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Eisenmenger Syndrome For Caesarean Section - A Case Report

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Introduction

Initially described by Victor Eisenmenger and redefined by Wood [1], Eisenmenger syndrome is essentially presence of high pulmonary vascular resistance associated with pulmonary artery hypertension (PAH) at, or close to, systemic pressure and associated with a reversed or bi-directional shunt at the aortopulmonary, interatrial or interventricular level [2]. Pregnancy in a patient with this condition causes deterioration due to the physiological cardiovascular changes of pregnancy. Such patients presenting for caesarean section is a major anaesthetic challenge which needs multidisciplinary management. We describe our experience with such a case.

Case report

A young lady, a known case of Eisenmenger syndrome with large ventricular septal defect (VSD), with 34 weeks gestation was admitted for safe confinement. Her obstetric score was G3A2. She developed pregnancy induced hypertension (PIH) at 20weeks of gestation, which was managed with alpha methyl DOPA and Labetalol. Her VSD was diagnosed at 12 years of age. She was not on any cardiac medications and was advised to avoid pregnancy. She was essentially asymptomatic, except for grade 2 dyspnoea on exertion during her third trimester of pregnancy. Her clinical examination revealed central cyanosis, Grade 2 clubbing and minimal bilateral pedal edema. Room air SPO₂ was 75 %. Laboratory investigations revealed Hb - 15.8 mg/dl, Hct - 53, Platelet count - 1.3 lakh /dl with all other baseline parameters in normal limits. ABG showed a pH - 7.36, PaCO₂ 54.5 mmHg, PaO₂ 54.5 mmHg and SPO₂ - 82-88%. ECG showed Right axis deviation with Right ventricular hypertrophy.

Echocardiography demonstrated a large VSD with bidirectional shunt, Severe PAH, moderate coarctation of aorta, dilated right atrium and ventricle with a good left ventricular systolic function and ejection fraction of 60%. Cardiologist evaluated her and categorised her as high risk with 50% peripartum mortality. A multidisciplinary team involving cardiologist, anaesthesiologist, obstetrician and paediatrician was formed to decide on the management of this patient and decision was made to monitor the mother and fetus till term and to do an elective caesarean section.

At 36 weeks of gestation she was posted for emergency caesarean section indicated by abnormal Doppler and severe PIH. After appropriate evaluation and consideration of risks and benefits of general and regional anaesthesia decision was made to proceed with a graded epidural anaesthesia. On table, patient was connected to the monitors including ECG, Pulse oximetry. Right radial artery was cannulated to monitor arterial blood pressure (ABP). Intravenous (IV) access was established through two 18 G cannulae placed on dorsum of right hand and in left external jugular vein. Cardiac output monitoring was done through Flotrac. Mechanical thromboprophylaxis was initiated. Extra precautions were taken to avoid air bubbles in the IV lines and to limit fluid administration. Infective endocarditis prophylaxis comprising of ampicillin and gentamycin was administered as prescribed by the cardiologist. Her baseline parameters were HR- 74/min, ABP - 120/70 mmHg, SPO2 on room air of 76% improving to 89% on oxygen mask. A phenylephrine infusion in a dose of 0.2-0.5 µg/kg/min was started.

An epidural catheter was placed in the L3 - L4 space and was fixed at 8 cm. Incremental doses of 0.375% Bupivacaine titrated against hemodynamic parameters was given. Over 30 minutes a total of 12 ml of 0.375% Bupivacaine and 100 µg Fentanyl was administered, which resulted in a loss of cold sensation up to T8 level. Surgery proceeded uneventfully and was completed in 45 minutes. On delivery of baby 20 Units of Pitocin diluted in 100 ml NS was given slowly. 600 ml of crystalloid was used. After surgery epidural catheter was removed, phenylephrine infusion was tapered and stopped and patient was shifted to intensive care unit. Postoperative analgesia was maintained with tramadol and morphine. Intermittent pneumatic compressions and heparin was used for thromboprophylaxis. After an uneventful postoperative period she was discharged on eighth postoperative day.

Discussion

Pregnancy imposes multiple physiological changes on the maternal body in order to compensate for the increased oxygen demands. This includes a fall in pulmonary vascular resistance (PVR) to increase pulmonary blood flow which fails to happen due to the fixed pulmonary vascular resistance in these patients. Further, the decrease in systemic vascular resistance (SVR) seen in pregnancy further decreases oxygenation by increasing the right to left shunt. In combination all these act to reduce maternal oxygenation further leading to fetal compromise. Maternal mortality in patients with Eisenmenger syndrome has been noted to be as high as 30%-50% [3]. Auto transfusion during labor and delivery can increase the volume load on the heart. Major causes of death in postoperative period are hypovolemia, thromboembolism and preeclampsia [4]. Eisenmenger syndrome with severe PIH is associated with extremely high risk.

Perioperative management of pregnant patients with Eisenmenger syndrome needs a multidisciplinary team approach with intensive monitoring extending well into the postoperative period since majority of maternal deaths happen in postpartum period.

Supplemental oxygen should be provided at all stages of labor. Intensive monitoring in the perioperative period is mandatory. Invasive monitors like pulmonary artery catheter have been at disadvantage due to the vascular complications in these patients with abnormal pulmonary vasculature. Pulse oximetry is the best monitor for assessing the right to left shunt [5]. IBP can demonstrate any sudden change in blood pressure. Adequate analgesia should be given to avoid sympathetic stimulation.

For patients presenting for caesarean section, the main anaesthetic goal would be to maintain the balance between SVR and PVR. The anaesthesiologist should focus on maintaining SVR and intravascular volume, avoiding vena caval compression, myocardial depression and an increase in PVR. Both regional and general anaesthetic techniques have been used, both with benefits and risks associated with each technique. Studies have shown that no single anaesthetic technique has been shown to be superior [6].

General anaesthesia can decrease the venous return due to IPPV and the inhalational agents can cause myocardial depression and a fall in SVR. Performing a rapid sequence induction will cause further harm by the sudden hemodynamic effects caused by the fast administration of the induction drugs. Regional techniques can cause a precipitous decrease in SVR, though a graded epidural produces a gradual fall in SVR [7]. Epidural anaesthesia has now been widely reported in the management of these cases [8,9]. Several of these patients are on anticoagulants which may prove to be a hindrance to choosing a regional technique.

Irrespective of the anaesthetic technique chosen the intensive monitoring should extend into the postoperative period because of the high maternal mortality in the postoperative period. Meticulous fluid management is needed, replacing any blood loss with blood products if needed.

The role of anticoagulation is controversial. Adverse outcomes have been seen with administration of heparin. Judicious pharmacotherapy, mechanical thromboprophylaxis and early ambulation can decrease the chances of thromboembolism and improve the maternal outcome [5].

Conclusion

Successful perioperative management of patients with Eisenmenger syndrome needs a multidisciplinary approach with intensive monitoring. The key to the anaesthetic management would be appropriately balancing the SVR and PVR. The high maternal mortality in the postoperative should always be kept in mind while maintaining an argus eye monitoring in the postoperative period as well.

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