Case Report

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Successful Pregnancy Outcome in Eisenmengers Syndrome with Severe Preeclampsia

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Abstract

Background: Eisenmengers syndrome consists of pulmonary hypertension with a reversed or bidirectional shunt at the atrioventricular or aortopulmonary level.Maternal mortality in the presence of Eisenmengers syndrome is reported to be up to 50% and increases more if there is comorbidity. *Case Report:* We report successful outcome in a primi with Eisenmengers syndrome with severe preeclampsia who underwent emergency lower segment caesarean section for foetal bradycardia. *Conclusion:* Pregnancy should be avoided in case of Eisenmengers syndrome, but one can expect good outcome even with other co morbidity with coordinated multidisciplinary approach with cardiologist, cardiothoracic surgeon, anesthesiologist and obstetrician with intense monitoring to detect adverse maternal and fetal outcome.

Keywords: Eisenmengers Syndrome; Severe Preeclampsia; Pregnancy; Outcome.

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Introduction

Eisenmengers syndrome is a cyanotic heart disease consists of pulmonary hypertension with a reversed or bidirectional shunt at the atrial, ventricular or aorto pulmonary level. Pregnancy ideally should be discouraged in a woman with Eisenmengers syndrome as it carries a high risk (30-50%) of sudden death [1]. Death can occur any time during antenatal period with no difference in incidence in any trimester. Sudden death commonly occurring during postpartum.

Here we report successful pregnancy outcome in a woman with Eisenmengers syndrome with severe preeclampsia.

Case Report

A 28 year old, primigravida with a known history of congenital acyanotic heart disease (ventricular septal defect) attended the antenatal OPD for the first time at 21 weeks gestation for routine check-up. She was asymptomatic. All the examination findings were normal except pan-systolic murmur on auscultation. Cardiology consultation was done and the ECHO showed Sub-pulmonic outlet VSD of 10mm with left to right shunt, severe pulmonary artery hypertension (PASP 117mmHg) and good biventricular function. She was prescribed tablet Torsemide 10mg twice daily & tablet Diltiazem 30 mg thrice daily which she continued. All other routine antenatal investigations including anomaly scan & fetal ECHO were normal. Patient was on regular follow-up. Fetal growth was assessed with serial USG. Pregnancy was progressing well until at 34 weeks 5 days of gestation the patient presented in the OPD with dyspnoea, orthopnoea, cough & headache. On examination, pedal oedema, cyanosis (central and peripheral) and clubbing was seen. Pulse rate was 130/min, Blood pressure-170/110 mmHg, Chest- B/L clear, CVS-S1 &S2 heard, and SpO2- 70% in room air (80% with O2). On per abdomen

examination uterus was 32 weeks size, relaxed with good FHS. Urine albumin was 4+. ECHO revealed VSD with bi-directional flow with severe pulmonary artery hypertension (PASP 130mmHg) and good biventricular function. With a diagnosis of primigravida at 34 weeks5 days gestation with Eisenmenger's syndrome with severe pre-eclampsia, patient was admitted in HDU & continuous Oxygen inhalation at the rate of 6L/min was given. She received Injection Labetalol, injection Frusemide, tablet Torsemide and Diltiazem were continued. Surfactant induction was initiated. Her routine investigation reports were within normal limit. USG showed- Single live fetus at 32 weeks with adequate liquor.Fundoscopy was normal. Close maternal and fetal surveillance was done. Patient delivered a live, male baby weighing 1.6 kgs by emergency LSCS with bilateral tubal ligation (indication-fetal bradycardia) under graded epidural anaesthesia at 35 weeks 2 days gestation. Intra-operative atonic PPH was seen which was managed conservatively. The post-operative period was uneventful. Patient received injectable antibiotics and thromboprophylaxis with injection Enoxaparin. The baby was kept in NICU under observation for 2 days and later was transferred to the mother. The patient was discharged on 11thpost-operative day after cardiology consultation with the advised for follow-up in post natal clinic and cardiology OPD. AT 6 weeks follow-up she was doing well with healthy baby.

Discussion

Incidence of Eisenmengers syndrome is about 3% of patients with congenital heart disease. Maternal Mortality in this setting is high but that associated with VSD (60%) is higher when compared to ASD or PDA [2]. Gliicher et al reported 34% Maternal mortality associated with normal delivery and 75% associated with LSCS [1]. A high incidence of maternal death (43%) was associated with hypovolumia,thromboembolic phenomenon and preeclapsia [3].

Hemodynamic changes associated with normal pregnancy add to the high maternal mortality in patients with Eisenmengers syndrome. There is increase in plasma volume which peaks at about 50% above baseline at 28-30 week, add to the burden of compromised right ventricle and may lead to right heart failure. The preexisting pulmonary vascular disease impede this increased flow of blood to the lungs and increases right ventricular work. In pregnancy decreased systemic vascular resistance increases the degree of the right to left shunting. Therefore pulmonary perfusion decreases and results in hypoxemia and deterioration of maternal and fetal condition.In such case, systemic hypotension leads to decreased right ventricular filling pressure and in the existence of fixed pulmonary hypertension, such decreased right heart pressure may be insufficient to perfuse the pulmonary arterial bed. This paucity may result in sudden profound hypoxemia and mortality.[3] Sudden hypotension can be a consequences of haemorrhage, or complication of conduction anesthesia or hypotension from a vasovagal response to pain may result in mortality.Maternal demise may also occur due to pulmonary embolism or in situ pulmonary infarction.

As Eisenmengers syndrome carries a very high threat to maternal life suddenly pregnancy is contraindicated with this syndrome. Patient should counsel regarding contraception or sterilization or medical termination of pregnancy at 10weeks of pregnancy. If patient choose to continue pregnancy continuous hospitalization is necessary with continuous oxygen administration, since hypoxemia is a potent pulmonary vasoconstrictor oxygen supplementation is recommended to maintain o2 saturation greater than 90% [4]. In cyanotic heart disease foetal outcome is correlating well with maternal haematocrit and successful outcome is unlikely with a haematocrit >65% [5].

Two studies reported good maternal and infant outcome with use of vasodilator (Sildenafil) for management pulmonary hypertension in pregnancy [6,7].

Third trimester foetal surveillance with obstetric ultrasonography and other test for fetal well being are important because at least 30% fetuses will be growth restricted. Maternal arterial partial pressure should be maintained at the level of 70 mm Hg or above. This is explained by a high incidence of spontaneous abortion, 30-50% risk of premature delivery and low birth weight as maternal hypoxemia disturbs fetal growth [1]. Over all foetal wastage with this syndrome is up to 75% [5].

There is no evidence to support any mode of delivery. Vaginal delivery associated with less blood loss but increase in maternal efforts.

Choice of anesthesia is controversial in these patients. Either regional or general anesthesia have been used [8]. Conduction anesthesia has its accompanying risk of hypotension and should be avoided. The conduction of anesthesia can increase the right to left shunt in these patients, regardless the choice of anesthesia. Therefore, maintaining the stable hemodynamic is more important than the choice of anesthesia.The use of epidural or intrathecal

morphine sulfate, a technique may be the best approach to anesthetic management of these patients [5].

Role of prophylactic heparin is not established. As Pitts et al. reported death of five women due to secondary haemorrhage out of seven who received prophylactic heparin post partum [9]. Prophylactic anticoagulation with subcutaneous heparin can be offered, because of the risk of both pulmonary and systemic thromboembolism.

Conclusion

Mortality is prohibitively high in patients of Eisenmengers syndrome with pregnancy. Appropriate counseling for contraception should be given to all patients. If patient conceives then therapeutic termination of pregnancy is recommended in first trimester. If woman wants to continue the pregnancy, this case report suggests, we can expect good outcome even with other co morbidity but it is possible with coordinated multidisciplinary approach with cardiologist, cardiothoracic surgeon, anesthesiologist and obstetrician with intense monitoring to detect adverse maternal and fetal outcome.

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Conflict of Interest

No

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