

## Tracheo-esophageal Fistula with VACTREL Anomaly: An Anaesthetic Challenge.

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### ABSTRACT

Tracheoesophageal fistula (TOF) and associated esophageal atresia (EA) in the neonate, presents during the first week of life. This congenital defect can be complicated by aspiration, respiratory distress, and other congenital anomalies. The knowledge and ability of the anesthesiologist to anticipate the challenges in managing neonates presenting for repair, plays an important role in their treatment and survival. Also, it is not uncommon for anesthesiologist to care for a patient later in life following repair of TOF. A familiarity with the immediate complications and long term outcome and sequelae after TOF repair is important to ensure the best patient outcome.

### Introduction

Tracheoesophageal fistula manifests in the neonate within hours to days of life. Considered a surgically correctable anomaly of the gastrointestinal and respiratory systems, TOF and the perioperative anesthetic considerations are of acute importance to the anesthesiologist. Prior to the first successful staged repair in 1939, esophageal atresia and associated TOF were uniformly fatal. Advancements in pediatric anesthetic techniques and monitoring, neonatology, and pediatric surgery have reduced mortality figures and survival is now higher than 90%<sup>1</sup>. Prematurity and severe associated congenital abnormalities continue to be the biggest contributors to mortality associated with TOF<sup>2</sup>.

### CASE REPORT

A 3 days old Full Term Neonate born to Primigravida by Caesarean delivery was posted for trachea-esophageal fistula repair by paediatric surgery department, patient had history of inability to pass feeding tube, recurrent regurgitation of oral feeds.

General examination revealed neonate with Low Birth Weight i.e. 2.1 kgs, with multiple congenital anomalies like Cleft lip, Cleft palate, Microtia, Syndactaly. Systemic Examination revealed Pansystolic Murmur on all four areas of heart, decreased air entry bilaterally with conducted sounds and crepitations on Respiratory Examination.

Investigations Revealed Normal Haemogram with Moderate lymphocytosis and Normal Electrolytes, Renal and Liver function Tests. Arterial Blood Gas estimation shown acidosis with anion gap of 13.6 mmol/l with SpO<sub>2</sub>

96% o air. On 2DECHO Multiple Cardiac Anomalies like ASD, VSD, PDA with Moderate Pulmonary Hypertension with preserved LV Systolic function of 70% detected. X-Ray Chest PA view revealed Coiling of Feeding tube in upper esophagus with blind esophageal end and TOF of Type-C variety.

Patient was taken for repair on Emergency basis, IV Access achieved, premedicated with Injection Atropine 0.1 mg Intravenously. Inhalational induction done with Sevoflurane 2-8% in incremental dose, Traceal Intubation done after applying Lignocaine jelly to oral cavity mucosa with uncuffed portex endotracheal tube No.3, intubated in Right main bronchus and withdrawn till air entry equalizes in both lungs, this is to place tube distal to the defect and to avoid gastric inflation. Patient was induced and maintained on Sevoflurane, and Atracurium used for muscle relaxation on Jackson-Rees circuit. Isolyte-P used as maintenance fluid calculated as per weight of the neonate. Intraoperative analgesia was provided with Injection Fentanyl 1µg/kg and paracetamol suppository. In left lateral position, Tracheo-esophageal fistula repair done with lateral Thoracotomy and positive pressure ventilation initiated after ligation of fistula. Intraoperative evidence of endotracheal tube blockade due to secretions immediately blocked Endotracheal tube removed and re-intubated with new tube. The surgery was completed uneventful and patient was reversed from neuromuscular blockade after confirming spontaneous attempts of Respiration and shifted to NICU on ventilator on SIMV mode. Patient was Extubated on 3<sup>rd</sup> Postoperative day and discharged subsequently.

## DISCUSSION

The Neonate of TOF are prone to dehydration and aspiration pneumonitis. Saliva and secretions accumulate in the upper esophageal pouch and normal swallowing is disturbed. Contamination of the lungs as a result of spillage from the pouch and/or aspiration of gastric contents through distal TOF results in atelectasis and pneumonitis.<sup>3</sup> Therefore chances of blockade of endotracheal tube are also more in these patients, so anaesthetist must have an eagle's eye on ETT.

The presence of atresia is usually confirmed by the inability to pass a nasogastric tube into the stomach. Clinical features after birth include excessive salivation, coughing, gagging, and choking, cyanosis and

regurgitation associated with attempted feeding. Pulmonary aspiration of gastric contents results in atelectasis and pneumonitis in neonates with EA and TOF. Because birth before term occurs in 30% to 40% of these neonates, respiratory distress of prematurity may also contribute to pulmonary impairment.<sup>4</sup>

Neonates with TOF and EA frequently have associated anomalies described by the acronym VACTERL. These anomalies and their associated incidences include Vertebral 17%, Anal 12%, Cardiac 20%, Tracheoesophageal fistula and Esophageal atresia, Renal 16%, Limb 10%) and other midline defects (cleft lip and palate 2%, sacral dysgenesis 2%, urogenital abnormalities 5%)<sup>5</sup> (Table 1).

Table 1:

TYPE	INCIDENCE	EXAMPLE
VERTEBRAL	17%	Scoliosis, Vertebral defects
ANAL	12%	Imperforate anus, Malrotation, Duodenal atresia
CARDIAC	20%	VSD, PDA, tetralogy of Fallot, ASD, Right-sided aortic arch
RENAL	16%	Renal agenesis/dysplasia, Hypospadias, polycystic/horseshoe Kidney
LIMB	10%	Radial anomalies, polydactyly, lower-limb defects

The Gross classification of EA and TOF describes 6 types of defects. Type A represents esophageal atresia without fistula. Type B illustrates esophageal atresia with proximal fistula. Esophageal atresia with distal fistula is classified as type C and is the most common type occurring in 80% to 90% of cases. Type D represents EA with proximal and distal fistula. TOF without atresia is classified as Type E. Type F represents esophageal stenosis.

Five types of TOF have been described based on the anatomic characteristics of the esophagus and the trachea. Type I represents esophageal atresia with no fistula. In type II, there is no atresia and a communication between the trachea and esophagus (H-type fistula) is present. Type IIIA has esophageal atresia and a communication between the upper portion of the esophagus with the trachea. In Type IIIB (Type C in the Gross classification system), esophageal atresia occurs with a blind upper pouch and the lower segment communicates with the trachea. This is the most common form of TOF. Type IIIC has atresia with both upper and lower segments communicating with the trachea.

Major co-existing cardiac anomalies are present in 20% of neonates with EA and TOF including ventricular septal

defect, tetralogy of Fallot, patent ductus arteriosus, coarctation of the aorta, and atrial septal defect. Post-natal echocardiogram is generally performed to identify such anomalies.

The neonate who presents for repair of EA and TOF represents a significant challenge to the anesthesiologist. Some of the difficulties encountered during anesthetic management include ineffective ventilation due to the endotracheal tube being placed in the fistula, massive gastric dilation, severe pre-existing lung disease from previous aspiration of gastric contents and/or respiratory distress syndrome of prematurity, and associated anomalies, particularly cardiac.

Anesthetic and surgical management focuses on ventilating the lungs without ventilation of the fistula. Techniques include awake tracheal intubation and avoidance of muscle relaxants and excessive positive pressure ventilation until the fistula has been controlled. Special attention to placement of the endotracheal tube is warranted, and gastrostomy, either preoperatively under local anesthesia or soon after induction is sometimes used to decompress the stomach and prevent gastric distentions<sup>4</sup> Hypoxemia may result from intubation of the right mainstem bronchus, endotracheal

tube obstruction by secretions or purulent drainage, bleeding, kinking of the bronchus or trachea, and atelectasis.

The goal of airway management is to ventilate the lungs adequately without ventilating the fistula. Ineffective ventilation, gastric distention or rupture, hypotension, or gastric reflux can all result from ventilation of the TOF. Current strategies to meet this goal include proper positioning of the endotracheal tube and catheter occlusion of the TOF. Problems may arise in maintaining the position of the endotracheal tube when the fistula is just above the carina. Patient movement or surgical manipulation may lead to subtle changes in the position of the tube and problems with ventilation.

Postoperative management includes admission to the neonatal intensive care unit whether or not the neonate is extubated. The need for postoperative ventilator support is based on the degree of respiratory impairment due to previous aspiration and/or respiratory distress of prematurity and associated anomalies.

Following repair of EA and TOF, gastroesophageal reflux occurs in 35% to 58% of patients probably due to intrinsic esophageal dysfunction. Postoperative esophageal motility disorders include abnormal peristalsis and impaired lower esophageal sphincter tone. In some patients with severe esophageal motility disorders and poor esophageal emptying, dysphagia may be a long term problem.

#### **SUMMARY**

The neonate who presents for repair of esophageal atresia and tracheoesophageal fistula can be especially challenging for the anesthesiologist. Anticipating potential perioperative problems and communicating with the surgeon are essential in the treatment of the

neonate with these congenital defects. Although patients with associated VACTERL anomalies have a poorer prognosis, survival rate of postsurgical repair is greater than 90%. Most children have good long term quality of life but are likely to return to the operating room later in life. Therefore, the anesthesiologist must be familiar with the peri-operative management of the neonate in need of TOF repair and the long term sequelae following repair. Lifelong problems such as gastroesophageal reflux, tracheomalacia, obstructive and restrictive ventilatory defects, airway reactivity, and recurrent pneumonia should be suspected in patients with a history of TOF repair.

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