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Tracheo-esophageal fistula in neonates; role of radiologist in the diagnosis and management

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Abstract
Tracheoesophageal fistula (TEF) is one of the most common neonatal emergencies. Polyhydramnios was the most frequent finding and stomach size and presence were not contributory to the diagnosis antenatally. The common presentation was excessive salivation and vomiting, respiratory distress after birth, recurrent pneumonia later in life. The incidence is 1 in 3000 to 1 in 4500 live births. Early recognition, prompt and efficient management of the cases was possible due to multidisciplinary approach by the radiologist, neonatologist, intensivist, and the paediatric surgeon. A precise surgical technique with proper mobilization of upper pouch and good anastomosis is key to successful outcomes.

Keywords: tracheo-esophageal fistula, neonates, diagnosis and management

Introduction

The incidence of tracheoesophageal fistula (TEF) is 1:3000–4500 of live births. TEF is a congenital anomaly of esophagus and trachea that manifests within the first few hours to days of life. The most common defect is esophageal atresia (EA) with distal TEF (type C/IIIB as described by Gross and Vogt). The risk factors are prematurity, low birth weight, difficult airway of neonate, associated respiratory distress because of repeated aspirations & congenital heart diseases (CHD), and large defects. Simple naso-gastric tube intubation is required to make diagnosis where difficulty is seen in intubating and curling of naso-gastric tube is diagnostic. Importantly, this approach to "confirmation" cannot determine the anatomic subtype of EA/TEF, the number or location of TEFs, the size of the gap between proximal and distal esophagus, or the presence of tracheomalacia.

Computed abdominal and lumbar ultrasound for vertebral anomaly, tomography scan, are necessary to recognize associated anomalies. We report TEFs in neonates, and 1 year experience in a tertiary care.

Material and Methods

Neonatal patients referred to our department for chest x-ray and contrast esophagogram whose detailed history were taken, like age, gender, ANC scan details, presenting complaints and sepsis were subjected to usg abdomen, lumbar ultrasound for associated anomalies and followed up for type of surgery, post op complication and recovery.

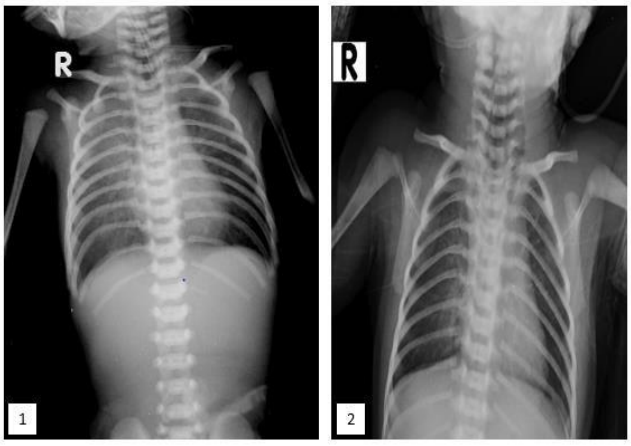

Fig 1 & 2: Frontal radiogram of chest showing coiling of nasogastric tube.

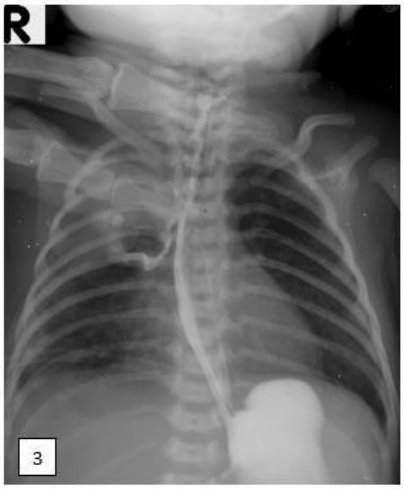

Fig 3: Frontal radiogram of chest showing post anastomotic leak of contrast into the mediastinum.

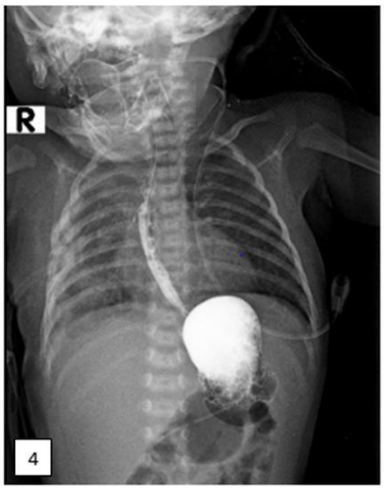

Fig 4: Frontal radiogram of chest showing post op re-repair with no leak of contrast

Results

Table 1: Baseline characteristics of neonates

Basic characteristics	Total=12
Gender	
Male	11
Female	1
Birth weight (g)	
≥2500	9
≤2499	3
Associated congenital anomalies	
Present	1 VATER
Absent	11
Type of esophageal atresia	
Type A	1
Type B	2
Type C	8
Type D	0
Type E	0
Type F	1
Surgical management	One stage operation
Outcomes	
Death	2
Complication: Sepsis and pneumonia	2
Anastamotic leak, Reoperation	2

12 cases of tracheo-oesophageal fistula were investigated and managed at our centre n=12.M=11(91.1%),(, F=1(8.33%) aged between 2 days3days n=11, to 22days n=1. All neonates presented with complaints of excessive salivation and vomiting. All the cases underwent chest radiogram (Figure 1 and 2), oesophagogram with oral non-ionic contrast media (Figure 3 and 4) and confirmed the diagnosis. One case was antenatally detected because of persistent small sized stomach and was confirmed to have oesophageal atresia & tracheo-oesophageal fistula.

Per-operative diagnosis of our cases was type a=1(8.33), b=1(8.33%)), c=8(66.6%), f-1(8.3%). One case had VATER anomaly, the most common was type III that is, EA with distal tracheo-oesophageal fistula n=8. (66.6%). The birth weight of our cases are ranged between --1800 to 2490 gms.

Two cases were reoperated for anastamotic leak (Figure 3). The mean age at presentation was 2.5 days in 11 cases one at 22days old.

Two cases died due to sepsis and pneumonia (16.6%).

Discussion

Patients of EA with TEF are prone to have other associated congenital anomalies, the presence of which adversely affects the outcome. TEF may be associated syndromes and anomalies such as VATER or VACTERL with frequent cardiac and anorectal defects [1]. The literature incidences of associations - 40% VSD, 30% PDA, and 20% anorectal malformation. The diagnosis of

EA may be suspected in ANC scan -the parameters are polyhydramnios, absent fetal stomach and Pouch sign. Presence of a small or absent stomach bubble on a routine ultrasound examination performed after the 18th week of pregnancy and the presence on an ultrasound of excessive amounts of amniotic fluid (polyhydramnios) raises further suspicion of EA. However, polyhydramnios alone is a poor indicator of EA because polyhydramnios has numerous, varied causes [13]. In cases where EA/TEF is not suspected or detected before birth, it may be suspected within a few hours of birth when an affected newborn is unable to swallow, has excessive mucous, or has breathing difficulties A classification system of EA/TEF into different subtypes.

Although this classification system is commonly used, it is not universal. Type C is the most common form. The other types affect less than 15 percent of individuals with EA/TEF. Under the classification, the subtypes include,

TYPE A

Only EA is present (there is no TEF). The esophagus is separated in two with both the upper and lower portions ending in blind pouches. This is sometimes referred to as pure esophageal atresia and accounts for approximately 8 percent of cases. We had one case of type A n=1

TYPE B

The lower segment of the esophagus ends in a blind pouch and the upper segment is connected to the trachea via a TEF. This form is rare, accounting for approximately 2 percent of cases. We had one case of type B n=1.

TYPE C

The upper segment of the esophagus ends in a blind pouch and the lower segment is connected to the trachea via a TEF. This is the most common type of EA/TEF occurring in approximately 85 percent of individuals. We had type C cases n-8.

TYPE D

A TEF is present connecting both the upper and lower segments of the esophagus to the trachea. This is the rarest form of EA/TEF affecting less than 1 percent of cases.

TYPE E

In this form, the esophagus is intact and connects normally to the stomach. However, a TEF is present connecting the esophagus and the trachea. This is also known as H-type fistula. (4 percent).

TYPE F

has been added ie esophageal stenosis (GrossRE; Surgery of infancy andchildhood. philadelphia, WB Sanders, 1953. We had type f case n-1.

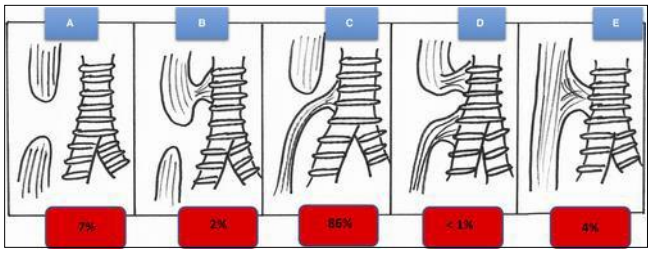

Fig 5

Gross classification of esophageal atresia.

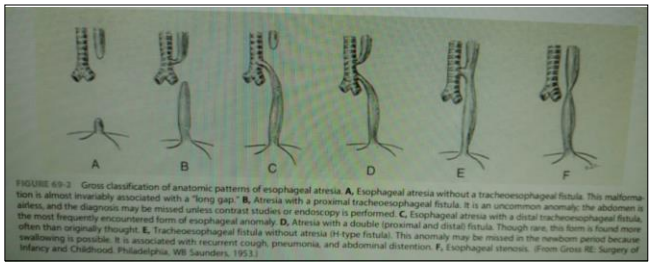

Fig 6

H-type TRACHEO-OESOPHAGEAL FISTULA (Type C) accounts for 4–5% of all congenital tracheoesophageal malformations [6]. The reported incidence varies from 1.8%[8] to 4.2%[9] amongst all types of tracheo-oesophageal fistula. We found an incidence of 66% over a period of 2 years. The classical symptoms of an H-TEF are recurrent chest infections, cyanosis and choking on feeding and abdominal distension. The investigation of an infant with a suspected H type TEF needs to bear in mind two objectives – first, confirmation of the diagnosis, and second, location of the site of the fistula – because this will influence the approach to subsequent surgical repair [2].

Associated congenital anomalies are more common with other variants of oesophageal atresia with TEF whereas H type TEF has a least association with about 30% of cases.6 The associated abnormalities reported in the major series are VACTERL/VATER association, Ventricular septal defect, Vascular ring, Fallot's tetralogy, Chromosomal anomaly, Duodenal atresia, Renal anomaly, Vertebral anomaly, CHARGE association, Goldenhaar's syndrome, syndactyly, oesophageal stenosis, Laryngeal cleft, Malrotation, Imperforate anus and Trisomy. We found no congenital anomaly in the first case while in the second case there was a small VSD along with anterior ectopic anus[3], Radiological and endoscopic procedures ie Tube oesophagograms, Tracheobronchoscopy, are complementary in both diagnosis and treatment of H-type TEF [2]. Sundar *et al.* did not find bronchoscopy or oesophagoscopy helpful, perhaps because skilled radiological demonstration of the fistula in each case made endoscopy unnecessary. Hays *et al.* in (1966) found oesophagoscopy unhelpful. Bedard *et al.* in 1974 showed the fistula at bronchoscopy in 12 out of 15 patients whereas found oesophagoscopy alone to be useless.

Upper gastro-intestinal endoscopy or rigid oesophagoscopy has no role in the diagnosis of an H type TEF as the oesophageal ostium of a TEF is simply too small and well hidden in the folds of mucosa of the oesophagus to be seen. Computed tomography and virtual bronchoscopy for the diagnosis of H-TEFs but whether this technique will be of clinical value remains to be seen. Prenatal diagnosis of TEF may be suspected from maternal polyhydramnios and absence of the fetal stomach bubble. In a study by Stringer *et al*., prenatal scans diagnosing EA had a sensitivity of 42% with a positive predictive value of 56% [5]. Ultrasound imaging may also reveal cardiac defects, which indicate a worse fetal prognosis.

Three-dimensional CT scanning also has been utilized for the diagnosis of TEF [4, 9, 12].

MR images could be used for diagnosis, and localisation of an H-type TEF could be detected safely and accurately in a sick preterm infant[11]. We have no experiences on both CT & MRI due to Surgeon, admn & pts constraints. Waterston's classification is still relevant in developing countries where it is a good predictor of survival as evident in literature. In this regard perhaps, the new preoperative risk classification proposed by Yagyu*et al*. suggested a better outcome

Common complications after EA and TEF repair in our series of - cases included anastomotic leak (16%), esophageal stricture (35%), and recurrent fistulae (3%). Esophageal stricture can be successfully managed with endoscopic balloon dilation. Tracheomalacia occurred in 15% of cases; 40% of these patients required surgical repair [12].

The first successful primary repair of TEF was performed by an American surgeon, Cameron Haight, in 1941. Because pediatric surgical centers now have a survival rate greater than 90% for these patients, the emphasis is now on reducing morbidity and enhancing these patients' quality of life[10]. Exact location of EA-TEF, the radiological as well as endoscopic procedures are recommend preoperative or intraoperative bronchoscopic guidewire trans-fistula placement which has several advantages. Firstly, such a wire can be passed through a flexible bronchoscope, and it can be identified on a chest roentgenogram so we can verify the exact location of the fistula. Secondly, the wire can be palpated intraoperatively, which facilitates and simplifies identification of the fistula and preservation of surrounding structures. Open surgical repair of TEF/EA involves a right posterolateral thoracotomy, fistula ligation, and the creation of a primary esophageal anastomosis[6]after thorough preoperative evaluation [10].

Motility disorders and respiratory function abnormalities are common after EA and TEF repair and warrant monitoring. A systematic review of long-term outcomes in adulthood after EA repair during infancy reported the following pooled estimated prevalences [8]:

Dysphagia: 50.3%, GERD with esophagitis: 40.2%, GERD without esophagitis: 56.5%, Respiratory tract infections: 24.1%, Asthma: 22.3%, Wheeze: 34.7%, Persistent cough: 14.6%, Barrett esophagus: 6.4%, Squamous cell esophageal cancer: 1.4%.

The differential diagnosis of TEF/EA to be considered are stricture or diverticulum, pharyngeal pseudodiverticulum, severe GERD, vascular ring, iatrogenic esophageal perforation, laryngo-tracheo-esophageal cleft, esophageal webs, esophageal duplication, congenital shortened esophagus, and tracheal agenesis or atresia.

Conclusion

Simple naso-gastric tube intubation and contrast esophagogram is required to make diagnosis where difficulty is seen in intubating and curling of naso-gastric tube is diagnostic. Contrast esophagram has been traditionally performed in order to detect the location of the dilated upper esophageal pouch in relation to the thoracic inlet and to detect a proximal TRACHEO-OESOPHAGEAL FISTULA but can give false-negative results (fistula is occluded by mucus), false-positive results (contrast leak from larynx). Moreover, this procedure involves radiation hazards and may be associated with complications, including aspiration pneumonia.

CT (SSD images) may have a complementary diagnostic role in congenital esophageal atresia facilitated appreciation of complex anatomic features of the anomaly by the surgeon, enabling a better orientation before surgery.

In cases of suspected neonatal EA/TEF, structural MRI is a safe, non-ionizing, non-sedated imaging method that allows for improved pre-operative planning, parental counseling, and post-operative evaluation of repair efficacy.

Source of Support

Nil

Ethical clearance

Obtained

Conflict of Interest

None.

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