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Epidural Anesthesia with Eissenmenger Syndrome Undergoing Caesarean Section : a Case Report

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ABSTRACT

Introduction: Heart disease in pregnant women can cause both morbidity and mortality. Eissenmenger syndrome is a pulmonary hypertension due to high pulmonary vascular resistance with a right-to-left or bidirectional shunt at the aortopulmonary, ventricular, or atrial level. To prevent hemodynamic instability and hypoxemia, it is essential to maintain a balance between PVR and SVR.

Case Report: We reported the case of a 28-year-old pregnant woman with a gestational age of 34-35 weeks, diagnosed with Eissenmenger syndrome due to shortness of breath and cyanosis in the extremities, classified as ASA III E. After epidural anesthesia was administered, a 1.5 hour cesarean section was performed and a male baby was born. Postoperatively, the patient was transferred to the ICU.

Conclusion: This case highlights the importance of maintaining a balance SVR and PVR, and continuous monitoring in pregnancy patient complicated by Eissenmenger syndrome. It requires a multidisciplinary approach involving obstetricians, anesthesiologists, and pediatricians.

Keywords

Eissenmenger Syndrome, Epidural Anesthesia, Pregnancy.

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INTRODUCTION

Heart disease in pregnant women can cause morbidity and even mortality. In the United States between 1991 and 1993, maternal mortality rates due to heart disease increased significantly. The incidence in Indonesia is also rising, as valve disorders caused by rheumatic fever remain high.[1,2]

Valve disorders in Indonesian women are often only detected during the first pregnancy, particularly when the hemodynamic burden increases at the end of the second trimester.[2] Heart disease in pregnant women, especially congenital heart disease during pregnancy, may involve abnormalities in the heart walls, leading to disruptions in blood flow.[1]

Victor Eissenmenger first described the "Eissenmenger complex" in 1897 in an article on congenital ventricular defects. In 1958, Wood redefined this syndrome as "pulmonary hypertension due to high pulmonary vascular resistance with a right-to-left or bidirectional shunt at the aortopulmonary, ventricular, or atrial level.[3] Patients with Eissenmenger syndrome can survive into adulthood, with a survival rate as high as 80% 10 years after diagnosis and 42% at 25 years. Maternal mortality in patients with Eissenmenger syndrome is very high, ranging from 23% to 50%. In this case, a 28-year-old pregnant woman, with a gestational age of 34-35 weeks, was brought to the hospital with complaints of shortness of breath and cyanosis in the extremities, with a physical status classified as ASA III E. Oxygen demand during pregnancy increases, potentially causing hypoxemia in both the mother and the fetus. Therefore, maintaining a balance

between pulmonary vascular resistance and systemic vascular resistance is essential to avoid hemodynamic changes that could lead to hypoxemia through a right-to-left shunt.[4,5]

Regional anesthesia techniques offer advantages in maintaining hemodynamics in such cases. In this patient, a cesarean section was performed using epidural regional anesthesia, with an operation lasting 1.5 hour, during which there were no hemodynamic changes. Postoperatively, the patient was transferred to the Intensive Care Unit (ICU). The primary goal of anesthesia in pregnant patients with congenital heart disease is to maintain stable hemodynamics in both the mother and the fetus. Here, we report a case of we report a case of management of labor in a pregnant patient with Eissenmenger syndrome.

CASE

A 28-year-old pregnant woman was referred to the emergency operating room at 34-35 weeks of gestation, G2P1A0. The patient complained of shortness of breath two days prior to hospital admission, accompanied by shallow breathing, cyanosis in both fingers, and the need to sleep with three pillows. She had an irregular history of prenatal checkups and had never monitored her blood pressure, with no history of systemic diseases such as diabetes mellitus. The patient was diagnosed with a ventricular septal defect (VSD) five years ago along with pulmonary hypertension. However, due to socioeconomic constraints, she did not regularly follow up for treatment at the hospital. Her obstetric history includes her first pregnancy in 2014, which was a full-term normal delivery assisted by a midwife, delivering a male infant weighing 2900 grams. This current pregnancy is her second.

On physical examination, the patient had Glasgow Coma Scale (GCS) score of 15 (E3M5V5), blood pressure 113/72 mmHg, pulse 104 beats/min regular rhythm, respiratory rate 24-26 breaths/min, SpO2 72% with non-rebreathing mask (NRM) at 10 L/min, and temperature 36.5°C. The Mallampati score was class I, with mouth opening of more than three fingers. Cardiac auscultation revealed a pansystolic murmur (+). The fundal height was 30 cm, corresponding to the gestational age, and the fetal heart rate was 135-140-144 beats/min. Cyanosis was observed in the extremities. Diuresis was 100 cc/hour, with clear yellow urine. Initial laboratory results showed hemoglobin (Hb) levels of 10.9 g/dL, hematocrit 32.7%, leukocytes 14,900/µL, sodium 140 mmol/L, potassium 4.0 mmol/L, calcium 4.45 mmol/dL, magnesium 1.8 mmol/dL, urea 17 mg/dL, creatinine 0.92 U/L, urine protein +3, prothrombin time (PT) 9.2 seconds, activated partial thromboplastin time (APTT) 22.1 seconds, aspartate aminotransferase (AST) 15 U/L, alanine aminotransferase (ALT) U/L, and random blood glucose 105 mg/dL.

The electrocardiogram (ECG) showed sinus tachycardia with a heart rate of 115 beats/min, regular rhythm, right axis deviation, and incomplete right bundle branch block (RBBB). The echocardiograph showed a 17 mm perimembranous VSD, a bidirectional shunt, severe pulmonary artery hypertension, severe tricuspid regurgitation, mitral regurgitation, a dilated right atrium, and normal biventricular function with an ejection fraction of 52%. The patient was then referred to the Anesthesiology Department as a G2P1A0 at 34-35 weeks with Eissenmenger syndrome. The assessment of the patient revealed American Society of Anesthesiologists (ASA) classification of III E. The patient was administered intravenous furosemide 20 mg/hour, dorner 20 mg twice daily and sildenafil 25 mg three times daily orally. 10 L/min oxygen was given in a semi-sitting position. The patient was instructed to fast before entering the operating room. Packed red cells (PRC) were prepared, and regional epidural anesthesia was planned, with postoperative care to be conducted in the ICU.

Pre-anesthetic evaluation must be performed carefully by considering both physical and laboratory examinations to determine the choice of anesthesia method. Monitoring of the fetus and the mother's vital signs includes blood pressure, fluid intake and output, cervical dilation, and frequency of uterine contractions. The Obstetrics and Gynecology Department referred the patient to Cardiology Departement due to an accompanying heart condition. A comprehensive examination was conducted by the Cardiology Department, including additional tests such as ECG and echocardiography. Oxygen administration of 6-10 liters per minute was provided for the oxygenation of the mother and fetus, along with 500 cc of Ringer's lactate fluid over 24 hours. A Foley catheter was inserted, and the patient was positioned semi-upright.

Continuous furosemide was administered at a dose of 10-20 mg per hour with a target urine output of 2 cc/kg body weight to reduce the cardiac load. Additionally, explanations were given to the patient and her family regarding the anesthesia procedure, fasting before surgery, and the postoperative care unit.

Preparation in the operating room included checking the anesthesia machine with its gas flow system and monitoring system, preparing STATICS, preparing medications and anesthesia equipment to be used, as well as resuscitation drugs and equipment. Additionally, a regional set for epidural anesthesia was also prepared. Once the patient arrived in the operating room, sphygmomanometer, ECG, and pulse oximeter were applied, and the anesthesia medical record card was prepared. During patient preparation, oxygen was administered at 6-10 L/min via a non-rebreathing mask, the intravenous line was checked to ensure it running smoothly, and the patient was then transferred to the gurney and operating table.

Epidural anesthesia was administered in a seated position, with the puncture at the L3-4 level, and 14 cc of 0.5% isobaric bupivacaine was given incrementally. After the epidural anesthesia, the patient was placed in a 45° head-up position. Blood pressure measurements and a test dose were performed to ensure that the epidural was not within a blood vessel, and a block level of T6 was achieved. The patient's vital signs during surgery showed systolic blood pressure of 110-120 mmHg, diastolic blood pressure of 76-81 mmHg, pulse 93-105 beats/min regular rhythm, and SpO2 82-84%. The surgery lasted 1.5 hours, with 500 cc of intraoperative bleeding and a urine output of 50 cc/hour. A male baby was born with an APGAR score of 7/9. When the baby and placenta are delivered, an additional 40 mg of intravenous furosemide is administered due to the autotransfusion that occurs at that time, which could increase the strain on the mother's heart. Afterward, the patient is transferred to the recovery room and given postoperative analgesics via epidural bupivacaine 0.125% and fentanyl 2 mcg/cc at a dose of 2.5 cc/hour (25 mcg/hour), along with 500 cc of Ringer's lactate over 24 hours.

During the recovery room stay, the patient was conscious and alert (compos mentis). The patient's vital signs showed a systolic blood pressure 112-115 mmHg, diastolic blood pressure 78-82 mmHg, pulse rate 100-107 beats/min regular rhythm, respiratory rate 22-24 breaths/min, and SpO2 of 82-84% with NRM at 10 L/min. The patient was given postoperative analgesia with epidural bupivacaine 0.125% and fentanyl 2 mcg/cc at a dose of 25 mcg/hour. The patient was transferred to the ICU once the ALDRETTE score was above 8. There were no signs of shortness of breath, increased pulse rate, or pain complaints, so the patient was moved to the ICU. While in the ICU, the patient was still compos mentis, with systolic blood pressure 118-121 mmHg, diastolic blood pressure 69-78 mmHg, pulse rate 92-108 beats/min regular rhythm, respiratory rate 22-24 breaths/min, and SpO2 of 82-84% with NRM at 10 L/min.

DISCUSSION

Eissenmenger syndrome is characterized by an uncorrected chronic left-to-right shunt, leading to left ventricular hypertrophy, increased pulmonary artery pressure, and right ventricular dysfunction. Left-to-right intracardiac shunting increases flow and pressure through the pulmonary vasculature, resulting in pulmonary vascular remodeling and eventually pulmonary vascular disease. Shunting at the level of the great arteries causes an increase in pulmonary artery blood flow and pulmonary venous return to the left atrium, leading to an increase in left ventricular end-diastolic volume and left ventricular stroke work, in line with the Frank-Starling mechanism. Shunting at the great artery level also decreases diastolic blood pressure due to blood flow into the low-pressure pulmonary circuit after aortic valve closure.[2,3]

Significant changes in maternal physiology are critical considerations when performing anesthesia procedures. The goal of anesthesia in Eissenmenger syndrome is to avoid hemodynamic changes that could worsen hypoxemia by increasing the right-to-left shunt. Cardiac output must be maintained, and systemic vascular resistance (SVR) should not decrease. Factors that reduce cardiac output include direct myocardial depression, loss of sympathetic stimulation to the heart, extreme changes in heart rate, and decreased venous return.[4,5] Additionally, maneuvers that increase pulmonary vascular resistance (PVR) must be avoided. In this patient, oxygen was administered to prevent an increase in PVR and to avoid hypoxemia. Dorner was given to manage the patient's pulmonary hypertension, while furosemide was used to reduce the cardiac

workload. The use of a Foley catheter was necessary to monitor fluid output and evaluate fluid intake and output.

Based on the physical examination and laboratory results, we chose regional epidural anesthesia. The risks associated with this include hypotension, radiculopathy, abscess, hematoma, back pain, dizziness, and neurological deficits. Factors that influence the effectiveness of epidural anesthesia include the type of local anesthetic used, age, gender, weight, the patient's physical condition, and their position during the procedure. For this patient, the epidural was performed in a seated position while the patient held a pillow. Blood pressure was measured beforehand to establish a baseline. After aseptic measures were applied, the puncture was made at the L3-L4 level using an epidural needle, and 15 cc of 0.5% bupivacaine was administered incrementally. The patient was then placed in a supine position. Blood pressure was measured again, and a test dose was administered to confirm whether the catheter had entered the epidural space or a blood vessel. In this case, the catheter entered the epidural space, indicated by the absence of an increase in heart rate and blood pressure.

The block height reached T6, which is preferable to T4, as T4 can cause bradycardia due to sympathetic block of the cardioaccelerator nerve fibers, which is particularly risky for high-risk patients.[3] Hemodynamic and respiratory changes are typically minimal with well-managed epidural anesthesia. The administration of oxytocin in bolus form at high doses in patients with heart disease can cause vasodilation and a decrease in SVR, compensatory tachycardia, and increased cardiac stress. Slow infusion of diluted oxytocin is generally well tolerated. Other uterotonic agents, such as ergometrine, can cause systemic hypertension and coronary vasoconstriction. Prostaglandin F2-alpha has the potential to cause severe pulmonary hypertension if large bolus doses are injected directly into the circulation.[5]

One factor that must be considered intraoperatively is placental blood flow, and any factors that reduce uterine blood flow must be avoided. The formula for calculating uterine blood flow is:

$$ubf = \frac{uap - uvp}{uvr}$$

ubf = Uterine Blood Flow
uap = Uterine Arterial Pressure
uvp = Uterine Venous Pressure
uvr = Uterine Vascular Resistance

Factors That Decrease UAP
Supine Position (Compression Of The Vena Cava)
Hemorrhage/Hypovolemia
Hypotension
Hypotension During Sympathetic Blockade
Factors that decrease UVP
Supine Position (Compression Of The Vena Cava)
Uterine Contractions
Drugs Causing Uterine Hypertonus (Oxytocin, Local Anesthetics)
Skeletal Muscle Hypertonus (Seizures, Valsalva Maneuver)
Factors that increase UVR
Endogenous Vasoconstrictors (Catecholamines, Vasopressin)
Exogenous Vasoconstrictors (Epinephrine, Ephedrine)
Local Anesthetics (In High Doses)

Eissenmenger syndrome is a complex pathophysiological condition characterized by cyanosis, communication between the right and left circulatory systems (such as an atrial septal defect/ASD, ventricular septal defect/VSD, or an aorticopulmonary anomaly) that allows bidirectional shunting, and pulmonary hypertension caused by irreversible increases in PVR.

Shunting at the level of the great arteries leads to an increased pulmonary artery blood flow and pulmonary venous return to the left atrium, which increases left ventricular end-diastolic volume and left ventricular stroke work, following the Frank-Starling mechanism. Shunting at the great artery level also results in decreased diastolic blood pressure due to blood flow into the low-pressure pulmonary circuit after the aortic valve closes. Low diastolic pressure reduces coronary perfusion, potentially causing ischemia due to an imbalance between reduced myocardial oxygen delivery and increased oxygen demand. As a result of this volume overload, the left ventricle eventually dilates and hypertrophies, increasing left ventricular end-diastolic pressure, which is followed by increased left atrial pressure. The final outcome is pulmonary edema due to pulmonary venous congestion and left heart failure.[3-5]

The increase in PVR raises right ventricular pressure, ultimately leading to right heart failure. Shunting at the atrium or ventricle level, if large, increases right ventricular volume, in addition to the hemodynamic effects caused by shunting at the great artery level. Prolonged exposure of the pulmonary vasculature to increased flow and pressure results in fixed elevated PVR. When PVR exceeds SVR, the shunt reverses, leading to cyanosis and erythrocytosis, known as Eissenmenger syndrome.[5]

CONCLUSION

Significant changes in maternal physiology are key considerations when performing anesthesia procedures. Comorbid conditions during pregnancy make both general and regional anesthesia techniques a unique challenge. Heart conditions during pregnancy increase the risk of morbidity and mortality for both the mother and the fetus.

This situation requires a multidisciplinary approach involving obstetricians, anesthesiologists, and pediatricians. Anesthesia management for pregnancy complicated by Eissenmenger syndrome often presents challenges. However, the most important principle is maintaining a balance SVR and PVR. Both general and regional anesthesia techniques must ensure that cardiac output and SVR are maintained, and increases in PVR are avoided. Continuous monitoring is essential not only in the preoperative and intraoperative phases but also in the postoperative period, as postoperative complications can significantly increase the risk of maternal mortality.

DECLARATIONS

Ethics approval and consent to participate. Permission for this study was obtained from the Ethics Committee of Bedas Cimaung General Hospital Bandung.

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The Authors agree to publication in Journal of Society Medicine.

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All authors significantly contribute to the work reported execution, acquisition of data, analysis, and interpretation, or in all these areas. Contribute to drafting, revising, or critically reviewing the article. Approved the final version to be published, agreed on the journal to be submitted, and agreed to be accountable for all aspects of the work.

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