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Case Report

Surgical management of neonate presenting with type 3 b 4 variety of TEF (KLUTH)

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Abstract

Introduction: TEF is a common neonatal surgical emergency that presents shortly after birth with drooling of saliva from the mouth, inability to feed, and respiratory distress with or without cyanosis.

The definitive treatment for this condition is surgery. Essentially the defect involves agenesis of a part of the esophagus with a fistulous communication with the trachea. There are a large number of anatomical variants of this anomaly, Kluth has described nearly 96 different varieties. The type B variety involves a blind-ending upper pouch with a fistulous communication between the lower pouch and the trachea. There are 20 subtypes described for type B anomaly.

Type 3b4 variety involves the overlapping upper and lower esophageal pouches. The long TEF courses parallel in proximity to the upper pouch (UP) and the trachea. This mandates a very meticulous dissection of the upper pouch from the trachea.

Case Presentation: A 1-day female presented with presumptive diagnosis of TEF. On exploration, after ligating the right azygous vein, we found that it was a type 3B4 variant of TEF. The dissection was modified. Careful separation of the fistula from the trachea and from the undersurface of the upper pouch was done. After completely freeing the fistula, rest of the surgery proceeded as is done normally.

Conclusion: In this case report, we describe how an on-table diagnosis of this variant forced us to modify the steps of dissection so as to prevent tracheal injury. Also, the the importance of doing pre-operative bronchoscopy in diagnosing such variants, although these cannot be over-emphasised. Thoracoscopic approaches are proving to be of high value in management of these cases with significant success rates and lower complications.

Keywords: Tracheoesophageal fistula, Anastomosis, Azygous vein, Neonates, Fistula.

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1. Introduction

The congenital defect of esophageal atresia with or without TEF can have very varied anatomy. Before proceeding with the surgical management of this anomaly, one needs an understanding of the multiple variations that may occur in this disorder.[1] The comprehensive description by Kluth amply reflects the multitude of defects in this condition. He has categorized it into 10 types, each having further subtypes.[1-3]

There are 20 subtypes of type IIIb EA with distal TEF (EA-TEF). This subtype is also sometimes described as cervical esophageal atresia[4] and has an overlapping of the upper and lower esophageal segments with lower esophageal pouch fistula ending high on the trachea near the thoracic inlet.[1] In advanced centers, it is possible to establish the high termination of the lower pouch into the trachea by preoperative bronchoscopy which can enable the surgeon to carry out a trans cervical repair. However, in those institutes where pre-operative bronchoscopy is not practiced routinely, the diagnosis will only become evident at exploration.[9,10]

In a case series reported by Kemmotsu H et al, three neonates with esophageal atresia and TEF were confirmed as having an unusually high position of the distal TEF by the preoperative bronchoscopy and contrast study. The trans cervical approach for repair was chosen. The skin was incised transversely at the right supraclavicular region, and the sternocleidomastoid muscle and the carotid sheath were retracted postero-laterally. After the division of the TEF, the suture site of the tracheal fistula was covered by a flap of the stern thyroid muscle that was inserted between the trachea and the esophagus, thus avoiding opposing suture lines.[4]

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Two of these patients are symptom-free while one has hoarseness of voice. These cases emphasize that pediatric surgeons should be aware of the presence of cervical esophageal atresia in which distal TEF is located above the clavicle, and suggest that cervical repair is feasible for the patient whose distal TEF moves up above the first rib on inspiration.[4]

In this case report, we describe a case of a neonate who was operated for EA-TEF. On exploration, the neonate was found to have TEF type 3 B 4. The repair was carried out via postero-lateral thoracotomy and TEF ligation with End-to-End esophageal anastomosis was done.

2. Case presentation

A 1-day female presented to emergency with complaints of respiratory distress and peripheral cyanosis. The child was admitted to the pediatrics department with a presumptive diagnosis of pneumonia. There was no complaint of dribbling of saliva.

On passing an NG tube, resistance was felt at 10 cm. On the x-ray, there was coiling opposite the T3- T4 vertebra with air seen in the stomach and small intestine. A diagnosis of EA with associated TEF was made and the patient was taken for exploration. Ultrasonography of abdomen and pelvic also done which found normal study. On 2D Echocardiography no any significant abnormality found. X-ray of spine, no any abnormality detected.

We started with standard Right posterolateral thoracotomy. on entering 5 th intercostal space, we proceeded extra pleural approach. After ligating the right azygous vein, we looked for the fistula and found it was a type 3B4 variant of TEF. (**Figure 1**)

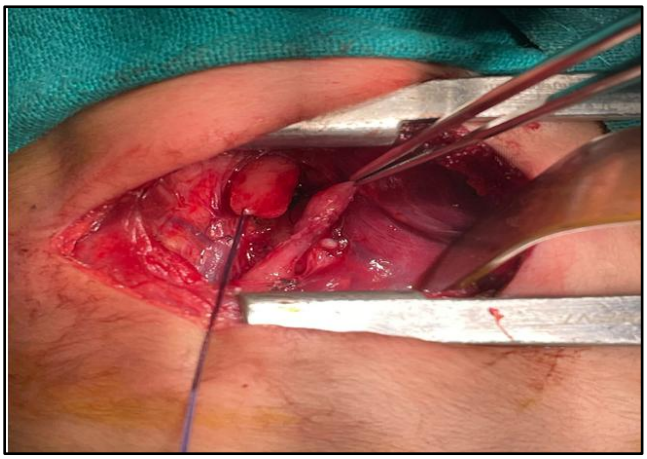

Figure 1: Intra-operative photo showing blind proximal pouch with stay suture attached and a long TEF held by a forceps.

The dissection continued, carefully separating the fistula from the trachea (taking care not to enter the trachea) and from the undersurface of the upper pouch. After completely freeing the fistula from the upper pouch and trachea, it was ligated with a PDS 6-0 suture.

The redundant fistula was excised from the lower pouch and end-to-end esophageal anastomosis was done. The posterior layer was sutured first using interrupted PDS 6-0 sutures.(**Figure 2**)

After that IFT no 6 placed via nostril was advanced beyond the anastomosis and an anterior layer of sutures was placed to complete the anastomosis. Right side 16 fr ICD was inserted and the wound was closed.

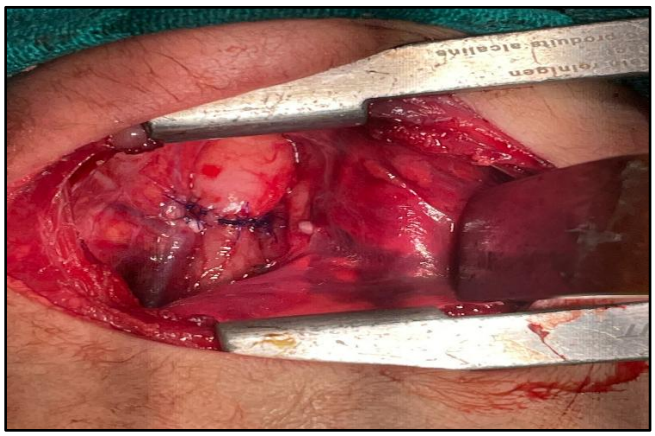

Figure 2: completed end to end esophageal anastomosis after careful dissection of both upper and lower pouch in a case with TEF type 3B4

The child was not extubated and shifted to the ward. The child was extubated on postoperative day 2.

The child was started on NG feeds on postoperative day 5. Feeds were initially given at 2ml every 3 hourly and along with feeds, oral lansoprazole and domperidone syrup were started. Since the child was tolerating NG feeds well, they were increased gradually in the following days.

On postoperative day 6, a chest x-ray was done and the ICD was removed. The wound site was healthy. The child was afebrile and passing stools.

On postoperative day 9, the child was started on breast feeds which the child accepted well. NG was removed the subsequent day and the child was discharged satisfactorily. At present the child is being followed up in OPD and is healthy and asymptomatic.

This case report shows that in centers where routine preoperative bronchoscopy is not done, intra-operative findings of TEF type 3B4 can be a cause of surprise to the surgeon. However, very careful separation of the fistula from the upper pouch as well as the trachea will prevent any on-table mishap and the child can have a successful outcome.

3. Discussion

The wide variations in the anatomy of TEF can present a serious challenge on the table to the pediatric surgeon. This problem is complicated by the fact that many times the

esophageal pouches are closely adherent to the trachea, bronchus, great vessels, vagus nerve, etc, and dissection from these structures can lead to their injuries.

Various anatomical classifications for TEF described are given in the table below.(**Figure 3**)[1]

Vogt (1929)	Gross (1953)	Kluth (1976)	Description of anomaly
1		I	Absent distal esophagus
2	A	II	Pure EA
3a	B	IIIa	EA with proximal TEF
3b	**C**	**IIIb**	**EA with distal TEF (20 subtypes)**
3c	D	IIIc	EA with proximal and distal TEF
	G	IV	Membranous atresia
		V	EA with esophageal duplication
		VI	Esophagobronchial communications
4	E	VII	H fistulas
	F	VIII	Esophageal stenosis
		IX	Tracheal atresia or agenesis with TEF
		X	Tracheoesophageal fissures or clefts

EA—Esophageal atresia, TEF—tracheoesophageal fistula.

Figure 3: Various anatomical classifications for TEF

Subtypes of IIIb with variations in esophageal segments	Subtypes of IIIb with variations in TEF	Subtypes of IIIb with laryngeal and tracheal anomalies	Rare subtypes of IIIb
$IIIb_1$-short gap EA TEF $IIIb_2$-long gap with fibromuscular band between the esophageal segments. $IIIb_3$-overlapping of the esophageal segments due to long upper pouch. $IIIb_4$-same as IIb_7, but with additional membranous atresia in the lower pouch. $IIIb_5$-same as $IIIb_1$, but with additional stenosis of the lower esophageal pouch. $IIIb_{10}$-lower esophagus and fistula being replaced by multiple cysts. $IIIb_{11}$-same as $IIIb_1$, but with multiple cysts at the lower esophagus. $IIIb_{15}$-same as $IIIb_1$, but with upper pouch duplication.	$IIIb_6$-high TEF with no overlapping of esophageal segments, due to high upper pouch. $IIIb_7$-high TEF with overlapping of esophageal segments. $IIIb_8$-same as $IIIb_4$, but the fistula is atretic. $IIIb_9$-same as $IIIb_1$, but with closed tracheal end of TEF. $IIIb_{12}$-same as $IIIb_1$, but with 2 distal fistulae opening into the trachea. $IIIb_{13}$-same as IIb_{12}, except that one fistula opens into right main bronchus.	$IIIb_{16}$-same as $IIIb_1$, but with laryngeal stenosis. $IIIb_{17}$-same as $IIIb_1$, but with laryngeal atresia. $IIIb_{18}$-same as $IIIb_1$, but with tracheal stenosis below the distal TEF.	$IIIb_{14}$-the external continuity of the esophagus is maintained with atretic segment between the two esophageal segments. $IIIb_{19}$-high EA with a common tube between the larynx and the stomach, with tracheal and esophageal characteristics. $IIIb_{20}$-same as $IIIb_1$, but with agenesis of both lungs and atresia of both bronchi.

Figure 4: Kluth classification showing subtypes of Type 3 TEF

The type III variety is the most common variant and within this type, there are 20 different subtypes as classified by Kluth.(**Figure 4**)[1]

Anomalies in tracheo-esophageal development result in a spectrum of congenital malformations ranging from, most commonly, esophageal atresia with or without trachea-esophageal fistula (EA+/-TEF) to esophageal web, duplication, stricture, tracheomalacia and tracheal agenesis.[11] With such varied anatomical presentations, they must present a serious challenge to the surgeon on the table. Coupled with the fact that in most centers especially in the developing world, pre-operative bronchoscopy is not routinely done to ascertain the site fistula and/or the anatomical variant of TEF. In such circumstances, it is imperative to follow the surgical principles as described by experts.

4. Conclusion

We have presented this case report to emphasize that in a case one encounters the Type 3 B 4 variant on the table, one must carefully locate both the upper and lower pouch as long overlapping pouches can sometimes confuse and appear as though the esophagus is in continuity. Secondly locating the fistula can be challenging as the fistula with the lower pouch is overlapped by the upper pouch. Thus, this case report emphasizes the surgical steps that need to be taken in case of seeing this ‘on-table surprise.

5. Source of Funding

None.

6. Conflict of Interest

None.

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