

# A Rare Case of Pregnancy with Eisenmenger Syndrome

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## Abstract

*A 29 year old primigravida patient admitted to Al Qassimi Hospital for induction of labor at 34 wks due to mild pre-eclampsia. She then went on to develop dyspnea. After a multidisciplinary approach in conjunction with obstetricians, anesthetists and cardiologists the patient was diagnosed as having Eisenmenger syndrome. She was managed in accordance with the protocols of the respective departments. The baby was delivered via an uncomplicated lower segment Cesarean section. Patient was managed in the ICU with multidisciplinary teams of doctors. She stabilized after the surgery and was discharged home in good health on the sixth post operative day on oral antihypertensives with a follow up in cardiology clinic and post-natal gynecology clinic.*

## Keywords

*Eisenmenger syndrome, Cardiac disease in pregnancy, Right to left shunt*

## I. Introduction

Eisenmenger Syndrome is a multi-system disorder defined as a development of pulmonary hypertension with a consequent right to left shunt and/ or bidirectional flow in response to a prior cardiac defect. This is usually associated with congenital cardiac

defects at the atrial, ventricular and/ or aortopulmonary level [1].

Pregnancy with Eisenmenger's Syndrome, has an estimated mortality that exceeds 50% [2]. If detected in time and managed appropriately, the mortality associated with this condition can be reduced. This report aims to highlight this condition and its presentation in a pregnant lady and how effectively she was managed.

## II. Case Details

A 29 yr old primigravida was admitted to Al Qassimi Hospital at 34 weeks gestational age, as she was a case of gestational diabetes controlled on diet with mild pre-eclampsia and obstetric cholestasis, as additional complicating factors. She had received antenatal steroids for lung maturity and was on ursodeoxycholic acid tablets, prior to admission. Patient had no significant past medical/ surgical history.

She had been decided for induction of labor, in view of developing pre-eclampsia. She had a blood pressure of 143/84 mm Hg with 1+ proteinuria on admission and anti-hypertensive treatment with Methyldopa 250 TID was initiated.

Next morning when she was being evaluated, before starting induction, patient started complaining of breathless at rest. On examination, peripheral and central cyanosis was noted. On further examination, she had a pulse 90/min, BP=137/80mm of Hg, respiratory rate=24/min, oxygen saturation=88 % ( on room air), with a raised JVP. On auscultation of the chest bilateral basal crepitations were heard and a pansystolic murmur was heard on auscultation of the cardiovascular system. On cardio-tocographic examination normal fetal heart tracing was recorded but irritability of uterus was present. Immediately, multidisciplinary care was initiated and consultant obstetrician was called in and medical, cardiac and anesthesia teams were involved in planning further care. After resuscitation with oxygen by mask (6L), oxygen saturation improved to 99%. Urgent ECG revealed right bundle branch block with T wave inversion in V1-V3, while cardiac

enzymes were normal. A CXR revealed cardiomegaly with bilateral pulmonary congestion. Patient was started on furosemide.

Relevant laboratory investigations were within acceptable ranges, except for a raised alt (49 IU/L) and uric acid (391mmol/l) and LDH (391 IU/l). The hematocrit was 44% (upper normal limit).

Arterial blood gas analysis revealed a pH- 7.37, pco<sub>2</sub>- 46, po<sub>2</sub>- 58 and hco<sub>3</sub>- 26. bedside echo findings revealed a large VSD with a diameter of 1.96cm with a right to left turbulent flow on Dopplers. The ventricular function appeared normal

The decision for urgent delivery was taken in view of maternal condition and an emergency LSCS was done. Prophylactic antibiotic cover was given prior to the surgery. A healthy male baby weighing 2.5kg was delivered with good APGARs and patient was shifted to ICU for post-operative care.

She was managed with a multidisciplinary approach with the contribution of cardiologist/ senior obstetricians and anesthetists. Methyldopa 250mg TID, Furosemide 40mg OD, enoxaparin 40 U SC injection, O<sub>2</sub> therapy and broad spectrum antibiotics were continued. No post-operative complications were noted. Stitch line was healing well and breast milk expression was encouraged from day 1 post op. The baby was reported to be doing well with no immediate complications and was shifted to NICU in view of prematurity.

A repeat 2-D ECHO was done for the patient that revealed a large premembranous VSD. A cardiac surgeon was also consulted.

On the fourth post-operative day, patient was shifted from the ICU to the wards. Her pre-eclampsia settled with systolic and diastolic

readings ranging from 150-120mm Hg and 81-69mm Hg respectively. Her average respiratory rate ranged from 18-31 breaths /min. She was discharged on the 6th post op day on oral antibiotics and anti-hypertensive medications.

She was counseled regarding her cardiac condition and the couple was advised against pregnancy till a definitive treatment was done. She was advised a follow up cardiac consultation after three months for planning further management.

### III. Discussion

It is important to note that this pregnant lady was an undiagnosed case of congenital cardiac defect that decompensated suddenly at 34 weeks of pregnancy into class three heart failure. Thankfully she was in hospital where she received immediate multi-disciplinary attention that resulted in successful maternal and neonatal outcome.

Literature has stated that pregnancy adds an extra cardiac burden in a patient with a congenital cardiac defect, more so in a patient with Eisenmenger Syndrome, where this load adds an increased risk of maternal mortality.

During the antepartum period, the decreased systemic vascular resistance associated with pregnancy increases the likelihood and the degree of right to left shunting. The pulmonary perfusion then decreases; which results in hypoxemia and deterioration of the maternal and fetal condition. In such a patient, systemic hypotension leads to decreased right ventricular filling pressure and in the presence of fixed pulmonary hypertension, such decreased right heart pressure may be

insufficient to perfuse the pulmonary arterial bed. This insufficiency may result in sudden profound hypoxemia and death. Such hypotension can result from hemorrhage or complications of conduction anesthesia and can lead to sudden death. Such an occurrence is the principal clinical concern in the intrapartum management of patients with pulmonary hypertension [3].

maternal mortality in association with VSD is higher (60%) than in association with atrial septal defect (ASD) (44%) and patent ductus arteriosus (PDA) (41.7%). the majority of maternal deaths occurred during or within the first week after delivery. only 25.6 per cent of all pregnancies reached term. at least 54.9 per cent of all deliveries occurred prematurely [4]. Eisenmenger syndrome is one of the every few conditions in which pregnancy is absolutely contraindicated. Mortality is typically from heart failure, sudden death presumably due to arrhythmias, or thromboembolic events.

A multidisciplinary approach is mandatory in a tertiary care setup. Close cardiovascular monitoring, with specific attention to volume status, is essential, as both hypovolemia as well as volume overload should be avoided [5].

One of the biggest risks is that of thromboembolism primarily due to the hypercoagulable state caused by pregnancy itself superimposed by polycythemia due to longstanding hypoxemia. Prophylactic anticoagulants are thus advised.

Contraception is essential for these patients, till definitive treatment is obtained. Sterilization is the best option, if family is complete but for a young patient, whereas progesterone only pills can be offered [6].

Combined oral contraceptive pills are contraindicated.

#### IV. Conclusion

Eisenmenger Syndrome outside of pregnancy is a challenging and arduous task to manage which becomes more complicated when occurring with pregnancy. It is therefore prudent to involve a multidisciplinary team in the management of such patients to allow for optimum maternal and fetal outcome. This case report therefore aims to highlight the same.

#### V. Disclosure of Interest

Dr Subul Bazmi collected the case details and was involved in summation of the case details. Dr Shalini Malhotra verified the case details and supervised the finalization of the case report. Dr Faez Zaman was involved in the management of the patient.

#### VI. Acknowledgements

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