

Anesthetic management of a patient with Eisenmenger's Syndrome for caesarean section

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ABSTRACT

The Eisenmenger's syndrome is presented with an abnormal communication between the systemic and pulmonary circulations and a predominant right to left shunt caused by increased vascular resistance in the pulmonary circulation. We report a successful anesthetic management of a patient with the Eisenmenger's syndrome who underwent emergency cesarean section under general anesthesia.

Key words: Eisenmenger Complex, pregnancy

INTRODUCTION

Victor Eisenmenger in 1897 coined the term Eisenmenger complex, which included large Ventricular Septal Defect (VSD) and pulmonary hypertension^[1]. Wood redefined this in 1958 as pulmonary hypertension with reversed or bidirectional shunt, at the atrio-ventricular or aorto-pulmonary level^[2]. The maternal mortality rate is high, with a cumulative risk of 30-70 %. These patients may survive into adulthood and thus the prospects of incidental surgeries are increasing posing challenges to the anesthesiologists^[3].

CASE REPORT

A 25-year-old primigravida (weight 50 kg, height 155cm) at 31-week and 2 days of gestation was referred to our hospital with complaints of difficulty in breathing, cough and chest pain for past seven days. The patient gave history that additional heart sounds were observed by a general practitioner noted in childhood, but no diagnosis or treatment was performed. She had no symptoms till a week prior to admission, when she developed severe fatigue, progressive cough and shortness of breath. Past medical history was non-significant. Physical examination revealed cyanosis and clubbing of her fingers. Vital signs were temperature 36.8°C, Heart Rate (HR) 120beatsmin⁻¹, Blood Pressure (BP) 140/100 mmHg, oxygen saturation by pulse oximetry (SpO₂) 78% on 6 Lmin⁻¹ of oxygen

by facemask and Respiratory Rate (RR) 28 breaths min⁻¹. Auscultation revealed a loud P₂ and a grade 4/6 pan-systolic murmur at the pulmonary area. There were jugular venous distention and lower extremity edema present.

Patient was shifted to intensive care unit treated by a multidisciplinary team of obstetricians, cardiologists and intensivists and started on noninvasive Continuous Positive Airway Pressure (CPAP) ventilation at 8cm H₂O. Arterial blood gas analysis on room air demonstrated pH -7.41, PaO₂-50 mmHg, PCO₂-25 mmHg, and SaO₂ 86%. Laboratory tests included hemoglobin (Hb)- 18.8 g·dL⁻¹, Hematocrit (Hct)- 58%, platelets-55000/mm³, Activated partial thromboplastin time, Prothrombin time, electrolytes, and serum creatinine levels were normal. Transthoracic echocardiography showed a 14mm sub-aortic Ventricular Septal Defect (VSD) with bidirectional flow, dilated right atrium (55mm) and right ventricle (29mm), right ventricular hypertrophy (12 mm), global

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hyperkinesia of left ventricle, moderate left ventricular systolic dysfunction, severe pulmonary regurgitation (gradient 70mmHg), mild tricuspid regurgitation and moderate to severe pulmonary artery hypertension (RVSP = 50mm Hg) with a left ventricular ejection fraction of 35%. Ultrasonography revealed mild intra-uterine growth retardation

The patient was given betamethasone for lung maturity. She was started on furosemide 20mg intravenous (I.V) twice a day and digoxin 0.25mg once a day. In view of her hypoxemic condition and compromised cardiac status, a caesarean section was scheduled and metoclopramide and ranitidine were used as aspiration prophylaxis. Antibiotics were administered preoperatively to prevent infective endocarditis. Upon arrival in the operating room, RR was 21 breaths min^{-1} , HR 82beats min^{-1} , BP 120/68 mmHg, and SpO₂ 89% on 100% oxygen. General anesthesia was chosen due to thrombocytopenia. Standard monitoring such as electrocardiography (ECG), pulse oximetry (SpO₂), end-tidal capnography (EtCO₂) along with Invasive Arterial Blood Pressure (IABP) was performed. Left uterine displacement was applied by a 15° left-tilt of the operation table. Modified Rapid Sequence Induction (RSI) was performed with titrate-to-effect etomidate (14 mg) to avoid dramatic hemodynamic fluctuations. Intubation was facilitated with rocuronium 30mg. intravenous lignocaine 1.5mg/kg bolus was administered 60sec prior to intubation and esmolol 3mg was given after intubation. Anesthesia was maintained with sevoflurane (1-2% end-tidal concentration) in oxygen. BP and heart rate were stable during the 45-minute operation, and SpO₂ remained 89-96% throughout the uneventful procedure. A male baby was delivered with Apgar scores of 8 at 1 min, 9 at 5 min, and 10 at 10 min. Following delivery of baby, fentanyl 125 μg , midazolam 1mg and esmolol 2 mg was given intravenously. Estimated blood loss was approximately 500 mL, and fluid administration was 750 mL with Lactated Ringer's solution. Urine output was 200mL. Oxytocin was administered intravenously 1unit every 30sec to a total of 5 units. There was no episode of hypotension. There were no other adverse events intraoperatively. She was shifted to intensive care where she was extubated in 2hours. She had completely uneventful recovery and was discharged on seventh post-operative day.

DISCUSSION

In Eisenmenger's syndrome, the amount of right-to-left shunt depends in part on the ratio of Systemic Vascular

Resistance (SVR) to Pulmonary Vascular Resistance (PVR). Regional anaesthesia causes sympathetic blockade that reduces SVR. If SVR decreases without a concomitant decrease in PVR, the amount of right-to-left shunt increases^[4]. Therefore, in our case, we considered it undesirable to induce a sympathetic blockade that might have resulted in an increased right-to-left shunt. We did not administer regional anesthesia as the patient had thrombocytopenia and our obstetric plan was to begin heparin perioperatively.

Though ketamine is preferred in these patients, and our patient already had tachycardia and sympathetic stimulation can lead to increased pulmonary hypertension^[3]. Titrated doses of etomidate were used as induction agent to minimize the risks of decreased SVR and cardiac depression. For maintenance of anaesthesia, nitrous oxide was avoided because it is a potent pulmonary vasoconstrictor^[5]. It was prudent to avoid nitrous oxide due to its added effect on pulmonary vascular resistance. Prevention of hypoxia, hypercarbia, acidosis and hypothermia are very crucial. Hypovolemia needs to be prevented, therefore fasting and intraoperative fluid were carefully planned^[6]. We used intra-arterial blood pressure monitoring as it has the advantage of continuous BP monitoring. Care was taken to avoid bubbles in lines and syringes because of the risk of air embolism. Maternal mortality remains increased in the first three to four weeks after delivery, so prolonged postoperative care in an intensive care unit setting may be needed^[6].

Management of patients with Eisenmenger's syndrome may include the use of supplemental oxygen, digitalis, diuretics, vasodilators, and anticoagulants. Supplementary oxygen for elective and emergency caesarean section is controversial for healthy patients^[7]. In patients with Eisenmenger's syndrome, oxygen is a pulmonary vasodilator which decreases the blood flow across the right-to-left shunt and thereby improves oxygen saturation, so it should be considered for patients with Eisenmenger's syndrome throughout the perioperative period. Maternal arterial oxygen tension should be maintained at 70 mmHg or above when possible^[5]. One must be careful in using digitalis with diuretics in patients who are hypoxemic as there is an increased risk of digitalis toxicity. Diuretics may be useful for patients with Eisenmenger's syndrome and severe right heart failure in order to relieve hepatic congestion or increased intravascular volume. This must be done cautiously to avoid decreasing preload below that required to maintain adequate cardiac output with

increased right ventricular after load. Because of possible teratogenicity and adverse effects on uterine circulation, pulmonary vasodilators including prostacycline analogs, phosphodiesterase inhibitors, and endothelin receptor antagonists are not recommended in pregnancy^[9]. Use of inhaled nitric oxide during labor has been recommended in Eisenmenger's syndrome. However, it was not available at the time in our hospital. In addition, for patients with severe polycythemia, one needs to consider the danger of increased hyperviscosity, which can lead to stroke and other related complications.

The role of anticoagulation in Eisenmenger's syndrome is controversial. Administration of antithrombotic drugs and early ambulation may increase survival in patients with Eisenmenger's syndrome. We did not use anticoagulation in the case because of the high risk of hemorrhage with thrombocytopenia and coagulopathy.

CONCLUSION

Parturient with cardiac lesions are high risk patients and requires adequate and meticulous planning for successful management of such high risk patients.

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