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Case Report

Anesthesia Management of Esophagostomy for the Patient with Double Outlet Right Ventricle

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ABSTRACT

Background: Esophageal atresia is a congenital disorder in which there is no esophagus because the proximal and distal esophagus is not connected. Babies with esophageal atresia can show several symptoms: foamy mouth, cyanosis, coughing and tightness, flatulence, oliguria, or worse, pneumonia symptoms. Accompanying anomalies occur in greater than 50% of neonates with esophageal atresia. Esophageal atresia is identified by ultrasound at 18 weeks of gestation, ultrasound, and Magnetic resonance imaging (MRI) of the fetal neck, or examination of a nasogastric tube in the neck of a newborn. The management of esophageal atresia is challenging. The main choice remains the surgical procedure, which usually involves making a stoma on the proximal esophagus and gastrostomy. However, surgery has risky complications.
Case: In this case, it was reported that a 22-day-old baby with tracheoesophageal fistula (TEF) type C with Ventricular Septum Defect and Atrial Septum Defect and Double Outlet Right Ventricle (DORV) underwent esophagostomy surgery with general anesthesia.
Conclusion: Anesthesia management with general anesthesia, intubation using intravenous ketamine 3 mg, fentanyl 3µg, atracurium 1.5 mg gives stability for esophagostomy in a patient with a double outlet right ventricle.

Keywords: anesthesia management, esophageal atresia, transesophageal fistula, Double Outlet Right Ventricle

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INTRODUCTION

Esophageal atresia is a condition in which there is no esophagus or closure and constriction of the esophagus Esophageal atresia (EA) is a hereditary disorder characterized by a defect in connection between the proximal esophagus and the distal esophagus.[1] EA can occur with tracheoesophageal fistula (TEF), a congenital disorder with a fistula between the trachea and the esophagus.[2]

Double Outlet Right Ventricle is a heart defect which is congenital where the two great arteries (aorta and pulmonary artery) partially or completely exits the right ventricle. In this situation, no blood vessels leave the left ventricle, and the oxygenated blood from the left ventricle is mixed with the blood in the right ventricle so that it looks like a Ventricular septum defect (VSD).[3]

Until now, the teratogenic agent that causing this EA still unknown. However, there have been reports linking EA in families with the risk of 2%.[4] EA also associated with trisomy 21, 13, and 18. The incidence of twins is six times more than non-twins.[5]

Today, many believe that the development of EA is not genetically linked. The debate over this embryopathological process is ongoing, but little progress has been made. The old theory held that lateral infolding divides the foregut into the esophagus and trachea, but in reality, the field of human embryology does not support this theory.[1]

There are variations in esophageal atresia based on anatomical classification. According to Gross of Boston, the variations of esophageal atresia and their frequency as mentioned below:[6]

- TEF Type A - esophageal atresia without fistulas or pure esophageal atresia (10%)
- TEF Type B - esophageal atresia with a proximal TEF (<1%)
- TEF Type C - esophageal atresia with distal TEF (85%)
- TEF Type D - esophageal atresia with proximal and distal TEF (<1%)
- TEF Type E - TEF without esophageal atresia or type H fistula (4%)
- TEF Type F - congenital esophageal stenosis (<1%)

Fetuses with atresia esophagus cannot swallow amniotic fluid effectively.[7] In fetus with atresia esophagus and TEF in distal location, amniotic fluid will flow to the trachea, fistula, and then into the intestine. This condition result is the polyhydramnios condition.[8] Polyhydramnios themselves can cause premature birth. The fetus should utilize amniotic fluid so that the fetus with esophageal atresia is smaller than its gestational age.[9] Aspiration pneumonia can occur when there is the aspiration of milk or saliva. If distal TEF occurs, the lungs have a risk of being exposed to stomach acid, and also, the air from the trachea can flow down to the fistula, especially when the baby cries or is receiving positive ventilation.[10] This condition can lead to perforation of the gaster, which is often fatal.[7] Studies on manometer manipulation of the esophagus have shown that the distal esophagus is often dysmotility, with poor peristalsis or peristalsis. This will lead to varying degrees of dysphagia after continuous manipulation of esophageal reflux.[1]

Several conditions are symptoms and signs of esophageal atresia, including:[11]

- Mouth foam (air bubbles from the nose and mouth)
- Cyanotic
- Respiratory Distress and Cough
- Pneumonia symptom caused by saliva regurgitation from a blocked esophagus and backflow of gastric acid through the fistula into the trachea
- Flatulence, because the air passes from the trachea to the fistula and then to the stomach and intestines
- Oliguria
- The patient often has other congenital anomalies, like a congenital heart defect.

Double outlet right ventricle (DORV) is a defect of the heart which is congenital where the two great arteries (aorta and pulmonary artery) partially or completely exits the right ventricle.[12] In this situation, no blood vessels leave the left ventricle, and the blood from the left ventricle is mixed with the blood in the right ventricle so that it looks like a VSD. Some cases reported that there is no VSD and the left ventricle is very hypoplastic. DORV is often referred as:[3]

- Taussig-Bing malformation
- Aortic extreme dextroposition of the Tetralogy of Fallot
- Eisenmenger's "Anomaly"

In general, the clinical picture that appears in neonates with DORV is that the baby gets tired quickly, especially when breastfeeding, shortness of breath, pallor, easy cold sweats, leg edema or ascites, central or peripheral cyanosis, clubbing fingers, barriers to growth and development.[13]

Patients with double outlet right ventricle with subaortic interventricular communication and stenosis artery pulmonary (fallot type) have a similar clinical picture with Tetralogy of Fallot patient.[3] The presence of systolic murmur in the neonates associated with the presence of stenosis artery pulmonary.[14] We can found cyanosis in the early months from birth and is progressive and may be accompanied by hypoxic spells.[3] Correction surgery on DORV made by placing an intra-ventricular barrier to direct blood in the left ventricle directly to the aorta and reducing flow obstruction to the right ventricle due to pulmonary stenosis.[15] In some cases with severe cyanosis at the onset of birth, prior shunt surgery may aid repair in subsequent operations.[16] Anesthesia management in patient with EA and DORV remain challenging for anesthesiologist. We present a successful anesthesia management in patient undergoing esophagostomy with DORV.

CASE

A 22-day-old baby 2.9 kg was born spontaneously with the help from a midwife. From the prenatal care history taking, the mother did not have any complaint during her pregnancy and did not have any history of the past illness like hypertension or diabetes mellitus. From the labor history, we know that the baby was born spontaneously with the help of a midwife and was found cyanotic after the baby was born. The midwife refers the patient to the Dr. Saiful Anwar General hospital for advance care.

When the patient arrives at the hospital, we discovered that the babies always drool and vomit when given breastmilk. The physical examination found that patients with patent airway on Continous positive airway pressure (CPAP), Positive end expiratory pressure (PEEP) 7, SpO_2 65-67%, rr 50x / min, rhonchi (+). The circulation obtained cold acral, Capillary refill time (CRT) 2 seconds, pulse 123 x/minute, murmur gr 2/6. Later, this patient was diagnosed with TEF Type C, Ventricular septum defect, Atrial Septum Defect, Double Outlet Right Ventricle, and was planned for an esophagostomy. VACTERL evaluation revealed cardiac abnormalities VSD, Atrial septal defect (ASD), and DORV. In addition, patients were assessed with ASA 4E, Neonate, Resp Failure, moderate VSD, moderate ASD, DORV, TEF type C.

The patient underwent anesthetic management with general anesthesia, intubation with intravenous ketamine 3 mg, fentanyl 3 µg, atracurium 1.5 mg. The operation lasted 3 hours, stable condition, bleeding at least 30 cm^3. During the operation, the patient was given vasopressor norepinephrine at 0.05 µg/ kg/minute. The patient was postoperatively tube-in at the Neonatal intensive care (NICU).

DISCUSSION

Patients with TEF and DORV is a complex problem in anesthesia management. They requires both airway and cardiovascular attention. The first is attention to the airway, especially when doing intubation. Wherever possible, we perform the Rapid sequence induction (RSI) technique in patients with TEF to minimize the risk of aspiration. We choose to give the patient rocuronium for the neuromuscular blocking agent to facilitate the intubation,[17] because we did not have any succinylcholine in our hospital.

In neonatal patients, an induction technique using inhaled gases would generally be chosen, but in these patients, we chose to use an intravenous agent in the hope that we could

balance systemic vascular resistance and pulmonary vascular resistance.[16] The choice of drug to be used is ketamine, taking into account that ketamine has the effect of increasing systemic vascular resistance so that it is expected that the shunt flow to the heart will not change. However actually, ketamine is also has a risk in this kind of patient. We know that ketamine will induce the hypersecretion of the mucous in the respiratory tract, so that we give the pre medication sulfas atropine that has an anti-sialagogue effect for this patient.[18]

To maintain systemic vascular resistance, we also provide a vasopressor, namely norepinephrine, hoping that the dominant alpha 1 activity can maintain systemic vascular resistance and reduce pulmonary vascular resistance, thereby preventing reverse flow.[19] In such patients, our target saturation is according to the basal saturation of the patient, 66-67%. We also prepare the milrinone for this patient if we need to reduce the pulmonary vascular resistance for this patient.[20]

The operation lasted for 3 hours, and we have stable hemodynamic during the operation. The saturation from this patient durante operation is between 61-70%, and the heart rate from this patient is stable between 130 – 140x/minute. After the operation was over, the patient safely returned to NICU ward.

CONCLUSION

TEF is a congenital disorder that requires special management in the induction of anesthesia, namely RSI, which requires fast onset drugs to prevent aspiration. DORV is a congenital heart defect that requires special handling when general anesthesia is performed because most anesthetics have a cardiac depressive effect. Sometimes, we need a vasopressor or inotropic or maintain the balance between systemic vascular resistance and pulmonary vascular resistance. The presence of both of these congenital abnormalities makes induction of anesthesia more difficult and maintenance of anesthesia during the operation so that it requires very close observation to maintain the patient's hemodynamic stability. Anesthesia management with general anesthesia, intubation using intravenous ketamine 3 mg, fentanyl 3µg, atracurium 1.5 mg gives stability for esophagostomy in a patient with a double outlet right ventricle.

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CONFLICT OF INTEREST

None

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