 of 21) in our series. However, only 14% (3 of 21 in our series) and all three referred cases developed severe symptomatic TM and needed surgical aortopexy. All postoperative TM infants had only Gross C types of EA. (Gross C type of TEF is blind upper oesophageal pouch without communication with trachea but lower oesophagus communicates with trachea making a fistula.) The relation of Gross C type of EA with postoperative TM was statistically significant ($p=0.02$) than other types. Whereas TEF communicating level ($p=0.5$) [even 82% (9 of 11) infants had fistula communication at 1-2 cm above the carina level], oesophageal gap ($p=0.2$) [even 64% (7 of 11) infants had less than 2 cm gap between the two oesophageal ends] and division of azygos vein ($p=0.19$) [about 53% (9 of 17) azygos vein divided group developed postoperative TM] could not show statistical significance with postoperative development of TM (Table I).

Preaortopexy investigation as aortography, contrast bronchogram, CT scan and MRI in severe symptomatic infants ($n=6$) showed lower tracheal compression either by right innominate artery ($n=3$) or by aortic arch ($n=3$).

Six severe symptomatic TM infants were managed with surgical aortopexy ($n=2$), aorto-right innominate arteriopexy ($n=1$) or pericardial flap aortopexy ($n=3$). 5 infants (83%) improved well and are asymptomatic in an average follow up of until 33.78 ± 35.32 months (13-92 months). One patient with multiple congenital anomalies died 9 months post-aortopexy due to congenital cardiac anomaly problem. Recurrence of the symptoms and failure of aortopexy technique were not complained among these infants until 8 months to seven and half years of follow up period.

Whereas five cases with mild to moderate symptomatic TM, were managed symptomatically, also improved the symptoms well and are clinically asymptomatic for more than 7.3 ± 3.8 years (ranging from 3 years 3 months to 12 years 4 months) in recent follow up.

Table I:

Conditions	Without TM (n=13)	With TM (n=11)	(Severe TM) (n=6)
a. Sex*			
Male	2	8	(4)
Female	11	3	(2)
b. Oesophageal atresia**			
C type	8	11	(6)
H type	3	0	(0)
A type	1	0	(0)
B type	1	0	(0)
c. Level of TEF			
Carina (1-2 cm)	9	9	(6)
Middle part of trachea	2	1	(0)
Not mentioned	2	1	(0)
d. Oesophageal gap			
1-1.5 cm	2	7	(4)
2 cm	4	2	(2)
> 2 cm	2	2	(0)
Overlapping	2	0	(0)
Not mentioned	3	0	(0)
e. Azygos vein			
Divided	8	9	(4)
Not divided	4	1	(1)
Records not available	1	1	(1)

* p = 0.004 ** p = 0.02

Discussion

Management of congenital EA and TEF has been well success in recent years, even then postoperative TM in some infants is still remaining a big challenge.³ Infants and children presenting with barking cough, stridor, expiratory wheeze, history of repeated respiratory infections, cyanotic spell or near miss syndromes after surgical repair of EA with TEF need to evaluate immediately with bronchoscopic examination to rule out the possibility and or severity of TM. Further management needs to plan accordingly. Compression of trachea by thoracic great vessels in infants with TM are the commonly observed problems by different authors.^{1,4,8,10,17,19} However tracheal compression by right innominate artery and aortic arch were the main reason of severe symptoms in our cases. Why this problem is common ? is still unanswered. Different authors have advocated various reasons for the development of postoperative TM but non of them are agreeable to all. The diversity of their opinions are:

a. Congenital softening or abnormality in the laryngeal and tracheal wall will loose its consistency in presence of compression by vascular rings or oesophageal atresia.^{1,9,11}

b. Lack of segmental blood supply to the lower oesophagus after surgical correction of EA and TEF division is responsible to develop tracheomalacia according to the severity of the ischemia developed.²

c. TM is exceptionally a primary disease¹¹ of generalized or localized weakness of tracheal wall resulting in excessive narrowing of the lumen especially during expiration and/or in raised intrathoracic pressure.^{13,19}

d. Embryological developmental alteration of trachea is responsible for TM.¹⁵

However, EA of A and H types did not show postoperative TM in our series, probably because of there were no tracheal structural anomalies. Whereas all of the postoperative TM-developed infants had only C type of EA. The fistula communicating with trachea in type C of EA, is expected to have developmental alteration. Such difference in anatomy may lead to physiological changes which maybe responsible to develop TM.¹⁵ This study also favours the possibility of anatomical differences and physiological changes. Kao *et al*⁹ suggested that size of oesophageal pouch or site of TEF communication always does not correlate with the degree of tracheal collapse. These facts suggest us to think about the possibility of either abnormal development or postoperative deviation of thoracic great vessels from normal position in EA with TEF infants. Further study and subsequent evaluation is necessary to explain them in detail.

In our experience, the presence of multiple congenital anomalies, surgical complications (leakage, abscess or adhesions) and multistage surgical manipulation in and around the trachea or oesophagus, have been developed postoperative TM more frequently than single stage procedures. The onset of postoperative TM found by other authors is from 3 weeks to 10 months but in our series it was 14 ± 12.5 weeks in an average (from 19-318 postoperative days). The incidence of postoperative TM was 38% in our series whereas others have mentioned from 7-37%.^{6,7,8,12,16,17} Among the TM-developed infants, only 14% needed surgical management for tracheomalacia in our series, however, Rideout *et al*¹ mentioned that 18% of TM among the postoperative EA with TEF infants needed surgical intervention in their series but they could not find significant relationship between TM with EA without TEF infants.¹⁵

The relationship of azygos vein division and without it with postoperative TM in our series could not show any statistical significance though the majority of the infants (53%) had azygos vein division during primary radical management. It suggests that surgical technique has very minimal role to develop postoperative tracheomalacia.

This study suggests that surgical procedure is only a relative risk factor but developmental alternation could have played major role to develop postoperative TM in EA with TEF infants. Avoidance of multi-staged procedure, minimum dissection and manipulation around oesophagus, TEF and lower trachea (around the carina level) and prevention of complications may be helpful in reducing the incidence of postoperative TM in EA with TEF infants.

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