Case Report

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ABSTRACT

Esophageal atresia (EA) is a congenital anomaly commonly found with tracheoesophageal fistula (TEF) of neonates in the first week of life. This anomaly can cause several complications including aspiration, reduction in respiration, and complication from other concomitant congenital anomalies, mostly from the heart origin. The treatment for this anomaly is a surgery. Intraoperatively, the patient may develop hypoxia due to lung retraction and hemodynamic instability from bleeding or hypothermia. Anaesthesiologists play an important role in the management of EA during the perioperative period. Careful examination of the preoperative period must be done to discover any other concomitant anomaly and complication. Good anticipation of any complication during surgery and continuous monitoring post surgery can elevate the prognosis of the patient.

Keywords: Congenital anomaly, tracheoesophageal fistula, concomitant anomaly, preoperative management, hypoxia, neonates


INTRODUCTION

Esophageal atresia (EA) and tracheoesophageal fistula (TEF) are found in 1 of every 3000 to 4500 births and remain a major challenge in pediatric surgery.¹ EA manifests in neonate within hours to days of life. Considered as a surgically correctable anomaly of the gastrointestinal and respiratory systems, the perioperative anaesthetic considerations are important to the anaesthesiologist.¹

Prior to the first successful staged repair in 1939, EA and associated TEF were uniformly fatal. Advancements in pediatric anaesthetic techniques and monitoring, neonate, and pediatric surgery have reduced mortality and the survival rate is now higher than 90%.¹,²

Prematurity and severe associated congenital abnormalities continue to be the biggest contributors to mortality associated with TEF.² Several classification systems of EA and TEF have been developed based on the presence of atresia and the location of the fistula location to the atresia. The Gross classification system describes EA with and without TEF, types A through F.³ Another well-known classification system describes the five types of TEF including types I, II, IIIA, IIIB, and IIIC. Regardless of classification type, the most common form of this anomaly is EA with distal TEF.³,⁴

Neonates with TEF and EA frequently have concomitant anomalies described by the acronym VACTERL (vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb). These anomalies and their associated incidences include vertebral anomaly (17%), anorectal anomaly (12%), cardiac anomaly (20%), renal anomaly (16%), limb abnormality (10%) and other midline defects (left lip and palate 2%, sacral dysgenesis 2%, urogenital abnormalities 5%).³ Rarely, EA and TEF may be associated with DiGeorge syndrome, Pierre- Robin syndrome, Holt-Oram syndrome, and polysplenia.³

CASE REPORT

A one-month-old male patient, 3160 grams, presented at Sanglah Hospital with a chief complaint of vomiting after every breastfeeding since he was born. Another congenital anomaly found was the absence of anal orifice. The patient had undergone colostomy-gastrostomy surgery on his second day of life without any complication. The patient received full nutrition from gastrostomy feeding. Clinical examination revealed normal respiration and cardiac function. Upon laboratory examination, there was a slight elevation of transaminase enzymes (SGOT 110 U/L, SGPT 72,9 U/L) and gamma-glutamyltransferase (172 U/L). Total bilirubin (1.07 mg/dL) and direct bilirubin (0.87 mg/dL) were also elevated.

The patient was scheduled for repair of EA under general anaesthesia. Premedication of sufasa atropine (0.1 mg) was given. Induction in this patient was done with Sevoflurane at the concentration of MAC 2.5%, alongside 5 mcg of
Fentanyl as an analgesis and 1.5 mg of Atracurium as a relaxant. Maintenance of anaesthesia was done with Sevoflurane. Controlled respiration was done in the left lateral decubitus position. Hypothermia was prevented with the use of infusion and blanket warmer. Central venous catheter was inserted in right subclavian vein.

Surgery consisted of thoracotomy and percutaneous esophagostomy. No tracheoesophageal fistula was found, only EA. The EA repair went on for three hours, with no complications and minimal bleeding. The patient was then monitored in the PICU department for three days with fentanyl 20 mcg/day as an analgesic. After three days, the patient was referred to the pediatric ward for another two days. After six days, the patient was discharged and scheduled for the next step surgery in one or two months.

DISCUSSION

Correction of EA and TEF is a major surgery. The procedure is done in a step-by-step surgery that usually precedes with primary gastrosomy, especially on neonates with low body weight (under one kilogram), no other concomitant anomaly, or unstable condition.\textsuperscript{1}\textsuperscript{,}\textsuperscript{6} Then the next step is thoracotomy to repair the main problem in the trachea and esophagus. Surgery is in left lateral decubitus position through the right fourth inter-thoracal space. The time of the surgery is between 2-4 hours with possible bleeding at 10 mL/kgBW.\textsuperscript{5}\textsuperscript{,}\textsuperscript{6} Post-surgery, the patient should be monitored in the intensive care unit. The mortality rate is between 1-20% according to comorbid and another anomalies. Complications include stricture (20-40%), leakage (10-20%), aspiration, and atelectasis.\textsuperscript{3}\textsuperscript{,}\textsuperscript{6}

The two main pathological entities in the neonate with TEF are dehydration and aspiration pneumonitis. Saliva and secretions are accumulating in the upper esophageal pouch, and normal swallowing is disturbed. Contamination of the lung as a result of spillage from the pouch and/or aspiration of gastric contents through distal TEF results in atelectasis and pneumonitis.\textsuperscript{6}

Clinical features and manifestations in the prenatal ultrasound can consist of excessive amniotic fluid or polyhydramnios with a sign of obstruction of the gastrointestinal tract. After delivery, the presence of atresia is usually confirmed with the inability to pass a nasogastric tube into the stomach. Clinical features after birth include excessive salivation, coughing, gagging, choking, cyanosis and regurgitation associated with attempted feeding. Pulmonary aspiration of gastric contents results in atelectasis and pneumonitis in neonates with EA and TEF. With preterm births occurring in 30% to 40% of these neonates, respiratory distress of prematurity may also contribute to pulmonary impairment.\textsuperscript{7}

Diagnosis of EA is usually made shortly after delivery by the inability to pass a nasogastric tube beyond 8 to 10 cm.\textsuperscript{8} Prenatally diagnosed with polyhydramnios and with no swallowing activity or visible stomach contents is highly suspicious of EA and TEF. Confirmation of the diagnosis at birth by chest x-ray shows a nasogastric tube curled up in the upper chest or neck.\textsuperscript{9}\textsuperscript{,}\textsuperscript{10}

Surgical repair is the definitive treatment for EA and TEF. Surgery is generally performed within 24 to 72 hours in otherwise healthy neonates. Delaying the surgical correction increases the risk of aspiration of saliva as a result of its accumulation in the upper esophageal pouch. Reflux of gastric acid through the lower pouch and a TEF can cause pneumonitis.\textsuperscript{11} A primary repair involves isolation and ligation of the fistula followed by primary anastomosis of the esophagus. A staged surgical repair is an alternative for neonates that are unable to tolerate surgery due to pneumonia and/or other congenital anomalies.\textsuperscript{11}\textsuperscript{,}\textsuperscript{12}

Anaesthesia management in this type of surgery includes preoperative and postoperative attention. Careful examination must be done on respiratory, cardiovascular, gastrointestinal, musculoskeletal, and hematology systems. From the respiration, neonates with TEF commonly have an insufficiency lung capacity problem, caused by prematurity or aspiration pneumonia. Chest X-ray and blood gas analysis must be obtained before surgery.

Abnormalities within the cardiovascular system usually include ventricular septal defect, patent ductus arteriosus, coarctation of the aorta, and atrial septal defect. Tetralogy of Fallot is also common. Pulmonary hypertension with the right to the left shunt is the more advanced comorbidity. Monitoring using an electrocardiogram, echocardiography, or coronary catheter can be an option. Abdominal distention is reduced with the orogastric tube. An anomaly in the musculoskeletal system also must be observed. Within the first 2-3 months of life, fetal haemoglobin is still the main cause of elevated oxygen-haemoglobin affinity. Conditions like anaemia can cause worsening of tissue oxygenation. Other supporting examinations include a routine blood test, electrolytes, and blood sugar to determine the metabolic condition of the patient.\textsuperscript{6}\textsuperscript{,}\textsuperscript{8}

A room temperature of 21-25°C, an infusion warmer, aluminium foil, and a blanket warmer must be applied to prevent hypothermia. The patients
IV fluid must be precisely calculated because of the abnormal renal function of the fetus. Ideally, a central vein catheter is applied. Before induction, suctioning of the orogastric tube must be done to prevent aspiration.

Induction in this patient was done with sevoflurane at a concentration of MAC 2.5 %, in accordance with the infant dose of this agent. Pediatric induction of anaesthesia is better done with an inhalation agent due to high minute ventilation that can elevate the distribution of the drug and a faster period of the clearance. Sevoflurane was chosen because it does not cause an elevation in the production of airway secretions, and therefore reduced the possibility of aspiration.  

Careful ventilation must be applied to prevent over-limit inspiration pressure and gastric inflation. The endotracheal tube can be inserted into the right main bronchial and then slowly retracted until a bilateral vesicular sound is obtained. The level of the endotracheal tube is rotated to the posterior side to prevent fistula ventilation. Leakage of the endotracheal tube is minimized to prevent exchange in ventilation due to the reduction of the lung compliance during surgery. High oxygen fraction is not recommended because it can cause retinopathy. Ventilation is given with a slightly higher frequency of breathing with a lower tidal volume. This is done to maintain the minute ventilation in the normal range and also to reduce the inspiration pressure to prevent any destruction of the lung parenchyma and to help the operator’s view. In this patient there was no TEF, so gastric inflation because of fistula ventilation can be eliminated.

During surgery, the lung will be retracted to broaden the surgeon’s view. This can cause desaturation especially if the dependent lung is not in optimal condition. If the desaturation worsens, the surgeon must be notified to stop the lung retraction, so the anaesthetist may provide better oxygenation for the lung to expand again. Extubation by the end of surgery must consider the severity of the anomaly and another comorbidity. Re-intubation can have a good quality of life. With proper management, many of these patients can have a good quality of life.

CONCLUSION

Neonates with EA and TEF can cause problems and challenges for anesthesiologists. Anticipation to the potent perioperative problems and good communication with the surgeon are the priority target. The patient prognosis and sequelae is determined by both the surgeon and the anaesthetist. The good prognosis rate postsurgery can be as high as 90%. With proper management, many of these patients can have a good quality of life.

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