

# Pregnancy with Eisenmenger's Syndrome

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## ABSTRACT

**Aim:** This case report is aimed at studying the maternal and foetal outcome in pregnancy with congenital heart disease complicated with Eisenmenger's syndrome (ES).

**Background:** Eisenmenger's syndrome is the presence of a reversed or bi-directional shunt at the aortopulmonary, interatrial, or interventricular level along with pulmonary artery hypertension (PAH). This causes severe maternal hypoxemia and life-threatening complications.

**Case description:** We report a rare case of 31-year-old primigravida with 34 weeks gestation diagnosed with congenital heart disease complicated with ES for the first time in the index pregnancy. On evaluation for her breathlessness at 34 weeks gestation, she was found to have a 23 mm ostium secundum ASD with bidirectional shunt with severe pulmonary artery hypertension with PASP of 155 mm Hg. Her pregnancy was further complicated with gestational thrombocytopenia and intrauterine growth restriction (IUGR). She was managed by a multidisciplinary team at our tertiary care centre with sildenafil (vasodilator), diuretics, oxygen therapy, antenatal steroids and ultrasound obstetric doppler surveillance for IUGR. She underwent an emergency caesarean section under epidural anaesthesia at 35 weeks gestation in view of breech presentation in preterm labour. Postoperatively she was monitored closely in cardiac ICU for 3 days and later shifted to ward. She was then discharged 2 weeks later with same medications continued and advice regarding need for cardiac follow up and contraception in order to avoid future pregnancies.

**Conclusion:** Eisenmenger's syndrome in pregnancy is associated with a high maternal morbidity and mortality. The ES patient with PAH should be monitored closely and managed in a tertiary facility with a multidisciplinary teamwork if they choose to continue their pregnancy for a favourable maternal and foetal outcome as in our case.

**Clinical significance:** Pregnancy is best avoided in patients with ES on account of very high maternal and fetal morbidity and mortality. Successful maternal and fetal outcome of pregnancies complicated with ES needs thorough management by multidisciplinary teamwork in a tertiary facility.

**Keywords:** Eisenmenger's syndrome in pregnancy, High-risk pregnancy, Maternal and fetal outcome in pregnancy with Eisenmenger's syndrome, Maternal mortality, Pregnancy with heart disease, Severe PAH in pregnancy.

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## INTRODUCTION

Eisenmenger's syndrome is the presence of a reversed or bidirectional shunt at the aortopulmonary, interatrial, or interventricular level along with PAH. It is uncommon in pregnant patients with congenital cardiac abnormalities, with an incidence of only approximately 3%.<sup>1</sup> Primarily ventricular septal defect, ASD, and patent ductus arteriosus in pregnant women can develop into ES.

## CASE REPORT

We report a case of a young patient who was 31-year-old primigravida with 34 weeks of gestation diagnosed with congenital heart disease complicated with ES for the first time in the index pregnancy and thus referred to our tertiary care center. She had a history of breathlessness on exertion for 3 months, which gradually progressed to breathlessness on ordinary physical activities. On evaluation, she was found to have ostium secundum ASD with bidirectional shunt with severe PAH. She did not have any family history of congenital heart disease. Grade II pedal edema was noted on examination. Her vital parameters were stable with a pulse rate of 98/min, respiratory rate of 20/min, and BP of 110/80 mm Hg in the right upper limb. Jugular venous pressure (JVP) was 3 cm H<sub>2</sub>O. A left parasternal heave was distinctly appreciated with a loud second heart sound P2 on auscultation. Vesicular breath sounds were noted bilaterally with equal air entry on both sides. However, her SpO<sub>2</sub> on room air was 90% on account of the bidirectional shunt in the heart. On obstetric examination, the

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fetus was in breech presentation with clinically mild IUGR with a lag of 2 weeks noted. Chest X-ray showed bilateral pulmonary congestion. ECG showed marked right-axis deviation suggestive of right ventricular hypertrophy (RVH). Echocardiography report revealed a 23 mm ostium secundum ASD with bidirectional shunt, moderate PR, LVEF of 50–55%, and suprasystemic pulmonary artery hypertension with PASP of 155 mm Hg. Also, she had coexisting gestational thrombocytopenia with a platelet count of 90,400/μL. Rest all laboratory parameters viz., LFT, RFT, and PT INR were normal. Obstetric ultrasound revealed findings consistent with IUGR of 3 weeks with an estimated fetal weight (EFW) of 1.8 kg and a normal obstetric Doppler study. The patient was managed by a multidisciplinary team of cardiologists, cardiovascular thoracic

surgeons, senior obstetricians, anesthesiologists, intensivists, and pediatrician. Cardiology opinion was taken, and the patient was started on tab Sildenafil 25 mg twice daily and tab Furosemide 40 mg once daily. The patient was given antenatal steroids for fetal lung maturity and was monitored with weekly doppler studies. However, the emergency cesarean section under epidural anesthesia was done at 35 weeks of gestation when the patient went into preterm labor with a breech presentation. The patient tolerated the operative procedure well and a female child with birth weight 1.54 kg and APGAR of 9/10 was delivered. The neonate was shifted to NICU for further care. The patient was monitored postoperatively in the cardiac ICU and received noninvasive oxygen supplementation, intravenous antibiotics, analgesics, and central venous pressure-guided fluids while NBM and later shifted to the ward on postoperative day 4. She continued to maintain SpO<sub>2</sub> of 88–90% on room air. Postoperatively, she continued with the same medications and the postoperative period was uneventful. A review cardiology opinion was taken and she was advised to continue tab Sildenafil 25 mg twice daily and tab Furosemide 40 mg once daily on discharge. She was then discharged 2 weeks later with advice regarding the need for cardiac follow-up and contraception in order to avoid future pregnancies. On a follow-up visit at 6 weeks with the cardiologist, she was reassessed and advised to continue the same medications.

## DISCUSSION

Congenital heart defects initially result in a left–right shunt, which develops into severe PAH. This eventually causes the left-to-right shunt to become a right-to-left shunt, resulting in ES with significant hypoxemia and cyanosis. Pregnancy with ES can lead to serious consequences such as heart failure, endocarditis, and thromboembolic accidents. Pulmonary thromboembolism and abrupt mortality are more likely to occur in the first few postpartum days. Our case presented with New York Heart Association grade II dyspnea and grade II pedal edema at 34 weeks of gestation. Pregnancy in our patient with ostium secundum ASD was complicated with a severe PAH, leading to ES with bidirectional shunt and IUGR of 3 weeks. Pregnancy with ES is associated with a very high maternal mortality (30–50%) as well as perinatal wastage is up to 75%.<sup>2</sup> Gleicher et al. in their study found that 30% of the fetuses in such cases were growth retarded. Hence, an ultrasound for third-trimester interval growth scans every 2 weeks is necessary and interval growth is monitored.<sup>2</sup> The anesthesia for patients with PAH and the mode of delivery are still controversial. However, our case needed an emergency cesarean

section under epidural anesthesia as she went into preterm labor with breech presentation. General anesthesia can further lower the systemic vascular resistance significantly, thereby increasing the right-to-left shunting. However, Mishra et al. found that epidural anesthesia reduced the chance of precipitous hemodynamic changes in the mother, and is safe to administer epidural anesthesia to patients with ES.<sup>3</sup> Diuretics, vasodilators, anticoagulants, and supplementary oxygen therapy are all used in the treatment of these patients. Prophylactic anticoagulant medication during the peripartum interval is still up for dispute. Oxygen acts like a pulmonary vasodilator, which in turn decreases the right-to-left shunt and thus improves oxygen saturation. The use of sildenafil as vasodilator in PAH cases is supported by Cartago et al.<sup>4</sup> Cartago et al. found that in two cases of ES patients, sildenafil monotherapy led to stabilization of maternal state and a positive clinical outcome.<sup>4</sup>

## CONCLUSION

Eisenmenger's syndrome in pregnancy is associated with a very high maternal morbidity and mortality. Hence pregnancy termination in the first trimester is highly advised, and effective preconception counseling is of utmost importance. If the ES patients with PAH choose to continue their pregnancy, they should be monitored closely and managed with a multidisciplinary teamwork in a tertiary facility for a favorable maternal and fetal outcome as in our case.

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