Case Report

Congenital tracheoesophageal fistula: An anesthetic challenge

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ABSTRACT

Managing a patient scheduled for congenital tracheoesophageal fistula (TEF) repair is challenging for the anesthetist. If an appropriate ventilation strategy is not employed, serious complications such as hypoxemia, gastric distension, and pulmonary aspiration can occur. We present the case of a 2-day-old male child, suffering from an isolated TEF without esophageal atresia (type H) scheduled for an open surgical repair performed by transthoracic approach (right thoracotomy). We successfully managed this intervention and herein report this case to demonstrate the multiple pre-operative, intraoperative, and post-operative complications regarding intubation that can occur while managing such a case.

Key words: H-type fistula, Intubation, Tracheoesophageal fistula

Congenital tracheoesophageal fistula (TEF) is a fistulous communication between the esophagus and trachea or a main bronchus. It has an incidence of 1 in 2500–3000 live births [1], of which H-type TEF (H-TEF), a rare congenital anomaly, has an incidence of about one in 100,000 live births [2]. This defect has survival rates of >90%, owing largely to improved neonatal intensive care, earlier recognition, and appropriate intervention [3]. Congenital TEF manifests within a few hours to a few days of neonatal life. It requires surgical correction which presents a major challenge to the pediatric anesthesiologist. Survival following TEF repair has improved over the years due to advancements in pediatric anesthesia. However, prematurity and associated cardiac anomalies significantly contribute to mortality in these neonates [4]. These may be classified into five subtypes based on the location of the fistula and the presence or absence of esophageal atresia (Fig. 1) [5]. H-TEF is a rare, life-threatening congenital anomaly, which accounts for 4–5% of all esophageal atresias/TEF [4,6]. The clinical features are variable, but the most common are recurrent respiratory symptoms, aspiration with cyanosis during feeding, and abdominal distension (Table 1) [7]. The above factors can lead to rapid respiratory failure, so early and well-planned management is necessary to prevent serious consequences.

CASE REPORT

A full-term good weight male neonate weighing 2.5 kg presented to us on the 2nd day of life with complaints of copious frothy mucus in the nose and mouth. Choking, cyanosis with feeding, signs of dehydration, and abdominal distension were also present.

On general examination, the neonate was found to be active with a pulse rate of 128 bpm regular, blood pressure of 60/38 mmHg, and SpO_2-95%. Systemic examination findings were as follows: Respiratory system showed bilateral conducted sounds. On central nervous system examination, the patient was active and crying. S1 and S2 heard on cardiovascular system examination but there was no murmur. Per abdomen was distended.

The chest X-ray findings confirm the presence of a TEF and the presence of fundic gas confirmed the absence of associated esophageal atresia (Fig. 2). A pre-operative evaluation to look for any other associated anomalies (VACTERL malformations) was negative. A 2D echocardiography was performed to rule out cardiac anomalies. Tracheal visualization of the fistula using rigid bronchoscopy has been favored historically to confirm the level of the fistula and guide a cervical or thoracic surgical approach [8-10].

Written informed consent was obtained and IV fluids (0.45% DNS), parenteral nutrition, and antibiotics were administered. All standard monitors, including pulse oximetry, electrocardiography, temperature, and non-invasive blood pressure monitoring were attached. The anesthetic management of a neonate undergoing TEF repair was very challenging. The patient was pre-oxygenated with 100% of oxygen at 5 L/min through a tight-fitting face mask for 3 min. Thereafter, premedication with I.V. Inj glycopyrrolate 10 mcg, midazolam 0.1 mg, emset (ondansetron) 0.2 mg, fentanyl sodium 5 mcg, dexamethasone 0.5 mg, and hydrocortisone 5 mg followed by induction with Inj thiopentone 10 mg i.v. The patient was gently ventilated manually to ascertain the ability

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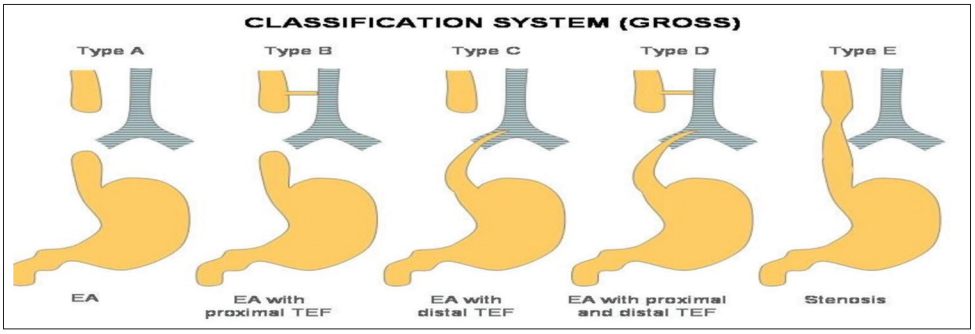

Figure 1: Classification of congenital tracheoesophageal fistula [5]

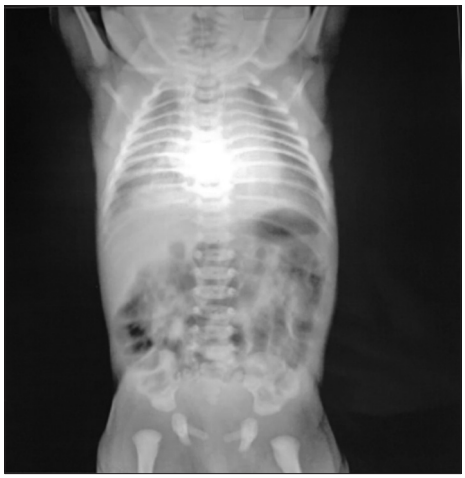

Figure 2: The chest X-ray findings showed the presence of a tracheoesophageal fistula and the presence of fundic gas

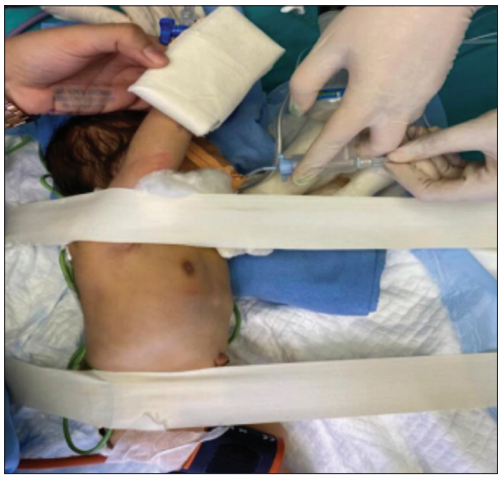

Figure 3: Positioning of the patient for right thoracotomy in the left lateral position with the right arm raised across the head and secured with padding, tapes, and gel blocks

to ventilate before giving the muscle relaxant. Succinylcholine 4 mg was injected and tracheal intubation with an uncuffed ETT No. 3 was carried out. Bilateral equal air entry was checked and confirmed and the patient was maintained on Sevoflurane and Inj Atracurium 1.25 mg after which positive pressure ventilation was started. The patient was positioned for right thoracotomy in the left lateral position with the right arm raised across the head and secured with padding, tapes, and gel blocks (Fig. 3). Thoracotomy was started (Fig. 4). A sudden episode of desaturation (56%) and bradycardia (60 bpm) was encountered, following which, a supine position was given and Inj Atropine 0.1 mg was administered. On examination, misplacement of the ETT in the fistula was identified, and hence, immediate reintubation was performed.

After surgery, during shifting, there was another episode of bradycardia (51 bpm) and desaturation (64%). Atropine and adrenaline were given in a dose of 1:1 lakh. Due to a mucus plug and secretions, there was a block in the ETT. The baby, therefore, had to be reintubated. Thereafter, the baby was shifted to the NICU and kept on mechanical ventilation for 2 days. Successful extubation was performed on day 3.

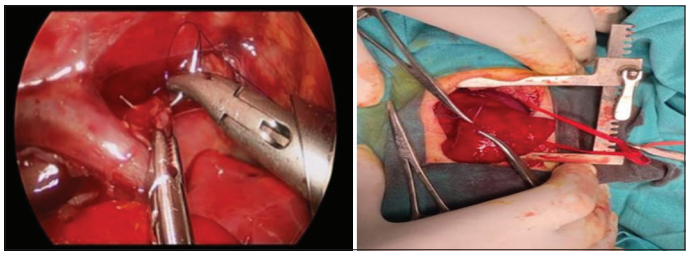

Figure 4: The thoracotomy procedure

DISCUSSION

In the above case, the baby was intubated twice due to two episodes of desaturation and bradycardia. Perioperative airway management in neonates undergoing TEF repair could be a challenge for anesthesiologists. There are basic concerns related to neonatal anesthesia due to their anatomic and physiologic differences from adults, such as greater difficulty in securing the airway, vulnerability to flip-flop circulation, less compliant ventricles, immature renal and hepatic function, susceptibility to develop hypothermia, need for very careful and strict fluid balance, risk of post-operative apnea in preterm infants, and risk of anesthetic overdose [11]. About 50% of infants are non-syndromic without other anomalies, and the rest have associated anomalies, most often associated with the vertebral, anorectal, (cardiac), tracheal, esophageal, renal, radial, and limb (VACTERL) syndrome [3].

The concerns specific to anesthetic management of TEF include the need to avoid endotracheal tube placement above or in the fistula to prevent gas insufflation into the fistula and stomach; poor lung condition due to aspiration of gastric contents and/or respiratory distress syndrome of prematurity; and associated

Table 1: Clinical features of patients with various types of congenital tracheoesophageal fistula [7]

Anomaly	Age at presentation	Predominant symptoms	Diagnosis	Treatment
Isolated atresia	Newborns	Regurgitation of feeding Aspiration	Eosophagogram* Plain film: Gasless abdomen	Surgery
Atresia+Distal TEF	Newborns	Regurgitation of feeding Aspiration	Eosophagogram* Plain film: Gasless abdomen	Surgery
H-type TEF	Infants to adults	Recurrent pneumonia Bronchiectasis	Esophagogram* Bronchoscopy+	Dilation±
Eosophageal stenosis	Infants to adults	Dysphagia Food impaction	Esophagogram* Endoscopy+	Surgery§

***Diagnostic test of choice, ±Primary therapeutic approach, §Secondary therapeutic approach, +Confirmatory test, TEF: Tracheoesophageal fistula**

cardiac or other congenital anomalies [12]. The orifice may be detected at bronchoscopy or when methylene blue dye injected into the endotracheal tube during endoscopy is observed in the esophagus during forced inspiration [3].

We need to place the tip of the endotracheal tube below the fistula but above the carina to ensure airway protection and avoid ineffective ventilation and massive gastric dilation, which can further result in gastric reflux, hypotension, and hypoxemia. For proper placement, the tube is inserted as far as possible and then is slowly withdrawn until bilateral air entry is present on auscultation. Auscultation over the stomach also helps to identify the correct location. A very important requirement while anesthetizing TEF patients is the ability to ventilate lungs without ventilation of the fistula [12]. To achieve this, it is preferable to avoid giving muscle relaxants before appropriately securing the airway. Either awake intubation or inhalation induction with spontaneous ventilation may be used to secure the airway, as positive pressure ventilation with a bag and mask may cause gastric inflation. The difficulty lies in maintaining the proper position of the ETT during surgical manipulation. During the procedure, the surgeon usually compresses the lung to mobilize the distal segment of the esophagus. This can result in desaturation which requires intermittent expansion of the lung. Other causes of intraoperative hypoxemia include endobronchial intubation; endotracheal tube obstruction due to kinking, secretions, or bleeding; kinking of bronchus or trachea; and atelectasis [12].

Postoperatively, these patients need vigilant monitoring and care in neonatal intensive care units. Accumulation of blood or secretions in the ETT can lead to airway obstruction, requiring frequent ETT suctioning. Close communication between the interventionist and anesthetist is of paramount importance, and intubation equipment like an appropriate-sized mask, endotracheal tube, laryngoscope, and a resuscitation bag should be easily available [13] in case the need for emergent re-intubation arises. It is imperative to realize the importance of a good communication system between various teams involved in patient care and the easy availability of good quality and appropriate-sized equipment such as ET tubes, bags, masks, and laryngoscopes. All these neonates require intensive monitoring in the neonatal intensive care unit. Routine care includes the use of appropriate analgesics, intravenous fluids, and antibiotics.

CONCLUSION

Anesthetic management of a neonate undergoing TEF repair is a challenging task. It may become more complex due to coexisting anomalies in other organ systems, especially cardiac anomalies. Good pre-operative assessment and preparation are required to identify problems and optimize the patient's condition. Neonates require stabilization and correction of fluid-electrolyte imbalance, hypothermia, hypoglycemia, and poor chest condition.

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