

CASE REPORT

Perioperative Management of Patients with Ventricular Septal Defect, Severe Tricuspid Regurgitation and Gerbode Defects

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ABSTRACT

Ventricular Septal Defect (VSD) is a congenital heart disease that causes the connection between left and right ventricles called a Gerbode defect. Manifestation of a Gerbode defect is damage to the opening tricuspid valve caused regurgitation of the tricuspid valve. Delay in diagnosis and intervention will affect pre-operative nutritional status and malnutrition.

We reported a boy aged 2 months, weighing 3100 grams with biliary atresia followed by VSD, severe TR, and Gerbode defect who will undergo the Kasai procedure. Preoperative physical examination showed GCS E4V5M6, SpO₂ 100%. The skin gets icteric all over the body and conjunctiva. The cardiovascular system has a regular I-II heart sound, 2/3 mid clavicular S noise as high as 2 ICS and a pansystolic murmur. The examination of the abdomen is slight distended. Child pug score 8. Hemoglobin value 6.7gr%, hematocrite 37%, APTT 44.8 seconds, SGOT 443 U / L, SGPT 560 U / L, total bilirubin 23.89 mg / dl, direct bilirubin 13.92 mg / dl, and indirect bilirubin 9.97 mg / dl.

The goal of anesthesia in VSD, Severe Tricuspid Regurgitation (TR) with Gerbode Defect is preventing excessive ventilation to avoid severe pulmonary hypertension. The choice of anesthetic agent is based on the patient's physiology and balancing pulmonary and systemic blood flow. Perioperative management of cases of VSD, TR Severe with Gerbode defect in the following report describes the importance of understanding the pathophysiology of VSD and Gerbode defects to obtain a good outcome.

Perioperative management of VSD patients, severe tricuspid regurgitation with Gerbode defect requires more supervision, especially to minimize the increase in PVR, maintain systemic vascular resistance (SVR) and avoid excessive ventilation to prevent severe pulmonary hypertension.

Keywords: ventricular septal defect; tricuspid regurgitation severe and gerbode defects

INTRODUCTION

Congenital heart disease (CHD) is an abnormal condition in the heart or large intrathoracic blood vessels that is acquired from infancy. CHD is at the top of the list of diseases in infants. Cyanotic CHD cases are known to be higher than the asyanotic CHD. In America, 1% of babies out of 40,000 live births suffer from CHD. CHD that is not detected early and handled properly will result in death in the first month of life (33-50%).^{1,2}

In developed countries, CHD can be detected at less than 1 month of age, whereas in developing countries like Indonesia, the majority of CHD cases are detected when the patient's condition begins to deteriorate. However, most 30% of babies with CHD show symptoms in the early weeks of life. One type of CHD is a ventricular septal defect (VSD) which occupies 20-30% of all CHD.^{2,4}

The type of ventricular septal defect (VSD) that causes the connection between the left and right ventricles to appear as a direct or indirect type is called a Gerbode defect. This defect occurs in less than 1% of CHD cases. One of the manifestations of a Gerbode defect is damage to the tricuspid valve gap causing regurgitation of the tricuspid valve. Delays in diagnosis and intervention in CHD cases will affect nutritional status which will lead to pre-operative malnutrition.^{3,5}

CASE ILLUSTRATION

Anamnesis

A boy aged 2 months with a body weight of 3100 grams has complaints of jaundice since birth, the longer it gets yellow to the eye area. Patients sometimes have a fever and then the fever goes down by itself. The stool is greenish yellow. Pee brown like tea. Complaints are not accompanied by a history of cough, runny nose, nausea and vomiting. The patient had a history of surgery with General Anesthesia (GA).

Physical Examination

From the physical examination, it was found GCS E4V5M6, other vital signs were within normal limits. Examination of the skin looks icteric all over the body, conjunctiva is icteric. In the cardiovascular system there is a regular I-II heart sound, 2/3 mid clavicle S sounds as high as ICS 2 and a pansystolic murmur is obtained. Abdominal examination found slight distended, supple, no tenderness. There is no palpable enlargement of the liver and spleen. Child pug score shows a value of 8 (moderate severe liver disease) with a risk of death of 10%.

Supporting Examination

Laboratory examination showed the value of Hemoglobin 6.7gr%, 37% hematocrit, leukocytes 7,800 / ul, PT 12.7 seconds, APTT 44.8 seconds, SGOT 443 U / L, SGPT 560 U / L, total bilirubin 23.89 mg / dl, direct bilirubin

13.92 mg / dl, and indirect bilirubin 9.97 mg / dl. Echocardiography shows VSD 2.5 cm diameter, TR severe, Gerbode Shunt. Thorax X-ray shows pneumonia and an enlarged heart with Left Ventricular Hypertrophy (LVH) configuration. The cystography shows a contrast filling of the cystic ducts, hepatic ducts, communis and right and left hepatic ducts. USG Abdomen shows cholestasis picture possibly due to biliary atresia, the structure of the other abdominal organs appear normal.



Figure 2. Cystography shows a contrast mingition of the cystic duct



Figure 1. Ultrasound Abdomen shows Cholestasis



Figure 3. Thorax X-ray shows pneumonia and LVH

Anesthesia Management

After obtaining consent from the family and there is no contraindication for the procedure, preoperative management while the patient is fasting ensure that intravenous fluids are sufficient with D5 ¼ NS 12.4 cc / hour by infusion pump. Furthermore, the patients received intravenous premedication

Midazolam 0.15 mg / kg and Ketamine 1 mg / kg before being separated from their families. In the operating room, a standard monitor was installed according to the ASA recommendations. Obtained the condition of the patient with a blood pressure of 65/35 mmHg, pulse rate 108 x / minute and SpO2 100%.

Prior to induction, the patient was given 10 cc crystalloid solution of D51/4NS. Analgesia Fentanyl 1 mcg / kg intravenously, atracurium 0.5 mg/kg, atropine sulfas 0.1 mg, induction with addition of Ketamine 1 mg / kg and Sevofluran 2 volume % in FiO2 60% versus airbar with hyperventilation. In addition, dexamethasone was given 0.1 mg / kg, ondancetrone 0.1 mg / kg. Atracurium 0.1 mg / kg intravenously was given as a muscle relaxant before the endotracheal tube insertion no. 3.0 oral non kinking. A foley catheter is also placed to monitor urine production.

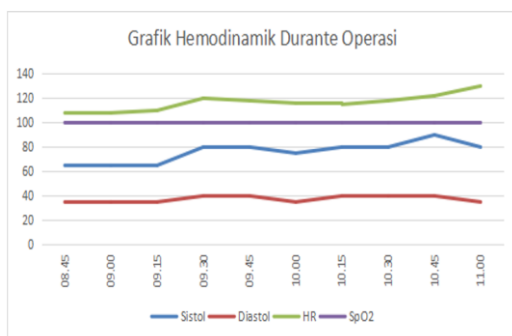


Figure 4. Hemodynamic Graph of Operation Durante

Anesthesia maintained with control ventilation of 2.5 volume % Sevoflurane in

50% oxygen versus airbar, Fentanyl 1 mcg / kg / hr intravenously continuous, D5 1/4NS fluid during surgery. The patient was stable during the operation which lasted for 2 hours with a pulse rate ranging from 110-120 x / minute, systolic in the range 60-80 mmHg, 100% SpO2, temperature 36,4°C. The total fluid intake during surgery is 50cc. Bleeding amounting to 10 cc. After confirming that the muscle relaxant effect wears off, the patient is extubated in a fully conscious state for 3 hours in the PICU room.

Management of postoperative

Management of postoperative analgesia of ketamine 0,12 mcg / kg / hour for 8 hours postoperatively. On the third day the patient was transferred to the ward and allowed to be outpatient on the 5th day.



Figure 5. Postoperative 3rd Day

DISCUSSION

Anesthesia management in VSD patients requires caution, because too deep sedation can cause hypercarbia and ultimately increase pulmonary vascular pressure (pulmonary hypertension). Pulmonary hypertension is defined as a sudden increase in PVR that causes pulmonary pressure to exceed mean arterial pressure, interferes with cardiac output and causes hypoxemia. Patients who have defects of the pulmonary and systemic circulation (VSD) need to pay attention after intubation not to excessive ventilation.^{6,7}

Low levels of carbon dioxide in the blood due to excessive ventilation will decrease PVR and result in increased blood flow to the pulmonary via the shunt, resulting in reduced systemic blood flow. A balance between the systemic and pulmonary circulation can be achieved by providing minimal oxygen concentrations and maintaining normal carbon dioxide levels. PEEP 5-10 cmH₂O can help limit excessive pulmonary blood flow by increasing the mean airway pressure (MAP).^{7,8,9}

Common problems that can be encountered in general anesthesia (GA) procedures are heart block, dysrhythmias, heart failure and infective endocarditis. Shunting has an important influence on anesthetic management. Inadequate anesthesia and stimulation of the sympathetic nervous system

can increase SVR, worsen left-to-right shunts and reduce systemic cardiac output in patients with large ventricular septal defects. GA provides better hemodynamic stability intraoperatively and postoperatively. Hypercarbia can be avoided by mechanical ventilation and adequate adjustment of tidal volume and respiratory rate. Inflation of the lungs with intermittent positive pressure ventilation (IPPV) causes the release of endogenous nitric oxide and prostaglandins leading to pulmonary vasodilation.^{7,10}

The main objectives during GA are to minimize the increase in PVR, maintain systemic vascular resistance (SVR), maintain optimal cardiac contractility along with close monitoring to detect arrhythmias in which these changes can lead to hypoxemia, cyanosis, and myocardial ischemia. Anesthetics for tricuspid regurgitation are designed to maintain cardiac output by reducing systemic and pulmonary vascular resistance to aortic and pulmonary outflow, and by maintaining careful venous return. Increased heart rate can help increase the output of mitral regurgitation, so bradycardia should be avoided. Isoflurane, halothane, or opioid anesthetics may be recommended.^{10,11}

Anesthetic management in VSD includes optimizing preload, avoiding hypovolemia, keeping SVR normal and preventing an acute increase in PVR. Given the

possibility of right ventricular dysfunction due to severe PH, inotropic supports such as adrenaline and milrinone are used during weaning on cardiopulmonary bypass.^{11,12}

Gerbode defect in VSD requires special supervision, especially in the use of anesthesia. Gerbode defect still remains after Kasai procedure. Anesthesia in a patient with Gerbode defect involves preventing altered shunt flow and maintaining cardiovascular stability and adequate breathing so as to maintain tissue perfusion and oxygenation. Increased vascular resistance in the lungs must be prevented to avoid airway obstruction, hypoxia, hypoventilation, acidosis and hypothermia. On the other hand, a significant reduction in pulmonary vascular resistance will increase shunt flow and should be avoided. The choice of anesthetic agent should be based on the patient's physiology and the goal of balancing pulmonary and systemic blood flow. Ketamine exerts beneficial effects in children with CHD and severe pulmonary hypertension by maintaining SVR and ventricular performance, without increasing pulmonary vascular resistance.^{12,13}

CONCLUSION

Perioperative management of VSD patients, severe tricuspid regurgitation with Gerbode defect requires more supervision, especially to minimize the increase in PVR,

maintain systemic vascular resistance (SVR) and avoid excessive ventilation to prevent severe pulmonary hypertension.

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