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Case Report

# Anaesthetic challenges in a patient with eisenmenger syndrome: A case report

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ABSTRACT

The term Eisenmenger syndrome describe a large left-to-right cardiac shunt resulting in pulmonary vascular disease and later reversal of the shunt. Occur in 8% of patients with congenital heart disease. Pregnancies carries very high risk of mortality and premature delivery. 20-30% pregnancies result in spontaneous abortion and premature delivery. Pulmonary microembolism and macroembolism have caused peripartum maternal death even after delivery. Anaesthetic management is challenging due to the patho-physiology of the shunt. Our case report is a 27-year-old female known case of Eisenmenger syndrome with chronic thromboembolism presented with inevitable abortion scheduled to undergo emergency evacuation under general anaesthesia.



## 1. Introduction

Eisenmenger in 1897 first described it as large Ventricular septal defect with dextroposition of the aorta.[1,2] In 1958, Wood then described Eisenmenger syndrome as pulmonary hypertension due to increased pulmonary vascular resistance with reversed or bidirectional shunt through a large ventricular septal defect.[3] Later he proposed the communication can occur at any level, can be at atrial level, aortic level or ventricular level.[4] Finally the peripheral vascular resistance is so high the shunt reverses and become right-to-left shunt having worse long term survival rate.[4,5] Eisenmenger syndrome seen in 8% of patients having congenital heart disease.[6] Most common death is due to sudden cardiac death and they face significant perioperative risk of the same.

## 2. Case Report

A 27-year-old, primigravida of gestational age 10weeks 6 days admitted with bleeding per vagina. Later diagnosed inevitable abortion posted for Emergency evacuation. She is a known case of Eisenmenger syndrome, severe pulmonary arterial hypertension, OS-ASD with bidirectional shunt and chronic thromboembolism diagnosed at the age of 20 years. Patient was on home oxygen therapy and heparin. And has stopped taking heparin 2 days before admission by herself. She also has complaints of exertional dyspnea NYHA class 3, with MET score of 1. Physical examination revealed peripheral cyanosis with weight of 40kg, height of 150cm. Jugular venous pressure not elevated and no associated pedal edema with BP of 111/79 Pulse rate 114, SPO2 with 6 liters of O2 was 82%. On auscultation chest was clear, cardiovascular- S1 and S2 heard with loud P2 and mid-systolic ejection murmur. Hemoglobin was 14.5 with D-dimer of more than 10,000 and other blood investigations were normal limit. Echocardiography showed No Regional wall motion abnormality, good left ventricular function, OS-ASD of size 21mm with biventricular shunt predominantly

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R to L shunt, severe pulmonary arterial hypertension, mild PR, mild MR, moderate TR, right atrium and main pulmonary artery dilated, no clots or effusion. She was on warfarin, furosemide, bosentan, sildenafil, metoprolol for the past 1 year. Subsequently warfarin was stopped and changed to Heparin during $1^{st}$ trimester.

Preoperative fasting was 6hrs, Tab. pantoprazole 40mg and Tab. metaclopramide 10mg were given. Advised high risk consent and to arrange ICU bed and ventilator. Infective endocarditis prophylaxis was administered. Patient was taken to Gynaecology Emergency operation theatre and on table pre-induction monitor such as 5 lead-ECG, Heart rate (HR), Pulse oximetry (Spo2), non-invasive blood pressure (NIBP), End-tidal $CO_2$ were attached. Urine output monitored. Intravenous lines secured with 20-gauge cannula on right and left upper limb with Ringer lactate on flow after making sure no air bubble enter inside the lines. Oxygen was administered in anatomical face mask with 6-liter flow rate. Premedicated with Inj. midazolam 0.5mg iv, Inj. ondansetron 4mg iv, Inj. glycopyrrolate 0.2mg iv, Inj. fentanyl 100mcg iv. Preoxygenation with 100% oxygen through mask via bain's circuit. Inj. ketamine 30mg iv, HR-130 per min NIBP-120/70mmHg Spo2-78%. Procedure started. Intraoperative period Spo2 maintained at a range of 74%-80%, heart rate increased to 130 per min, NIBP at 130/80mmHg. Rest of the intraoperative period was uneventful. Procedure over with a total duration of 30 minutes. Infused total 100ml ringer lactate. Patient awake, Spo2-80% at 6 litres of oxygen changed to simple face mask, NIBP of 110/80 mmHg, Chest- clear, air entry bilaterally equal. Obeys commands. Shifted to Gynaecology ICU in oxygen trolley after 10 min accompanied by gynaecology resident. Advised Cardiology evaluation. Patient was asymptomatic and was started on Tab. rivaroxaban 15mg once daily and iv antibiotics. She was later discharged on post-operative day 6.

## 3. Discussion

The anaesthetic challenge is to avoid decrease in arterial blood pressure by maintaining cardiac output and systemic vascular resistance.[7] While inducing the patient there will be decrease in systemic vascular resistance followed by increase in right to left shunt leading to decrease in systemic oxygen saturation and also causes decreased left ventricular filling with right ventricular encroachment.[7,8] Hence ketamine is the ideal agent to use as it does not reduce systemic vascular resistance.[4,9,10] Anaesthesiologist must avoid factors that increase pulmonary vascular resistance which include cold, hypercarbia, acidosis, hypoxia, $\alpha$-adrenergic agonists.[9] Nitrous oxide not used due its interference with pulmonary vascular resistance.[8] Both intra-arterial and non-invasive blood pressure monitors can be used.[9] In our patient, as there is no anticipated major fluid shift hence non-invasive blood pressure preferred and monitored every 3 minutes. As central venous catheterization has potential risk of infection and air embolism and no major blood loss is anticipated in our case, peripheral cannula was placed on both arms. To avoid paradoxical embolization, all intravenous catheters must be free of air bubbles.[4,10,11] General anaesthesia is preferred than regional anaesthetic techniques due to later causing profonde hypotension due to autonomic blockade.[4,8,10–12] Intermittent positive pressure ventilation was avoided as it decreases pulmonary blood flow and increase the right to left shunt.[4,12]

Pregnancy carries high risk of mortality about 30-45% and it positively correlate with degree of pulmonary hypertension.[2,6] Pregnancy itself decreases the systemic vascular resistance and increase the intracardiac shunting and patient additional with thromboembolism can interfere already damaged pulmonary circulation.[7] Patient on prophylactic anti-coagulant has high risk for bleeding and regional anaesthesia would be contraindicated.

Consideration should be given to postoperative observation in an intensive or intermediate care. Avoidance of extreme changes in heart rate, oxygen flow rate and early mobilization is advised.[4]

## 4. Conclusion

Knowledge about the anaesthetic challenges and complication in an Eisenmenger syndrome patient and advocating a strict a plan adds to a successful anaesthetic management.

## 5. Source of Funding

None.

## 6. Conflict of Interest

None.

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