

# Pregnancy with Eisenmenger Syndrome: A Challenge to Obstetrician

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## ABSTRACT

Eisenmenger syndrome is defined as the development of pulmonary hypertension in response to a left-to-right cardiac shunt with consequent bidirectional or reversal (right-to-left) of shunt flow. Maternal mortality in the presence of Eisenmenger syndrome is reported to be 30-50%. If the patient continues her pregnancy against advice, a well-coordinated multidisciplinary team approach is advocated. Here, we report a case of pregnancy with Eisenmenger syndrome and its successful outcome.

**Keywords:** Pregnancy, Eisenmenger syndrome, maternal mortality

Congenital heart disease patients are reaching reproductive age due to improved healthcare facilities and more of them are conceiving. Eisenmenger syndrome involves pulmonary hypertension with a reversed or bidirectional shunt at the atrial, ventricular or aortopulmonary level.

Eisenmenger syndrome in pregnancy is usually associated with high mortality rates of around 30-50%.

Such patients are advised against pregnancy or to interrupt pregnancy before 10th gestational week, but if they continue pregnancy against advice, a well-organized multi-specialist care is required.

Here, we report a successful pregnancy in a woman with Eisenmenger syndrome.

## CASE REPORT

A 30-year-old woman, G<sub>3</sub>P<sub>1</sub>A<sub>1</sub>, gestational age (GA) 36 weeks, presented to our setting with chief complaints of pain abdomen for 6 hours and breathlessness (dyspnea on less than ordinary activity and orthopnea) for 3 days. She had marked limitation of physical activity (New York Heart Association [NYHA] Class III).

**Obstetric history:** Previous lower segment cesarean section (LSCS) 4 years back (Indication – cephalopelvic disproportion [CPD]), miscarriage in first trimester 3 years back.

**Past history:** She had no history of dyspnea previously. She had a previous uneventful cesarean section. Her symptoms were much worse in this pregnancy.

Recently diagnosed with pulmonary arterial hypertension (PAH) and atrial septal defect (ASD).

Echocardiography revealed moderate-sized ASD, severe PAH, moderate pulmonary regurgitation (PR) and tricuspid regurgitation (TR). Routine antenatal investigations were within normal limits.

**General examination:** Blood pressure (BP) - 124/80 mmHg, pulse rate - 92/min, Pallor +, edema -, cyanosis ++.

**Systemic examination:** CVS: S1 normal, S2 single-narrow split, pulmonary ejection systolic murmur, enlarged right heart.

**P/A:** Uterus 36 weeks size, longitudinal, cephalic, moderate contractions, FHS + tachycardia, scar tenderness present. As per cardiological review, she was treated with propped up position and oxygen inhalation. Injection ampicillin 2 g and injection gentamicin 80 mg were given as infective endocarditis prophylaxis.

Decision for emergency LSCS was taken. Her cesarean section was performed. A healthy baby of weight 2.9 kg was born. Her bilateral tubal ligation was performed. Intraoperative and postoperative period was uneventful. Continuation of same cardiac treatment was done. Patient was discharged on 8th post-op day in a good condition with a healthy baby.

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## DISCUSSION

The hemodynamic changes occurring in pregnancy are not well-tolerated among women with Eisenmenger syndrome. Most women with Eisenmenger syndrome have a delicately balanced state and this balance must not be disrupted. In women with Eisenmenger syndrome and a low cardiac output state, the right ventricle is compromised, which may fail to meet the demands of increasing blood volume and cardiac output in pregnancy.

A fixed pulmonary vascular resistance and failure to increase pulmonary blood flow may not be able to hold an increase in cardiac output. A compromised cardiovascular system may not be able to tolerate massive fluctuations in blood volume, pre- and postpartum. The decrease in peripheral vascular resistance during pregnancy can amplify right-to-left shunting, and aggravate maternal hypoxemia and cyanosis.

Pregnancy is a cause of significant mortality in women with Eisenmenger syndrome. A systematic review of published studies from 1978 to 1996 examined maternal mortality rates in women with Eisenmenger syndrome and demonstrated mortality rates of 56%. The degree of maternal hypoxemia is a key predictor of fetal outcome. Pre-pregnant arterial oxygen saturation of 85% or less is tied to live birth rate as low as 12%, while saturation of 90% or more is associated with a 92% live birth rate. This can be explained by an increase in spontaneous abortions, risk of premature delivery of around 30-50% and low birth weights.

Maternal mortality, in association with Eisenmenger syndrome, has been reported to be as high as 30-50%. Gleicher et al reported a 34% mortality associated with vaginal delivery and a 75% mortality associated with cesarean section.

Mortality is high if pregnancy is continued. Therefore, abortion is the treatment of choice for women who have Eisenmenger syndrome. For patients who wish to continue gestation, hospitalization in the second trimester is highly recommended.

Intrauterine growth restriction is observed in 30% of pregnancies on account of maternal hypoxemia. Furthermore, premature labor is encountered in around 50-60% of cases and the high perinatal mortality rate of around 28% is often attributed to prematurity. In a study conducted among women with Eisenmenger syndrome, 47% delivered at term, 33% delivered between 32 and 36 weeks, and 20% delivered prior to 31 weeks of gestation.

Continuous administration of oxygen, anticoagulation and pulmonary vasodilator is disputed. While there is a scarcity of controlled trials, a Brazilian series of 13 pregnancies revealed that there was improved maternal mortality (23%) with oxygen therapy, heparin before delivery and warfarin after 48 hours. About 60% of infants were live births, mostly premature. There seems to be no evidence to support the choice of vaginal or cesarean delivery based on cardiac reasons. However, vaginal delivery is tied to a lower average blood loss at the cost of escalated maternal effort.

Mortality among patients with Eisenmenger syndrome who conceive remains high. All patients should receive appropriate advice regarding contraception. If a patient becomes pregnant, clinicians must offer therapeutic termination of pregnancy. However, if pregnancy is continued against medical advice, treatment strategies mentioned above may help, with prolonged hospital care, pre- and postpartum.

## CONCLUSION

Although pregnancy should be discouraged in women with Eisenmenger syndrome, it can be successful with careful monitoring and a well-coordinated multi-specialist care, as elucidated in this case.

## SUGGESTED READING

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