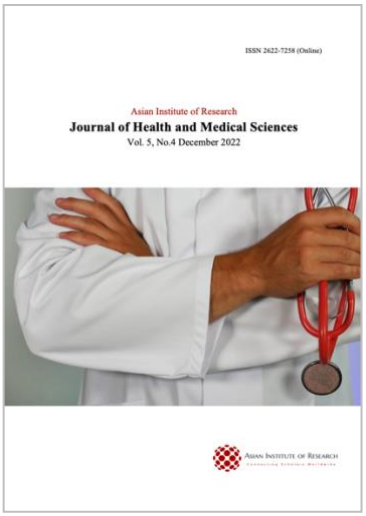

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Anesthesia Management of Sectio Caesarian Patients with Eisenmenger Syndrome and Fetal Distress

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Abstract
Brief overview: 27 year old woman, G2P1A0 gravida 35-36 weeks with congenital heart disease (ventricular septal defect) that progresses to Eisenmenger syndrome, respiratory failure and fetal distress underwent an emergency caesarean section. Management: Anesthesia management was performed under general anesthesia with post-operative care in ICU. Prior to induction, pre-oxygenated with 100% O2 was performed, followed by Rapid Sequence Induction with Ketamine 70 mg, and Rocuronium 50 mg. After the patient had fallen asleep the Sellick maneuver was performed, intubated with ETT no. 6.5. Anesthesia maintenance with Sevoflurane 1 vol%, and 100% oxygen. Fentanyl 50 µg was given after the baby was born. Outcome: In this patient, general anesthetic technique was preferred over regional anesthetic technique for better maternal and fetal outcomes because this patient had already experienced respiratory failure and fetal distress, so a caesarean section was decided as soon as possible. The operation lasted for two hours, with SpO2 during the operation reaching 85% and a live baby boy was born with APGAR scores at the 1st ,3rd and 5th minute 6. Postoperatively, the patient remained intubated and was transferred to the intensive care unit with vital signs blood pressure 122/80 mmHg, pulse 96 beats per minute, SpO2 82-85%.

Keywords: Congenital Heart Disease, Cesarean Section, Eisenmenger Syndrome, General Anesthesia, Pregnant Women

1. Introduction

Eisenmenger syndrome (SE) is a heart disease disorder with pulmonary hypertension and bidirectional blood flow. This case was first reported by Victor Eisenmenger in 1897 in a 32-year-old male patient with a ventricular septal defect and in 1958 by Paul Wood in a larger sample of a population of patients with congenital heart disease with increased vascular resistance. Congenital heart diseases that can cause Eisenmenger syndrome include ventricular septal defect, atrial septal defect, atrioventricular septal defect, persistent ductus arteriosus, transposition of the great vessels. (Duan et al., 2016; Yuan,2016)

With this structural defect, pulmonary blood flow will increase in the early phase, which will then cause increased shear stress on pulmonary blood flow resulting in an increase in pulmonary blood pressure and fibrosis. This situation will cause changes in deoxygenated blood flow from the right side of the heart to systemic blood flow

which will cause cyanosis and decreased pulmonary blood flow. The final phase of this condition is called Eisenmenger syndrome (Lopez et al., 2020; Yentis & Steer, 1998).

Obstetric patients with Eisenmenger syndrome have a mortality rate of 30-70%. Eighty percent of deaths occur on the second and thirtieth day after parturition (Cole, Cross, & Dresner, 2001; Dasgupta, Das, Majumdar, & Basu, 2016; Yuan, 2016). In pregnant women with Eisenmenger syndrome, a decrease in systemic vascular resistance that occurs according to the physiological changes of pregnant women will be followed by an increase in vascular vascular resistance which will cause an increase in pressure on the right side of the heart and the occurrence of shunts and hypoxemia (Lopez et al., 2020). This will increase the mortality rate in patients with Eisenmenger syndrome. Eisenmenger syndrome is included in category IV in the WHO pregnancy risk criteria and is a contraindication to pregnancy (Duan et al., 2016; Suk-Young et al., 2007). Where the cause of death can be caused by hypovolemia, arrhythmia, myocardial infarction, and thromboembolism. Therefore, prevention and termination of pregnancy are recommended for pregnant women with Eisenmenger syndrome (Duan et al., 2016; Lopez et al., 2020; Popat & Russell, 2001; Yuan, 2016).

The goal of anesthetic management is to prevent hemodynamic changes and decrease systemic vascular resistance to prevent shunting of blood flow from the right heart to the left (Cole et al., 2001; Dasgupta et al., 2016). Eisenmenger syndrome patients with advanced pregnancy require multidisciplinary coordination involving the anesthesiology, obstetrics, pulmonology, cardiology, neonatology and intensivist departments to improve the prognosis for both mother and baby where hypoxaemia that occurs will cause stunted fetal growth and premature delivery which will increase the rate of morbidity and mortality in infants (Duan et al., 2016; Lopez et al., 2020; Popat & Russell, 2001).

2. Cases

The patient was identified as a 27-year-old woman with gestational status G2P1A0 gravida 34-35 weeks. The patient came with complaints of shortness of breath since 3 days of SMRS which was felt to be getting heavier on 1 day before hospital admission. Complaints of shortness of breath felt continuously, did not decrease with rest, improved with a semi-sitting position. Complaints of shortness of breath did not decrease with the right and left side position. Complaints of shortness of breath accompanied by cough but not accompanied by fever.

History of shortness of breath during strenuous activity that improved with rest was felt since 2 years before hospital admission. History of frequent awakening at night 2-3 hours after falling asleep due to shortness of breath where this complaint improved with a sitting position since 1 week of before hospital admission. The patient is more comfortable sleeping using 2 pillows since 3 days of before hospital admission. The patient has a history of swelling in both legs that comes and goes since 1 month before hospital admission.

The patient has a history of bluish coloration of the lips and fingertips when crying or doing activities. The patient's history of growth and development is known to be smaller than his age. The patient is known to have congenital heart defects since childhood. The patient is not routinely checked for treatment for heart disorders. History of intermittently stopping breastfeeding when the patient still in childhood is unknown. The patient's parental pregnancy history is also unknown. In her first pregnancy, the patient was admitted to the RSHS 3 years ago and underwent Sectio Caesarea surgery, echocardiography examination and was advised to do right heart catheterization but the patient never came back for control. The patient does not routinely take medication to treat his complaints. The patient also never went to the doctor for control during his second pregnancy, only checked twice with the midwife. The history of the patient's first child, a woman with a birth weight of 1900 grams, born prematurely, was born by caesarean section.

The patient came to the hospital with somnolence, blood pressure at 100/70 mmHg, with a pulse rate of 96 beats per minute. The patient experienced respiratory distress with an increase in respiratory rate of 40-42 breaths per minute and a decrease in SpO2 to 70% with a non-rebreathing mask of 15 lpm. Based on physical examination, the patient's anthropometric status was weight 40 kg, height 155 cm and an additional heart sound was found with a grade 3/6 systolic murmur at the lower left border of the sternal line.

Upon admission, the patient underwent laboratory examinations, chest X-rays, and COVID-19 PCR swabs. In the patient's laboratory results, a decrease in Hb was found at 11.0 and other laboratory results were within normal limits. The chest X-ray shows bilateral pneumonia and cardiomegaly without engorgement. The patient had an echocardiographic examination in 2018 with the results: Site solitus, Multiple VSD, perimembranous diameter 9-11 mm, Bidirectional shunt dominant R to L shunt, PDA (-), Dilated RA, RV, Normal LV systolic function with global normokinetic at rest, High probability of PH, Suspect ASD secundum, LVEF : 61%

Based on preoperative examination, patient was diagnosed with G2P1A0 gravida 35-36 weeks + Maternal Congenital Heart Disease + Eisenmenger Syndrome + Respiratory Failure + Former Cesarean Section + Fetal Distress with physical status ASA IV E.

Upon arrival in the operating room, the patient's clinical condition deteriorated. Standard monitoring equipment was installed, namely oxygen saturation, electrocardiography, blood pressure, temperature and vital signs were assessed. Patients with somnolence consciousness, blood pressure 97/68 mmHg, pulse rate 102 beats per minute, respiratory rate 45 breaths per minute, SpO2 60% with non-rebreathing mask oxygen 15 liters per minute. The fetal heart rate was examined with the results of 120-80-100 beats per minute, irregular.

Anesthesia management in our patients was planned under general anesthesia and in the postoperative cardiovascular intensive care room. Pre-oxygenated with 100% O2, and Rapid Sequence Induction with Ketamine 70 mg, Rocuronium 50 mg, Sellick maneuver, intubation with ETT no. 6.5. Anesthesia maintenance with Sevoflurane 1 vol%, given oxygen: air with FiO2 100%. After the baby was born, Fentanyl was given 50 μg.

The operation lasted for two hours, the hemodynamic duration of surgery SpO2 reached 85% (Figure 2). The baby boy was born alive with APGAR scores in the 1st minute 3 and 5th minute 6 and the birth weight was 2200 grams. Oxytocin was given 20 IU in RL 500 ml 20 drops per minute after the baby was born. Intraoperative bleeding 400 ml with a urine output of 50 ml. The patient received 500 ml of crystalloid fluid intraoperatively.

Postoperatively, the patient remained intubated with sedative and was transferred to the cardiovascular intensive care unit with vital signs, blood pressure 122/80 mmHg, pulse 96 beats per minute, SpO2 82-85% with Jackson Reese 10 liters per minute. The patient received the analgesic fentanyl 25 μg/hour IV and was transferred to the cardiovascular intensive care unit. The patient is then treated in the intensive care unit with close monitoring of hemodynamic conditions, respiratory conditions and complications that can occur. During treatment in the cardiovascular intensive care unit, the patient experienced complications of Hospital Acquired Pneumonia and a pulmonary hypertensive crisis occurred. On the 11th day the patient worsened and died in the cardiovascular intensive care unit.

3. Discussion

Pregnancy is a condition that causes hemodynamic changes in the body to meet the increased metabolic needs of the mother and fetus. Changes in cardiovascular physiology that occur during pregnancy will affect the morbidity and mortality rate that occurs in pregnant women with Eisenmenger syndrome such as heart failure, syncope, and death (Duan et al., 2016; Lopez et al., 2020). In the cardiovascular system, there will be an increase in intravascular volume, an increase in cardiac output, a decrease in systemic vascular resistance and hypotension due to the aortic caval syndrome (Arif, Wahab, & Tofani, 2019).

In the first trimester there is a 40-70% decrease in systemic vascular resistance resulting in an increase in stroke volume and cardiac output. This increase in cardiac output reaches its peak in the second trimester, reaching 25-50% and is followed by an increase in pulse rate. Blood volume will increase by 30-50% until the end of the trimester (Gehlot, Verma, & Raiger, 2021). The progressive increase in plasma volume will increase the burden on the right ventricle so that right heart failure can occur (K. W. Arendt & Lindley, 2019; Arif et al., 2019). In the third trimester, the supine position will reduce cardiac output due to compression of the inferior vena cava due to the enlarged uterus (Gehlot et al., 2021).

During labor, uterine contractions will increase stroke volume, cardiac output up to 30-50% higher before the onset of labor. This condition of hemodynamic changes cannot be tolerated by patients with congenital heart disease conditionsl (Canobbio et al., 2017; Rao & Ginns, 2014). Where 80% of deaths occur between the second and thirtieth day after delivery (Cole et al., 2001).

Table 1: Physiological changes in pregnancy (Arif et al., 2019)

Parameter	Changes
Neurologiy	
MAC	-40%
Respiratory	
Oxygen consumption	+20-50%
Airway obstruction	-35%
FRC	-20%
Minute Ventilation	+50%
Tidal Volume	+40%
Respiratory Rate	+15%
PaO2	+10%
PaCO2	-15%
HCO3	-15%
Cardiovascular	
Blood Volume	+35%
Plasma Volume	+45%
Cardiac Output	+40%
Stroke Volume	+30%
Heart Rate	+20%
Sistolic Blood Pressure	-5%
Diastolic Pressure	-15%
Peripheral Vascular Resistance	-15%
Pulmonary Vascular Resistance	-30%
Hematology	
Hemoglobin	-20%
Thrombosit	-10%
Clotting Factor	+30%-50%

The main principle of the anesthetic technique used is to prevent hemodynamic changes that increase the right-to-left shunt and cause hypoxaemia (Cole et al., 2001). This can be done by preventing a decrease in systemic vascular resistance, venous return and cardiovascular depression (Gehlot et al., 2021). If there is a decrease in systemic vascular resistance, the right-to-left shunt will get worse which will trigger hypoxia and aggravate pulmonary hypertension where in pregnant women there has been an increase in cardiac output and pulmonary blood flow (Gurumurthy, Hegde, & Mohandas, 2012). Decreased functional residual capacity and increased oxygen demand due to pregnancy also predispose to maternal hypoxia (K. W. Arendt & Lindley, 2019; Arif et al., 2019).

Both general and regional anesthesia techniques have advantages and disadvantages, and both techniques carry the risk of increasing right-to-left shunts (Gehlot et al., 2021). The anesthetic technique chosen, both general and regional anesthesia, has the same principle, which is to maintain the flow of cardiac output and prevent a decrease in systemic vascular resistance (Arif et al., 2019; Gurumurthy et al., 2012). If regional anesthetic techniques are used, the advantage is that cardiovascular system depression can be avoided, but sympathetic nervous system blockade can occur which will cause a decrease in systemic vascular resistance which will increase the occurrence

of right-to-left shunts (Soefviana & Zulfariansyah, n.d.). In this case, the general anesthetic technique was chosen with the consideration that general anesthetic technique can provide better oxygenation than regional anesthesia technique because this patient has respiratory failure (K. W. Arendt & Lindley, 2019; Gehlot et al., 2021). The thing that must be considered is the use of anesthetic drugs that can reduce systemic vascular resistance such as volatile anesthetics, propofol, barbiturates. In this case, we used ketamine because ketamine does not reduce systemic vascular resistance so it is safe and effective in patients. Ketamine is also considered safe because it keeps spontaneous respiration and laryngopharyngeal reflex intact so that the patient can maintain his or her airway (Arif et al., 2019). The relaxant agent used is rocuronium at a dose of 0.9 -1.2 mcg/kgbw, due to its rapid onset of 60 seconds (Fawcett, 2019). Lipophilic opioids such as fentanyl are components of general anesthetics that can be used because they suppress the neuroendocrine stress response to surgery (Arif et al., 2019). The use of N2O gas should also be avoided because it can increase pulmonary vascular resistance (Gurumurthy et al., 2012; Yen, 2015).

After delivery, the filling volume of the heart increases significantly because of the loss of compression of the inferior vena cava by the enlarging uterus. This will cause an increase in stroke volume, pulse rate, cardiac output which will increase the risk of heart failure (Katherine W. Arendt, Muehlschlegel, & Tsen, 2012). In general, pregnant women with unstable hemodynamic conditions before delivery will experience a worsening in the early postpartum period, namely in the first 24-48 hours, therefore an intensive monitoring room is needed where these patients receive treatment in the cardiac intensive care room (K. W. Arendt & Lindley, 2019; Canobbio et al., 2017).

4. Conclusion

Anesthesia in pregnant patients with Eisenmenger syndrome requires appropriate perioperative anesthetic management. An anesthesiologist must prioritize maternal and fetal outcomes. Regional and general anesthetic techniques have advantages and disadvantages. In this patient, general anesthesia is preferred over regional anesthesia because of the better maternal and fetal outcomes because these patients already have respiratory failure and require anesthetic techniques that further reduce the decrease in systemic vascular resistance.

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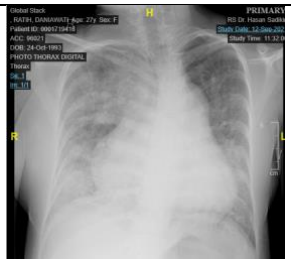
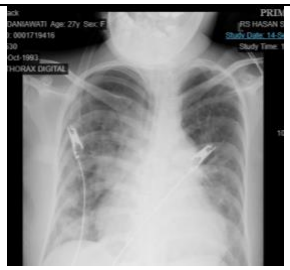
Pictures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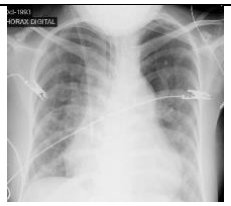
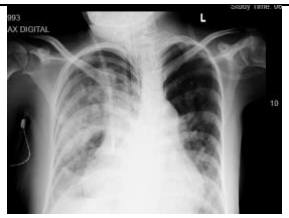
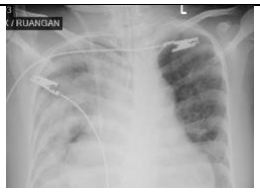
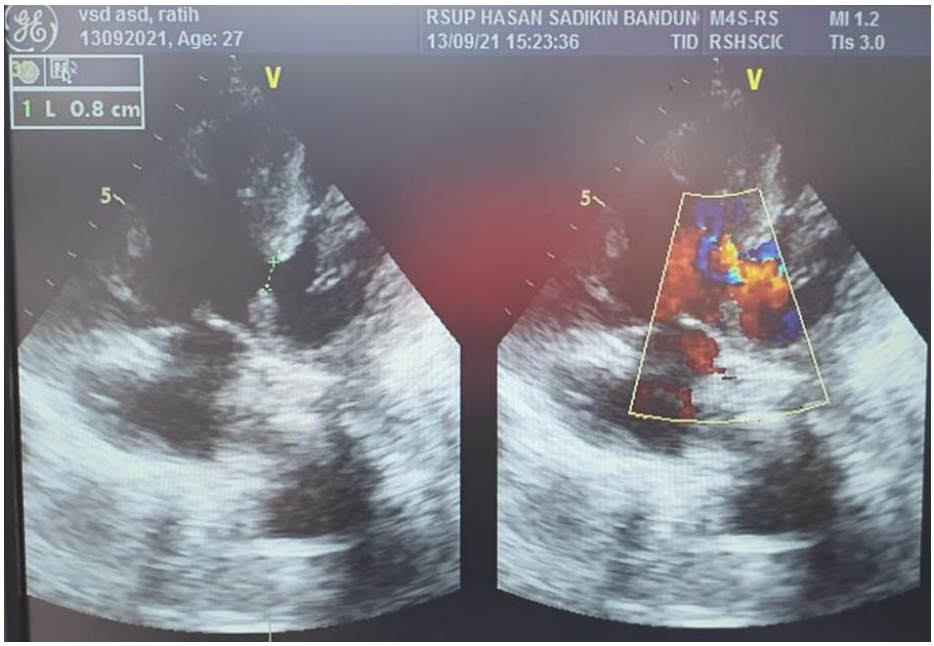
		
Day -7	**Day -9**	**Day -11**

Picture 1: Thorax X-ray



Picture 2: Patient`s Echocardiography