

**EISENMENGER SYNDROME IN PREGNANCY- A REVIEW ARTICLE****P. Priyadarshini\***

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**ABSTRACT**

Eisenmenger syndrome is very rare in pregnant women. Debates remain concerning the management of Eisenmenger syndrome in this patient population and the prognosis is unclear in terms of maternal and fetoneonatal outcomes. Epidural analgesia is preferred for Cesarean section as it alleviates perioperative pain and reduces the pulmonary and systemic vascular resistances. Maternal mortality in the presence of Eisenmenger syndrome is reported as 30-50% and even up to 65% in those with Cesarean section. The major causes of death could be hypovolemia, thromboembolism and preeclampsia. The purpose of this article is to discuss the debates of Eisenmenger syndrome in pregnancy and the possible resolutions.<sup>[1]</sup>

**KEYWORDS:** Eisenmenger Complex, Pregnancy Complications, Hypertension, Pulmonary and systemic vascular resistance.

**INTRODUCTION**

Pregnancy is a cause of significant mortality in most published series of women with Eisenmenger syndrome. Eisenmenger syndrome is very rare in pregnant women. Debates remain concerning the management of Eisenmenger syndrome in this patient population and the prognosis is unclear in terms of maternal and fetoneonatal outcomes.<sup>[2]</sup> Pregnancy should ideally be avoided in a woman with Eisenmenger syndrome concerning the high maternal mortality rate and probable poor prognosis of the baby. A short labour and an atraumatic delivery under epidural block are preferred in the women with a strong desire of pregnancy.

**Definition**

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.

Initially, left-to-right intracardiac shunting is associated with increased flow (and sometimes transmitted pressure) through the pulmonary vasculature. This results in pulmonary vascular remodeling and leads to pulmonary vascular disease. The pulmonary arterial hypertension and associated elevation in right heart pressures result in reversal of the shunt with either right-to-left or bidirectional flow, which is called Eisenmenger syndrome.<sup>[3,4]</sup> Congenital heart defects that can lead to Eisenmenger syndrome include: atrial septal defects, ventricular septal defects, persistent arterial ducts, as

well as more complex defects such as atrioventricular septal defects, truncus arteriosus, aortopulmonary window, complex pulmonary atresia, and the univentricular heart. As a result of the right-to-left shunt, patients are chronically hypoxemic, hence cyanotic.

**Effects of pregnancy-related hemodynamic changes**

The hemodynamic changes of pregnancy are usually poorly tolerated in women with Eisenmenger syndrome.) Most women with Eisenmenger syndrome are in a precariously balanced state and an important principle of care is to not disrupt this balance. In women with Eisenmenger syndrome and a low cardiac output state, the compromised right ventricle may not meet the demands of increasing blood volume and cardiac output associated with pregnancy. In addition, a fixed pulmonary vascular resistance with a resulting inability to increase pulmonary blood flow may not accommodate an increase in cardiac output.<sup>[5,7]</sup> Similarly, large fluctuations in blood volume both pre and post partum may not be tolerated by an already compromised cardiovascular system. The fall in peripheral vascular resistance that occurs during pregnancy can augment right-to-left shunting, worsening maternal hypoxemia and cyanosis (1). During pregnancy, the blood becomes more hypercoagulable and in the cyanotic patient the risk of deep venous thrombosis, pulmonary infarction, and/or paradoxical embolus and stroke increases.

**Maternal Cardiac Complications**

Pregnancy is a cause of significant mortality in most published series of women with Eisenmenger

syndrome.<sup>[7]</sup> A systematic review of published studies from 1978-1996 examined maternal mortality rates in women with Eisenmenger syndrome and demonstrated mortality rates of 56%. A more recent review suggested that mortality remains high. Most complications occur near term and early (1st week) post-partum, and therefore extended post-partum hospital observation is suggested. Mortality is typically from heart failure, sudden death presumably due to arrhythmias, or thromboembolic events.<sup>[2]</sup> During pregnancy, it is important to watch for cardiac symptoms including increasing fatigue, worsening peripheral edema, palpitations, chest pain that could reflect right ventricular ischemia, and/or volume overload and presyncope/syncope with exertion reflecting a decrease in cardiac output. However, other complications as described can also occur, particularly thromboembolism.

### Fetal Complications

Miscarriage is common in cyanotic women. Intrauterine growth restriction is seen in 30% of pregnancies as a result of maternal hypoxemia.<sup>[6]</sup> Premature labour is found in 50-60% of instances and the high perinatal mortality rate (28%) is due mostly to prematurity. In one study of women with Eisenmenger syndrome, 47% delivered at term, 33% between 32 and 36 weeks, and 20% before 31 weeks of gestation.

The mortality of patients with Eisenmenger's syndrome who become pregnant remains prohibitively high. Appropriate advice regarding contraception should be given to all patients.<sup>[5]</sup> If a patient becomes pregnant, therapeutic termination should be offered. If pregnancy continues against medical advice, treatment strategies as outlined above may be helpful, with prolonged hospital care both pre and post partum.

### Management

Some patients, however, may benefit from nocturnal supplementation, although oxygen therapy is most useful as a bridge to heart-lung transplant.

### Fluid Balance and Climate Control

Patients should avoid sudden fluid shifts or dehydration, which may increase right-to-left shunting. They should also avoid very hot or humid conditions, which may exacerbate vasodilation, causing syncope and increased right-to-left shunting.

### Right Heart Failure

Right heart failure is often present in patients with Eisenmenger syndrome, but limited treatment options are available for this complication. Typically, digoxin and diuretics have been used. Diuretics, specifically loop diuretics, are used for symptomatic relief of congestion. However, diuretics should be used cautiously in these patients, given their preload-dependent state.

Central nervous system (CNS) events can occur secondary to paradoxical embolus, CNS venous

thrombosis, intracranial hemorrhage, or brain abscesses in the setting of endocarditis.<sup>[4]</sup> Endocarditis prophylaxis, the use of air filters with all intravenous catheters during hospital admissions, and adequate management of hyperviscosity can help to decrease these potentially fatal complications.

### CONCLUSION: CNS EVENTS

Pregnancy should ideally be avoided in a woman with Eisenmenger syndrome because of a high maternal mortality rate and probable poor prognosis of baby. These patients with continuing pregnancy should be assessed by a combined and experienced multidisciplinary team, including obstetric, anesthetic, cardiology, pediatric and neonatal physicians. Epidural anesthesia is preferred in Cesarean section<sup>[2]</sup>. A short labour and an atraumatic delivery under epidural block are preferred. The use of heparin and oxygen therapy presumably positively influenced maternal and infant outcomes.

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