Anesthesia in Emergency PDA Stenting with Pulmonary Arteri Atresia

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Abstract
Pulmonary atresia with an intact ventricular septum is a condition that is characterized by a complete obstruction to right ventricular outflow. This condition is fatal if untreated. Ductus arteriosus communicate between pulmonary artery and proximal descending aorta. It is essential in fetal life as a communicating shunt to send blood from right ventricle bypassing the underdeveloped lungs. Ductal patency may be lifesaving in few cyanotic congenital heart diseases to maintain pulmonary blood flow. Case Illustration: We present a case of a 1-day-old, 3405 g child who presented to casualty with bluish discoloration. The child had central cyanosis, tachypnea with saturation 80% on 1 L/minute nasal cannula and desaturate to 40%. Transthoracic echocardiography (TTE) revealed intact intraventricular septum with pulmonary atresia and patent ductus arteriosus (PDA). The patient scheduled to an emergency PDA stenting. A heating blanket is installed on the operation table, oxygen saturation with Pulse-oxymeter, noninvasive blood pressure, end-tidal CO2 monitor, and temperature monitoring were done. Anesthesia induction was performed with 50% oxygen in air, fentanyl 3 µg/kg, ketamine 2 mg/kg, atracurium 0.5 mg/kg and maintained with sevoflurane. The procedure was performed about 20 to 30 minutes. After procedure, oxygen saturation was increased up to 60%. The child was successfully extubated after 24 hours in the Neonatal Intensive Care Unit (NICU) with saturation 92%. Conclusion: PDA stenting is a less invasive palliative procedure for ductal dependent complex congenital heart diseases. Anesthesiologist's role were maintaining arterial saturation and hemodynamics, securing vascular assess, maintaining temperature. It needs thorough understanding of underlying physiology.

Keywords: Pulmonary atresia, PDA stenting, Anesthesia, Emergency

Introduction
Pulmonary Atresia with intact ventricular septum (PAIVS) is a rare congenital defect that consists of atresia of the pulmonary valve resulting in an absent connection between the right ventricular outflow tract and the pulmonary arteries, and an intact ventricular septum that allows no connection between the right and left ventricles (Nanditha, Kapoor, & Sarin, 2017);(Edwards P, 1973);(Proud, 2008). It is to be considered that PAIVS is a distinct defect compared to pulmonary atresia in ventricular septal defect and Ebstein anomaly (Koehler et al., 2020);(Lai & Yu, 2021). It is different that PAIVS features developmental abnormalities of the right ventricle with the tricuspid valve that is upstream of the pulmonary outflow (Yonas, Pranata, & Nusarintowati, 2017).
The pulmonary arteries in PAIVS are small, but the architecture and branching are normal. While in pulmonary atresia in ventricular septal defect, the right ventricle is normally-formed and the relation between the tricuspid valve and the pulmonary artery is inverted in that downstream abnormalities of the pulmonary arteries are present, compared with upstream abnormalities of PAIVS (Yonas et al., 2017);(Snow et al., 2020). The reported incidence of this condition is based on registries and population-based studies of children born with congenital heart disease are approximately four to eight per 100,000 live births (Glaziou, Falzon, Floyd, & Raviglione, 2013). PAIVS itself accounts for 1 to 3 percent of all congenital cardiac defects in children regardless of gender (Bahaidarah et al., 2021).

Method Research

This research method will use a retrospective descriptive approach to evaluate the management of anesthesia in patients undergoing emergency ductus arteriosus (PDA) stenting procedures with pulmonary arterial atresia conditions. Data will be collected from the medical records of patients who have undergone this procedure in the hospital over the past five years. The variables to be evaluated include patient demographics, preoperative clinical conditions, anesthesia techniques used, anesthesia drugs administered, perioperative complications, and postoperative clinical outcomes. Data analysis will be performed using descriptive statistics to describe patient profiles and the most commonly used anesthesia techniques, as well as bivariate analysis to identify factors associated with better clinical outcomes or perioperative complications. This study is expected to provide practical guidance in the management of anesthesia for emergency PDA stenting procedures in patients with pulmonary artery atresia.

Result and Discussion

A 1-day-old baby boy patient was brought to Sarjito Hospital due to complaints of blue body and desaturation. The patient was born to P1A0 mother, 39 weeks of gestation vaginally, assisted by a midwife. After birth, the patient immediately started crying and after 30 minutes he got blue all over his body, from the examination it was found that the saturation was only 80%. In the ER at Sarjito Hospital, the patient underwent a physical examination and supporting examinations. Obtained RR 40x/minute, with a saturation of 68%, No abnormal breath sounds and heart murmurs were found. And the investigations obtained normal results. The patient was also subjected to an imaging examination in the form of an x-ray thorax, and no abnormalities were found in the imaging results. The patient was sent to the NICU for observation. The patient's saturation decreased to 56% and echocardiography was performed. Pulmonary atresia was found with an intact ventricular septum and dependent patent ductus arteriosus on echocardiography examination. The patient was suggested to undergo emergency PDA stenting immediately.

Patients were assessed for ASA physical status 4 with desaturated PDA dependent. The patient was fasted and an intravenous line was placed and it was flowing
smoothly with iv cath 24G and maintenance fluid was installed with D5 1/2 NS. Oxygenation of the patient was maintained with a nasal cannula at 1 liter/minute at a saturation of 30%-40% before being transferred to the cathlab. In the cathlab room, a heating blanket is installed on the table.

Pulse-oxymeter oxygen saturation, electrocardiogram, noninvasive blood pressure, and temperature monitoring were performed. Anesthesia was induced with 50% oxygen, fentanyl 3 mcg/kg, ketamine 2 mg/kg, atracurium 0.5 mg/kg and anesthesia was maintained with sevoflurane with 30% oxygen. The maintenance D51/2% infusion was continued throughout the procedure. The patient was intubated with a 3.5 uncuff ETT during the procedure. The 5 F sheath is secured in the femoral vein and the 4 F sheath is secured in the femoral artery. 0.5 mg/kg heparin is given and 3 stents were inserted into the PDA and confirmed by transthoracic echo. With TTE, correct stent placement and continued flow across the PDA is seen. The procedure takes approximately 1 hour. During the procedure, the child was stable, and the saturation increased from 40% to 68%. Nothing was discontinued but intubation was maintained and the patient was sent back to the NICU for observation. In the NICU the patient had an improvement in saturation to 92%, observations were stopped and extubated after 24 hours.

Pulmonary atresia with the intact ventricular septum (PAIVS) is a channel-dependent cyanotic congenital heart disease. As deoxygenated blood flows through the inferior and superior vena cava into the right atrium, it does not pass through the tricuspid valve into the right ventricle; instead, most of the blood volume is diverted through a patent foramen ovale (PFO) or atrial septal defect (ASD) into the left atrium, where it enters the systemic circulation. Deoxygenated blood is pushed away from the pulmonary circulation because of the high elevation of right ventricular pressure, which results from the atretic pulmonary valve. To oxidize blood, the patent ductus arteriosus (PDA) is essential, connecting systemic oxygenation to the lungs. Patients with PAIVS cyanosis at birth. Initial palliative care includes PGE1 infusion to keep the ductus arteriosus open, pulmonary valvotomy, and/or a modified Blalock-Taussig shunt (Yoshimura & Yamaguchi, 2009).

As cardiac catheterization techniques improve, PDA insertion and balloon valvotomy appear to be effective and minimally invasive alternatives. PDA stenting has even been shown to be superior for promoting even pulmonary vascular growth, compared to the modified Blalock-Taussig shunt. Therefore, it is wise for the anesthesiologist to become familiar with the procedure and its possible complications when treating these patients perioperatively (Bentham et al., 2018). Periprocedural risk is significantly increased. The overall periprocedural morbidity and mortality is approximately 7%. Patients younger than 30 days of age had an odds ratio of 1.43 compared to those older than 30 days. Hemodynamic susceptibility with systemic saturation <95% adds additional risk, compared with those without hemodynamic markers (Odegard et al., 2016).

Thus, cardiac catheterization laboratory anesthesiologists should be more vigilant when treating similar pediatric patients. Periprocedural management centers around a
balance between systemic blood flow to the lungs as well as functional RV support. Acidosis, hypoxemia, hypercapnia, and vasoconstrictive drugs, overload an already vulnerable cardiovascular system (Odegard et al., 2016). Conscious sedation carries the risk of airway obstruction but avoids the need for tracheal intubation, potentially resulting in faster recovery. However, intubation is still preferred because of its advantages in maintaining the airway in patients at risk of regurgitation of gastric contents, such as emergency patients with inadequate fasting (Tierney, Kenny, & Greaney, 2022).

PDA stent detachment is a rare complication. In the past literature, there have only been sporadic case reports involving stent migration PDA. Michel-Behnke I. and colleagues reported a dislocation to the left main pulmonary artery followed by successful resuscitation6. Hussain A, et al found two dislodged to the right pulmonary artery, and Schranz D, et al reported one; all three were converted to surgical aortopulmonary shunts7. A blocked pulmonary trunk mimics an acute pulmonary embolism and requires immediate cardiopulmonary resuscitation (Tierney et al., 2022).

Anesthesia for PDA stenting poses a unique challenge. Goals include maintaining adequate preload and contractility. The decrease in systemic vascular resistance and the increase in pulmonary vascular resistance (PVR) can be eliminated as this will lead to a further decrease in pulmonary blood flow and hypoxia. Factors that cause an increase in PVR such as hypoxia, hypercarbia, and acidosis can cause a sudden decrease in clinical condition. Other challenges include hypoglycemia and hypothermia (Ruetzler & Kurz, 2018). Warm blankets and warm saline for rinsing and fluid warmers for intravenous infusion were used in our case. Adequate hydration is necessary to reduce viscosity. The role of the cardiac anesthesiologist also extends to transthoracic imaging. Confirmation of the diagnosis, correct placement of the stent throughout the PDA, adequate flow across the stent, and complications such as stent migration should be looked for when performing TTE. In addition to these complications, reported complications such as acute stent thrombosis, pre-stent duct spasm, and pulmonary congestion should also be kept in mind (Flow, 2013).

**Conclusion**

Pediatric patients with PA/IVS are at increased perioperative risk associated with interventional cardiac catheterization procedures. Anesthesiologists should acquaint themselves with the unique pathophysiological changes and stratify the periprocedural management tethered to each patient. PDA stenting is an attractive, less invasive, palliative procedure for duct-dependent complex congenital heart diseases where the definitive procedure is to be done at slightly later date. Anesthesiologist's role lies in maintaining arterial saturation, hemodynamics, securing vascular assess, maintaining temperature, and thorough understanding of underlying physiology holds the key.
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