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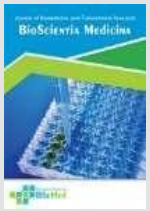

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Anaesthetic Management for Caesarian Section Complicated with Eisenmenger Syndrome Concomitant Severe Preeclampsia: A Case Report

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A B S T R A C T

Background: Although conception is discouraged in women with Eisenmenger syndrome, in inevitable circumstances, careful and meticulous planning of anesthesia can help the parturient survive the ordeal of a cesarean section. Anaesthetic management is aimed at avoiding haemodynamic changes that increase right-to-left shunt, thereby worsening hypoxemia. This study aimed to present a case of a pregnant woman with Eisenmenger syndrome who underwent caesarean section under general anesthesia management with severe preeclampsia. **Case presentation:** A 42-year-old pregnant woman (gravida 3, para 1, abortion 1) was admitted to our hospital at 38 weeks gestation with the chief complaint of shortness of breath and severe preeclampsia. Physical examination revealed cyanotic with clubbing fingers and elevated jugular venous pressure. Laboratory investigations did not reveal any abnormalities. Electrocardiography (ECG) shows normal sinus rhythm and right atrial deviation. General anesthesia was chosen due to obstetric complications, namely severe preeclampsia. The patient was premedicated with a gastroprotective agent, ranitidine 150 mg. At follow-up anesthesia 36 hours after surgery, the patient scored for maximum perioperative comfort and satisfaction. **Conclusion:** A multidisciplinary approach is needed to secure maternal and fetal survival for patients with Eisenmenger syndrome. Termination of Caesarean section under general anaesthesia may be the method of delivery that can be used, but care must be taken to avoid excessive and rapid blockade, dehydration, and hypoxaemia.

1. Introduction

Eisenmenger syndrome (ES) belongs to the classification of congenital heart disease (CHD), which manifests as pulmonary hypertension with bidirectional or right-to-left blood shunts in the intracardiac or aortopulmonary circulation.[1] This phenomenon was first described by Victor Eisenmenger in 1897.[1] Pregnancy in women with Eisenmenger syndrome is associated with a high mortality rate.[2,3] Therefore, women with ES are recommended to avoid pregnancy and terminate if proven pregnant rather than perform any delivery procedures.[2] A multidisciplinary approach with close communication between health professionals is needed in managing care, especially delivery in pregnant women with Eisenmenger syndrome.

Anesthesia for patients with pulmonary hypertension is controversial.[4] Both general and regional anesthetic techniques for pregnant women with ES have been reported. The basic difference between the two techniques is in general anesthesia with intubation and mechanical ventilation, while in regional anesthesia, maintenance of spontaneous breathing is carried out.[3] However, both central neuraxial anesthesia and general anesthesia have the potential to increase right-to-left shunts, so the clinician's expertise in analyzing needs and patients

may be required. This study aimed to present a case of a pregnant woman with Eisenmenger syndrome who underwent caesarean section under general anesthesia management with severe preeclampsia. The primary goals of anesthesia were to minimize pulmonary vascular resistance (PVR) secondary to hypercarbia, hypoxia, acidosis, stress, and pain and to avoid hemodynamic changes that could increase right-to-left shunts, thereby increasing the severity of hypoxemia.

2. Case Presentation

A 42-year-old pregnant woman (gravida 3, para 1, abortion 1) was admitted to the emergency unit at 38 weeks gestation with the chief complaint of shortness of breath (dyspnea grades II-III) and severe preeclampsia (proteinuria +3). Fifteen years earlier, a baby boy with a birth weight of 2600 grams at term was born vaginally. During her second pregnancy, eight years earlier, the patient had an abortion at 8 weeks. Significant past medical history for heart disease is not known. The patient is currently not on any medication. The patient had New York Heart Association (NYHA) grade 1 dyspnea in the first trimester but increased to NYHA-III in the third trimester.

Physical examination revealed cyanotic with clubbing fingers and elevated jugular venous pressure. Her respiratory rate is 18 times/minute, her blood pressure is 140/85mmHg, and her pulse rate is 75 times/minute. On auscultation, a loud grade III/IV S2 systolic murmur was heard over the precordial with right ventricular elevation, but no additional lung sounds were heard. All risks were explained to the patient's family, and informed consent was obtained for further investigations to be carried out (632/V/HREC/2022).

Laboratory investigations did not reveal any abnormalities. Electrocardiography (ECG) shows normal sinus rhythm and right atrial deviation. A plain chest X-ray revealed cardiomegaly. Preoperative arterial blood gas analysis showed a pH of 7.381, PCO_2 27.2 mmHg, PO_2 45.5 mmHg, HCO_3 15.7 mmol/L, and SaO_2 79.4%. Echocardiography showed biventricular enlargement, 3.4 cm ostium secundum type ASD with dominant right-to-left shunt, dilated pulmonary arteries with severe pulmonary hypertension (35.25 mmHg), moderate tricuspid and mild mitral regurgitation with an ejection fraction of 70%. Abdominal ultrasound examination by an obstetrician showed a live fetus without intrauterine growth restriction (IUGR) and no congenital abnormalities.

General anesthesia was chosen due to obstetric complications, namely severe preeclampsia. The patient was premedicated with a gastroprotective agent, ranitidine 150 mg orally, the night before surgery. Infective endocarditis prophylaxis was given. In the operating room, the patient was placed on a standard monitor, arterial line, and central venous catheter. Severe preeclampsia caused a decrease in systemic vascular resistance (SVR), so prophylaxis for decreasing SVR was not given. In the other cases, our hospital gave prophylactic dopamine infusion 10 µg/kg/min was started to maintain the systemic vascular resistance (SVR) in Eisenmenger syndrome without another concomitant disease. Blood pressure (BP) and central venous pressure (CVP) before induction were 130/90 mmHg and 10 mmHg, respectively.

Rapid sequence induction (RSI) with the prescribed anesthetic dose was performed after we considered the risk of aspiration to hemodynamic instability. Intravenous induction using low-dose ketofol (propofol 50 mg and ketamine 30 g) titrated for anesthetic and hemodynamic effects. After 3 minutes of pre-oxygenation, SpO_2 increased to 99%. Subsequently, the patient was intubated endotracheally with 40 mg rocuronium. Anesthesia was maintained with sevoflurane (end-tidal concentration 1-2%) in oxygen and infusion of fentanyl at a rate of 0.08-0.10 g/kg/min. Blood pressure, stable heart rate (94-123/40-67mmHg, 79-120 beats/minute), and SpO_2 remained 67%-76%, and CVP 10-12 mmHg for 1 hour 45 minutes of operation. The evaluation of fluid balance was monitored during surgery. The patient received 1200 mL of crystalloid (including 400 mL

preload). The remaining 800 mL is infused to maintain the intravascular volume in line with the CVP. Urine output was 100 mL, and intraoperative blood loss was about 700 mL (460 cc fluid balance).

A baby boy within a normal Apgar score with a weight of 2.6 kg was safely born. Intravenous infusion of 15 units of oxytocin is given slowly over 30 minutes. After delivery, tubal ligation is performed. Intraoperative analgesia was obtained with morphine 6 mg i.v. After surgery, the patient was transferred to the intensive care unit (ICU) for monitoring and mechanical ventilation requirements and given a 1 gram/hour iv for postoperative analgesia. At follow-up anesthesia 36 hours after surgery, the patient scored for maximum perioperative comfort and satisfaction.

On the first postoperative day, the uterus was well contracted without active vaginal bleeding, her vital signs were stable (SpO_2 was maintained at 88% with mechanical ventilation), and a chest examination revealed soft wet crackles, and the fluid correction was performed with furosemide 40 mg every 8 hours in combination with vasopressor 0.05 mcg/kg/hour was carried out until the fluid balance reached the target by evaluating pulmonary and systemic pressures. The patient was planned to be extubated at 24 hours postoperatively, starting with nebulized PGE-2 inhibitor 20 mcg every 8 hours and lidocaine 2 ampoules to maintain a dilated condition of the respiratory tract and prevent the risk of a pulmonary embolism due to premature extubation.

The patient's condition gradually stabilized and showed a positive trend during the 5 days of hospitalization. Evaluation of complications of Eisenmenger syndrome continues to be carried out, such as arrhythmias, pulmonary artery rupture, thrombosis, and embolism. Doppler ultrasound evaluation in the pedis region was performed and did not show deep vein thrombosis or other abnormalities. The patient was discharged after undergoing treatment for 6 days after surgery and routinely controlled at an outpatient clinic.

3. Discussion

Eisenmenger syndrome (ES) is a complex combination of cardiovascular pathophysiology that includes clinical cyanosis and mixed right and left cardiac circulation (two-way shunt and pulmonary hypertension).[5] Clinical findings include polycythemia, cyanosis, clubbing, syncope, congestive heart failure, and hemoptysis. In addition, pregnancy is not well tolerated, with maternal mortality reaching 30% to 40%, especially if cesarean or vaginal termination is required.[5,6] Diagnosis of ES is made by clinical findings and confirmed work-up examination. Doppler ultrasound until contrast echocardiography can also be helpful in confirming the diagnosis. Eisenmenger syndrome treated with sildenafil as monotherapy leads to stabilization of the maternal condition and favorable clinical outcomes.[7] Pregnant women with ES may present with low oxygen saturation, dyspnea, fatigue, dizziness, and even right heart failure. Pregnant patients, especially multigravida patients with Eisenmenger syndrome, are rare because this condition is associated with a high mortality rate at term. Peak mortality during delivery and during the first week postpartum is caused by embolism, arrhythmias, right heart overload, myocardial infarction, and acute decrease in systemic vascular resistance. Plasma volume increases in early pregnancy and reaches its peak near term due to a large increase in cardiac preload. The mass of red blood cells also increases during pregnancy but to a lesser extent than the increase in plasma volume. This causes anemia and decreased oxygen-carrying capacity, so pregnant women are susceptible to hypoxia.[8,9] The increase in plasma volume is directly proportional to the increase in cardiac output (CO), which increases from the fifth week of gestation and reaches 35-40% above baseline by the end of the first trimester. Increased CO is caused by increased heart rate, increased cardiac preload, decreased afterload, increased myocardial contractility (most likely due to sex hormones), and left ventricular ejection. Several sex hormones, including estrogen, progesterone, testosterone, and dehydroepiandrosterone (DHEA),

have pulmonary vasodilating effects and may exert antiproliferative effects. On the other hand, estrogens have also been found to increase smooth muscle and neointimal cell proliferation, thereby aggravating pulmonary vascular remodeling.[10]

As a compensatory response, SVR and PVR decrease by up to 40%, resulting in a decrease in biventricular afterload and allowing to accommodate increased CO. Systemic and pulmonary arterial blood pressures and filling pressures remain virtually unchanged because the decrease in afterload is compensated for by the increase in CO.[11] Pregnancy-induced vasodilation and increased cardiac output can increase right-to-left shunts exacerbating pre-existing hypoxia leading to more pulmonary vasoconstriction. Cardiovascular changes associated with pregnancy can take several weeks to return to normal. Cardiac output decreases to just below pre-delivery values at 24 hours postpartum and returns to pre-pregnancy levels only between 12 and 24 weeks postpartum. Pulmonary pressure and resistance return to pre-pregnancy values about six weeks after delivery. Therefore, patients with cardiac disorders remain at risk for several weeks postpartum.[12]

The principle of anesthetic management of ES patients is to maintain a balance between systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR). Appropriate monitoring (e.g., central venous pressure, pulmonary artery catheter, transesophageal echocardiography) can assist in achieving this goal.[13-15] Conventionally, general anesthesia has been recommended for these patients because the relative sympathectomy that occurs with regional anesthesia tends to decrease SVR and increase right-to-left shunts. Maintenance of preload, contractility, normal SVR, and sinus rhythm is important. In addition, maneuvers likely to increase PVR should be avoided. These include (1) hypothermia, (2) acidosis, (3) hypercarbia (hypocarbia tends to decrease PVR; however, if the lower pCO_2 is the result of increased ventilation, then the increase in mean airway pressure will tend to increase PVR), (4) alveolar and pulmonary arterial hypoxia, (5) increased sympathetic output, and (6) sympathetic agents such as epinephrine and norepinephrine. In addition, prophylactic antibiotics are required because of the risk of bacterial endocarditis. In general anesthetic respiratory management, intermittent positive pressure ventilation causes a decrease in venous return and cardiac output and an increase in pulmonary arterial pressure, which together result in an increase in right-to-left shunts.[16] During anesthesia, the patient was ventilated with volume-controlled ventilation (Fabius GS Premium, Germany) with the following settings: tidal volume 8 ml/kg, respiratory rate 12 breaths per minute, positive end-expiratory pressure (PEEP) 5 cmH_2O, and a fraction of inspired oxygen (FiO_2) 50%. This is done because the anesthesiologist assesses oxygen as an effective pulmonary vasodilator.

Almost all general anesthetics have a negative inotropic effect and can impair RV systolic function. Controlled ventilation provides pulmonary mechanics, hemodynamics, and blood gases in postoperative patients.[17] Physiologically, PVR has a U-shaped relationship with lung volume, the lowest value corresponding to functional residual capacity. This makes PVR a major determinant of RV afterload, related to lung volume bimodally. The total resistance of pulmonary circulation depends on the balance of the vascular tone of its two components: the alveolar vessels and the extra-alveolar or parenchymal vessels. Overdistention, on the other hand, causes dead space ventilation (the lung area is ventilated but not perfused), causing hypercapnia. In our patient, induction of anesthesia was performed with propofol mixed with ketamine (ketofol), because propofol decreases PVR and SVR, and the reduction in SVR is sometimes so sudden and severe that it can exacerbate right-to-left shunts, whereas ketamine can increase SVR more than PVR and with thereby reducing right-to-left shunts. Ketamine is not used as the sole induction agent because it causes tachycardia, increased myocardial oxygen consumption, and decreased diastolic filling, leading to low coronary perfusion and decreased cardiac

output, and inhibition of uteroplacental blood flow. Thus, ketofol was administered to counteract propofol-induced side effects such as bradycardia and hypotension. In addition, the analgesic effect of ketamine reduces opioid requirements. Central venous pressure was demonstrated in our case to maintain a stable filling pressure in the presence of general anesthetic-induced vasodilation.

The immediate postpartum period appears to be the most critical time, during which most deaths occur. In the first 72 hours postpartum, hemodynamic changes reach their peak: circulating blood volume and, consequently, CO_2 is increased by autotransfusion of blood from the contracting uterus and by shifting extravascular fluid into the systemic vessels. In addition, PVR is acutely increased, whereas RV systolic function is reduced. All phenomena can cause acute heart failure in patients with impaired RV function. It is important that these critical patients remain monitored for several days in the ICU.

4. Conclusion

A multidisciplinary approach is needed to secure maternal and fetal survival for patients with Eisenmenger syndrome. Termination of the cesarean section under general anesthesia may be the method of delivery that can be used, but care must be taken to avoid excessive and rapid blockade, dehydration, and hypoxaemia.

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