

Ventricular septal defect (VSD) with Eisenmenger Syndrome in a pregnant female- A case report

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ABSTRACT

Eisenmenger syndrome (ES) is a condition where a previous congenital heart disease with right to left shunt eventually because of development of pulmonary arteria hypertension converts into a left to right shunt bypassing the lungs. Patients who were previously acyanotic become cyanotic and hypoxic. ES when associated with pregnancy increases the maternal and foetal risk. There is a 50% risk of sudden maternal death in ES. The overall estimated foetal wastage is reported to be up to 75%. Therefore patients with ES are advised not to conceive and if they become pregnant should terminate pregnancy by the end of first trimester. Treatment of these patients requires a multi-specialist care when termination of pregnancy is inevitable. We present an antenatal case of VSD with ES.

Keywords: Pulmonary, hypertension, trimester, acyanotic, cyanosis

1. INTRODUCTION

Eisenmenger's syndrome (ES) is development of pulmonary arterial hypertension with a reversal of shunt at the aortopulmonary or atrioventricular level like ventricular septal defects, atrial septal defect, patent ductus arteriosus, D-transposition of the great vessels, and surgically created aorto pulmonary connections (Wood, 1958). The severity of progression of ES depends on the size of intracardiac defect and its location (Beghetti & Galiè, 2009). ES eventually leads to complications like right ventricular (RV) failure, right ventricular hypertrophy and patients usually develop heart failure, dyspnoea, cyanosis, clubbing and arrhythmias.

ES is very rare in pregnancy with an incidence of around 3% with congenital heart defects (Rathod & Samal, 2014). As in pregnancy there is increased circulatory burden and this results in high maternal mortality rates around (30-50%) in pregnancy with ES (Avila et al., 2014; Gleicher et al., 1979; Jones et al., 1965; Yentis et al., 1998). The major causes of death in ES are right ventricular failure, pulmonary hypertension crisis, arrhythmia, and stroke.

The management and maternal and fetoneonatal outcome of Eisenmenger syndrome associated with pregnancy is unclear. Eisenmenger syndrome results in around 30-50% of maternal mortality and Caesarean section results in around 65% of maternal mortality.

Epidural analgesia is of choice in caesarean section as it reduces perioperative pain and decreases systemic vascular along with pulmonary resistances. Hypovolemia, preeclampsia, thromboembolism are the major causes of mortality. As Eisenmenger results in high maternal mortality and bad prognosis of the baby it's better to avoid pregnancy in Eisenmenger syndrome. A short labour and a non-traumatic delivery under epidural anaesthesia is the modality of choice in the women who eagerly want to become pregnant.

2. CASE REPORT

24 year old female primigravida with 35.4 weeks of gestational age presented with dyspnoea and palpitations. There was no history of giddiness, haemoptysis, chest pain, syncope. On examination patient had BP 100/60 mm of Hg, and pulse rate was 90 bpm, regular and saturation was 82% on room air, there were no signs of heart failure and patient had clubbing, cyanosis, and low volume pulse. CVS examination revealed a holosystolic murmur in the left lower sternal border with loud pulmonic component of second heart sound on auscultation.

Investigations revealed Hb- 14 gram%, TLC 7,800/ mm. CXR showed cardiomegaly with prominent pulmonary conus (fig.1). ECG showed right axis deviation, right atrial enlargement and right ventricular enlargement with RV strain (figure 2). 2 D echo revealed VSD with eisenmengerization (fig. 3).

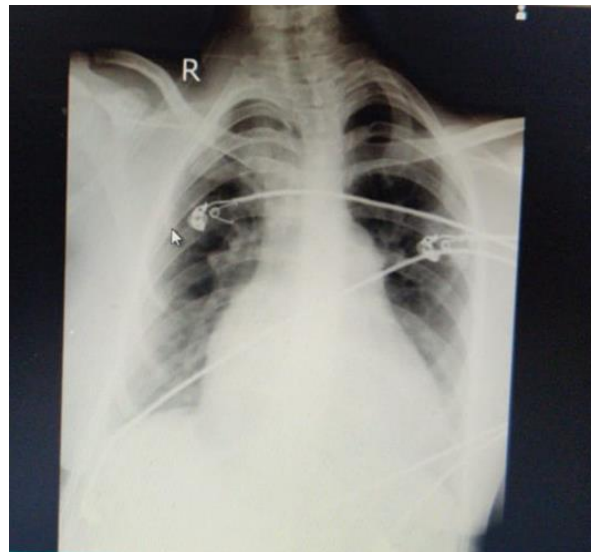


Figure 1 Chest x ray showing cardiomegaly with dilated pulmonary artery

She was diagnosed as a case of Eisenmenger syndrome with severe PAH with 20 mm mid muscular non-restrictive VSD. She was taken for elective caesarean section at 36 weeks 4 days period of gestation under combined spinal and epidural anaesthesia, Intraoperative period was uneventful. A male baby weighing 1900 grams was delivered by vertex presentation; baby cried immediately after birth had good APGAR score, baby was shifted to mother side after initial assessment by neonatologist. Post operatively patient and baby both were stable. Patient was treated with the fluids to maintain the pulmonary artery hypertension and diuretics were avoided to prevent the fall in PAH, and to prevent the inferior venacaval collapse. The patient was started on Tab. Bosentan 62.5 mg OD and was discharged.

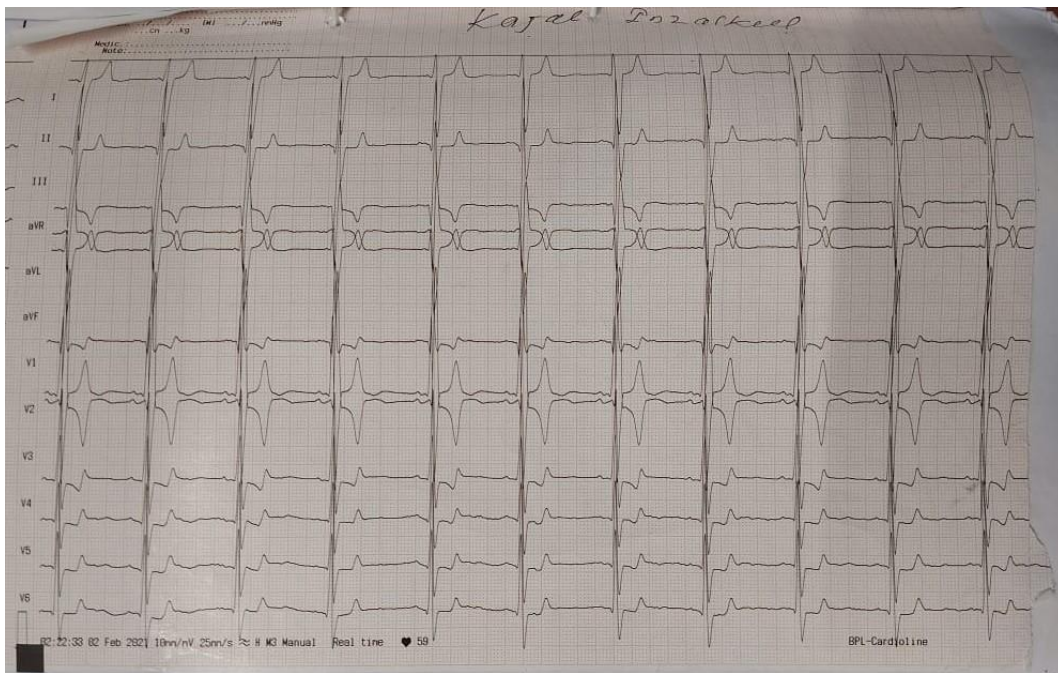


Figure 2 ECG showing: Right axis deviation, Right atrial enlargement and right ventricular enlargement with RV strain

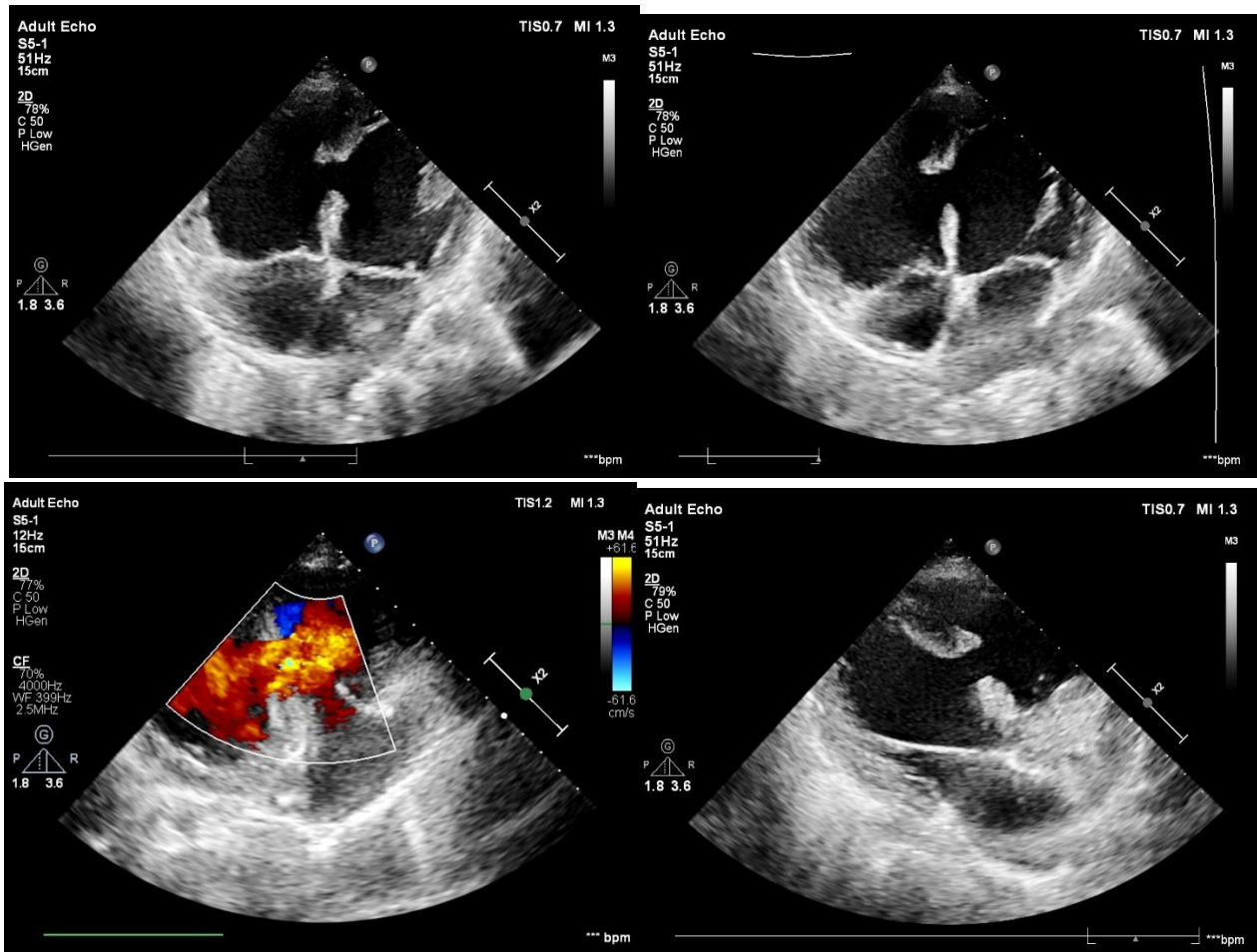


Figure 3 2d echo was done s/o VSD with severe pulmonary hypertension with Eisenmengerization

3. DISCUSSION

Eisenmenger syndrome is a condition that occurs as a complication of congenital heart defects like VSD, PDA, ASD. Chronic left to right shunts lead to increased pulmonary artery blood flow, resulting in gradual hyperkinetic pulmonary arterial hypertension (PAH) and reversal of shunt causing cyanosis and clubbing. Eisenmenger syndrome patients are vulnerable to hemodynamic changes like fall in systemic vascular resistance (SVR) and increase in right-to-left shunting that may result in circulatory collapse induced by anaesthesia or surgery. Other complications associated with surgery include excessive bleeding, deep vein thrombosis, postoperative arrhythmia and paradoxical emboli.

According to the study Yentis et al., (1998) maternal mortality was 40 percent and foetal death was 8 percent in the pregnant individuals with Eisenmenger syndrome. Another study Gleicher et al., (1979) revealed a 34 percent mortality rate for vaginal birth and a 75 percent mortality rate for Caesarean procedure. Seventy percent of deaths occur between days 2 and 30 after delivery or at the time of delivery (Brach et al., 1997). Normal blood volume increases by up to 50% during pregnancy, and increases even more during labour with each uterine contraction (Hegewald & Crapo, 2011). In persons with Eisenmenger syndrome, increased pulmonary artery pressure mixed with lower systemic resistance increases hypoxia. Because pregnancy is a prothrombotic state, it is not recommended in Eisenmenger syndrome. In these cases, pulmonary embolism and abrupt cardiac death can occur (Madden, 2009).

According to many researches, these patients have a high maternal and foetal mortality rate. Endothelial receptor blockers such as bosentan, ambrisentan, and macitentan (O'Callaghan et al., 2011) are currently more potent vasodilator medications. As these medicines are teratogenic (category X), they must be stopped before the patient becomes pregnant. If a patient is experiencing symptoms of heart failure, oxygen therapy and diuretics should be administered.

Vaginal delivery is the preferred method of delivery, with enough pain relief provided by intravenous analgesics or low-dose epidural anaesthetic. Anaesthesia must be administered with caution because hypotension can be fatal in these patients (Cunningham & Lindheimer, 1992). Vaginal birth has a number of advantages, including less blood loss, a lower chance of infection, and a lower risk of clot formation; however, it also has a number of drawbacks, including increased discomfort and tension, as well as increased sympathetic flow, which puts more strain on the ventricles.

4. CONCLUSION

More research and reporting of such cases need to be done, so that more physicians are made aware of these complications. It will help in early diagnosis and well as treatment and also help in reducing the mortality and in turn reduce the burden on the healthcare system.

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Author Contribution

SNB and SB initiated the idea of publication and contributed the intellectual content for the development of the manuscript. MR, KS, YT, GA, PR, TS, DS reviewed, proof read and edited the manuscript.

Informed consent

Written & Oral informed consent was obtained from all individual participants included in the study. Additional informed consent was obtained from all individual participants for whom identifying information is included in this manuscript.

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Conflicts of interest

The authors declare that there are no conflicts of interests.

Data and materials availability

All data associated with this study are present in the paper.

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