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# Successful pregnancy outcome in a case of eisenmenger syndrome: A Case Report

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**Abstract**

Eisenmenger syndrome is rare in pregnant women. Maternal mortality is high in Eisenmenger syndrome which is about 30-50% and up to 65% in case of caesarean section. There is foetal wastage up to 75% and 30% of foetuses are complicated with foetal growth restriction. Hence termination of pregnancy is advised in this condition. However only few literature with good maternal and foetal outcome is available in pregnancy with Eisenmenger syndrome. Here we report a successful pregnancy outcome with Eisenmenger syndrome.

**Keywords:** Eisenmenger syndrome, pulmonary hypertension, successful pregnancy, cardiac disease, congenital heart disease.

## Introduction

Eisenmenger syndrome is defined as pulmonary hypertension with reversal of shunt which occurs as a result of uncorrected ventricular septal defect, atrial septal defect or patent ductus arteriosis[(1)]. It was first described by Victor Eisenmenger in 1897. Eisenmenger syndrome has an incidence of 3% in pregnant women with congenital heart disease[(2)]. Main pathogenesis is the decrease in systemic vascular resistance which increases the right to left shunt, leading to sudden circulatory collapse[(3)]. Hence it is a devastating disease and very rare in pregnant women[(2)]. Eisenmenger syndrome is associated with very worse maternal and neonatal outcome[(4)]. Foetal wastage can be as high as 75% with 30% of foetuses with IUGR. Here we present a case of Eisenmenger syndrome diagnosed in pregnancy with successful obstetric outcome[(5)].

## Case Report

A 25year old Primigravida, presented to OPD at 32weeks period of gestation, referred with an ultrasound report showing severe foetal growth restriction and severe oligohydramnios. She had no specific complaints. She had past history of heart disease in childhood for which she had no proper treatment or follow up.

On Examination, she was not anaemic, with no cyanosis or clubbing. Her heart rate was 84/min, regular in rhythm and with good volume, blood pressure – 110/80mmhg, Spo2- 90%. Cardiovascular System Examination revealed Loud P2, Systolic murmur present. RS was normal. Abdominal examination – uterus 26weeks, relaxed, Foetal heart sound – 130bpm. Ultrasonography showed single live intrauterine pregnancy corresponding to 24weeks with growth lag of 7weeks with absent diastolic flow in the umbilical artery. Cardiologist opinion was sought. Echocardiography revealed atrial septal defect with severe pulmonary artery hypertension with pulmonary artery pressure of 128mmHg. She was diagnosed with Eisenmenger syndrome.

**Course**

Patient was administered a full course of corticosteroids for foetal lung maturity. She was managed in the cardiac high dependency unit under multidisciplinary team care. She was planned for termination of pregnancy considering the prognosis of the foetus and was induced with 1 dose of PGE2 gel with a plan for vaginal delivery as morbidity and mortality is high following caesarean delivery. She was given oxygen, maintained in fowler's position and provided adequate analgesia. Her vital signs were continuously monitored throughout the labour. She was given Inj.MgSO4 for neuroprotection. She progressed in labour and delivered a single live preterm baby of weight 614grams with Apgar score of 3/10, 8/10 by assisted breech delivery. Baby was shifted to NICU for preterm care.

Following delivery patient was comfortable, with stable vitals, SPO2 – 96% in 15litres of oxygen. Cardiologist opinion was sought. She was started on antibiotics, diuretics, and Inj.heparin 5000IU BD and then continued on T.Acitrom. Her Hb was 17.2gm/dl hence phlebotomy was done. Patient was advised contraception and to avoid further pregnancy and was discharged on postnatal day 14.

**Follow up**

Baby was discharged after 2months with weight of 1.5kg.Patient is now currently on Bosentan and furosemide. Both the mother and baby are healthy.

**Discussion**

Incidence of Eisenmenger syndrome among congenital heart disease is 3%[6]. Eisenmenger syndrome is associated with high risk of sudden maternal death. Sudden death is mainly because of hypovolemia and thromboembolic phenomena. Mortality rate is as high as 50% in pregnancy with Eisenmenger syndrome[4]. The pathogenesis is due to decrease in systemic vascular resistance which increases right to left shunt which adds on to the hypoxemia. Hypoxemia causes foetomaternal deterioration and polycythaemia. Resulting polycythaemia will lead to hypercoagulable state and thromboembolism[3].

In pre-pregnancy counselling of these patients with Eisenmenger syndrome, it is advised to avoid conception, explaining the risks associated with pregnancy. If the patient conceives, it is advisable to opt for termination of pregnancy in first trimester considering high incidence of maternal mortality[1]. However if the patient is desirous of continuing the pregnancy, a multidisciplinary consultation explaining the prognosis is necessary[5]. In patients who wants to continue the pregnancy, admission is required in second trimester[6]. In the third trimester judicious monitoring of the foetus with ultrasound and Doppler is mandatory, since risk of IUGR is 24%[7]. The incidence of premature delivery of foetus ranges from 30% to as high as 86% reported in various studies[3].

Management of patient with Eisenmenger syndrome includes Oxygen therapy, Diuretics, Digitalis and anticoagulants[3]. Maternal hypoxemia is the very important predictor of foetal outcome. There will be decrease in oxygen saturation below 85%. Because of hypoxemia there is reduced oxygen supply to the foetus which may lead to spontaneous abortion, low birth weight and growth restriction[6]. Increase in plasma volume in pregnancy can also increase the workload to the heart and result in heart failure. There is also further increase in pulmonary vascular resistance which increases the hypoxia resulting in decrease oxygen supply to the foetus which leads to foetal growth restriction[1]. Hence oxygen therapy will help to improve the maternal and foetal hypoxia. Diuretics may be useful for patients in heart failure as it decreases the cardiac load[3]. Digitalis is also used to manage heart failure in these patients[9]. Role of anticoagulation in this patients is controversial, it is beneficial in preventing thromboembolism[10].

In anticipated iatrogenic preterm birth, sometimes antenatal steroids are indicated[10]. Using Sildenafil and L-arginine in these patients throughout pregnancy had shown better clinical improvement in some studies[11]. Caesarean section leads to high risk of mortality[4]. Hence vaginal delivery assisted by forceps is recommended. Gleicher et al, found 34% mortality associated with vaginal delivery and 75% associated with caesarean section[12]. Monitoring of these patients in early postpartum period is crucial, as it is assumed as the most dangerous period with 70% of death occurs in early postpartum period[3]. There is worsening of the pre-existing hypoxia in Eisenmenger syndrome during pregnancy this is due to physiological changes in pregnancy, due to decrease in systemic vascular resistance there is increase in right to left shunt which adds on to the hypoxemia[6]. So to compensate this hypoxemia there is raise in haemoglobin and haematocrit. Hence Phlebotomy is recommended in these patients to decrease the haematocrit[13].

**Conclusion**

Women with Eisenmenger syndrome should be made aware of the significant risk of mortality in pregnancy and should receive good contraceptive advice. Hence pre-conceptional counselling plays a major role in any cardiac disease complicating pregnancy. Pregnant women with Eisenmenger syndrome should be offered termination due to its associated risk of 50% mortality. Any unexplained IUGR in the early third trimester of pregnancy, should be evaluated for underlying cardiac disease. Early diagnosis and prompt management by a multidisciplinary team in a tertiary care centre can give a successful pregnancy outcome in patients with cardiac disease.

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