

CASE REPORT

Challenging Management of Pregnancy Complicated by Eisenmenger Syndrome; A Case Report

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Abstract:

Women with diagnosed case of Eisenmenger syndrome are usually advised to avoid pregnancy because of high mortality rate 30% to 50%, which increase up to 65% in case of caesarean section. Literature survey showed, few case reports on successful pregnancy outcome with proper multidisciplinary approach in tertiary hospital those who wanted to continue pregnancy with informed written consent about high mortality rate patient with Eisenmenger syndrome who become pregnant are at a particular risk of rapidly progressive cardiopulmonary decompensation, thrombotic complications and sudden death^{2,3}, because pregnancy related physiological changes are poorly tolerated in Eisenmenger syndrome. Prevention and early termination is advisable^{3,5}. For patient who present in later stages of pregnancy, carefully coordinated multidisciplinary care involving experienced specialists from obstetrics, anesthesiology, cardiology, neonatology and critical care medicine is necessary to optimize the chances of survival for both mother and baby. We here present a case report of a 25-year-old 2nd gravida, P:1(VD-IUD) admitted in Fetomaternal medicine department, BSMMU with 36 weeks pregnancy with uncorrected ventricular septal defect(VSD), Eisenmenger syndrome, severe pulmonary hypertension (94mmHg), left ventricular dysfunction (EF-<39%), Fetal growth retardation (Wt-1600gm), oligohydramnios (AFI-5.6cm), Gestational diabetes mellitus (on diet) with Hypothyroidism. By meticulous multidisciplinary approach Caesarean section done and shifted to Coronary care unit (CCU). She delivered a growth restricted (birth wt1500gm) male baby with Apgar score: 8 and managed in Neonatal care unit (NICU).

Key Words: Eisenmenger syndrome, Pulmonary hypertension, Ventricular septal defect, Multidisciplinary approach, pregnancy.

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Introduction:

Eisenmenger syndrome was so named by Dr. Victor Eisenmenger 1897, defined as pulmonary vascular obstructive disease that develops as a consequence of a large preexisting left to right shunt due to high pulmonary vascular resistance with reversal or bidirectional shunt.

A healthy heart and lungs got enough reserve power to cope up with increased load of hemodynamic changes during pregnancy. But in Eisenmenger syndrome with a damaged heart along with damaged lungs, fails to do so. Therefore, deterioration of cardiac function that is cardiac decompensation occurred during pregnancy. Patients with Eisenmenger syndrome has profound effect both for the

mother and fetus. Preterm delivery, Fetal growth restriction, fetal congenital heart disease (4.5% vs 0.6%), fetal death because of chronic hypoxia. Some patients with Eisenmenger syndrome may be able to go through pregnancy labor and delivery without major complications but sudden death in early postpartum period may occur due to decreased peripheral vascular resistance with right to left shunt and pulmonary embolism from silent illio-femoral thrombus. Pre pregnancy counselling, management during pregnancy and around delivery should be conducted in an expert center by a pregnancy heart team can minimize complications associated with this highly challenging pregnancy with Eisenmenger syndrome.

We here present a patient who referred from a local hospital at 36 weeks of pregnancy with uncorrected VSD, Eisenmenger syndrome, severe pulmonary hypertension, polycythemia, Fetal growth restriction, oligohydramnios, Gestational diabetes mellitus and hypothyroidism. She underwent Caesarean section with proper preoperative, peroperative and postoperative management by pregnancy heart team. Baby was managed in NICU.

Case Report:

A 25 -year-old 2nd gravida (para: 1 [VD]IUD+0) women got herself admitted in Fetomaternal medicine department, BSMMU, on 24th September, 2024, with non-corrective VSD with Eisenmenger syndrome with the complaints of slowly progressive dyspnea on exertion. Upon physical examination she had dyspnea on exertion with central and peripheral cyanosis, digital clubbing, blood pressure 220/80 mm of Hg and one plus peripheral oedema. Cardiovascular examination showed louder P2 and systolic murmur in pulmonary area, respiratory rate 20/min at rest. Per abdominal examination symphysial fundal height 30 weeks gestation, FHS : 144b/min and regular. Trans abdominal U.S.G revealed single fetus weight 1600 gm, AFI -5.6 cm

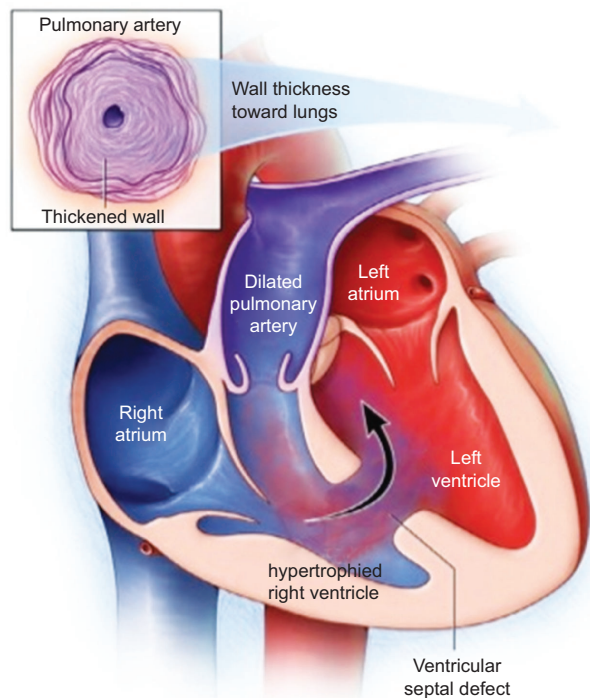


Figure 1: Pathophysiology of Eisenmenger

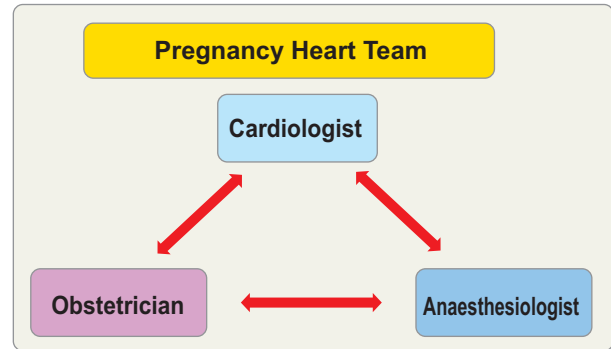


Figure 2: Pregnancy Heart Team

without any congenital anomaly. Echocardiography showed a large muscular ventricular septal defect 20mm in diameter, right to left shunting, pulmonary artery systolic pressure 94 mm of Hg, left ventricular ejection fraction <39% and dilated right ventricle. Therefore, multidisciplinary team consisting of obstetrician, anesthesiologist, cardiologist intensive care physician and neonatologist assigned to fully monitor the patient. Initially patient was kept in HDU with bed rest, oxygen administration by mask 5 L/min, sildenafil 50 mg 12 hourly, oral folic acid and calcium supplements. As patient admitted at 36 weeks with FGR, oligohydramnios and severe pulmonary hypertension Caesarean section under epidural anesthesia was decided to optimize patient's safety on 26th September. During surgery her initial oxygen saturation was 85po2 with 15L/min oxygen administration, but after delivery of the baby, oxygen saturation was 90pao2 with oxygen administration 5L/mi. Surgical procedure was uneventful and a male baby, weight 1500gm, Apgar score 8 was delivered and shifted to neonatal intensive Patient shifted to coronary care unit immediately after surgery. Patient condition comparatively remains stable after delivery. She was managed by Triple antibiotics, pain relieved by epidural analgesia and vasodilator Sildenafil 50mg 12 hourly and Levothyroxine 50mcg daily. On 3rd post operative day about 500 ml blood drained out by venesection to reduce hematocrit level. She was in Coronary care unit for one week and discharged on 8th postoperative day with oral antibiotic for 7 days, calcium, vitamins and Sildenafil 100mg. Given the risk of further decompensation, she was advised to resume regular close follow up after hospital discharge and to implement safe contraceptive method.

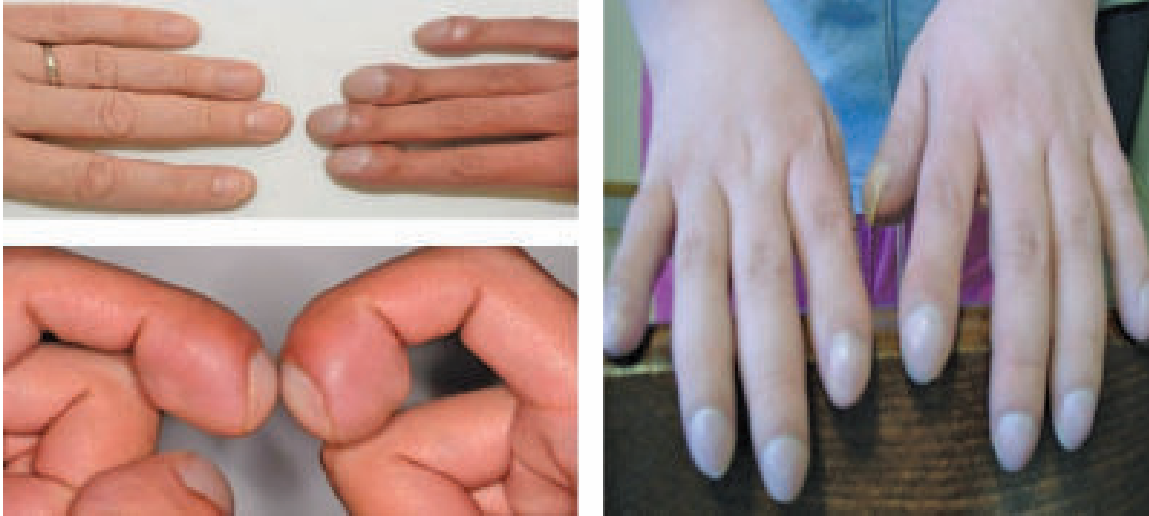


Figure 3: *Digital Clubbing*

Discussion:

Eisenmenger syndrome is a rare complication of congenital heart disease during pregnancy with a high mortality rate 30% to 50% and even up to 65% in these with Caesarean section. Within first few days of post-partum there is a high risk of sudden death may be due to hypovolemia, thromboembolism and eclampsia.^{3,4} The overall neonatal mortality 13% mainly due to prematurity. Pregnancy in women with uncorrected VSD with Eisenmenger syndrome presents significant maternal and fetal risks due to hemodynamic changes of pregnancy. Eisenmenger syndrome increases pulmonary vascular resistance, causing right to left shunt, leading to polycythemia, pregnancy further stresses the cardiovascular system, increasing the risk of heart failure, arrhythmia, thrombotic events and sudden death. The decision to proceed caesarean section under epidural anesthesia was based on the need to minimize maternal cardiovascular stress, also alleviates perioperative pain, reduces peripheral and systemic vascular resistance causing less tachycardia, less myocardial oxygen consumption and reduction of right to left shunt.⁴ General anesthesia could increase pulmonary vascular resistance and exacerbate right to left shunt, causing severe hypoxia and sudden cardiac decompensation.⁶ A multidisciplinary approach, involving collaboration, between obstetrician, cardiologist, anesthesiologist and neonatologist was crucial in ensuring safety both for the mother and baby. Now a days more pregnant women with Eisenmenger syndrome survive because of multidisciplinary approach in a tertiary Centre. The first postpartum week is considered a period of maximum mortality. Thus, intensive monitoring is necessary as most hemodynamic changes of pregnancy resolve by two weeks postpartum.^{5,6} Fetal outcome is

closely related to the hematocrit level. For a successful pregnancy outcome hematocrit should be less than 65% and the arterial oxygen saturation higher than 70%. When arterial oxygen saturation is maintained more than 90% in pregnancy with Eisenmenger syndrome, 92% of the fetus survive, but when oxygen saturation fell below 85% survival falls to 12%.¹ Though our patient referred at 36 weeks of pregnancy with uncorrected VSD, Eisenmenger syndrome, severe pulmonary hypertension, left ventricular dysfunction, polycythemia, FGR (Fetal Growth Restriction), prompt and proper management by pregnancy cardiac team saved both mother and baby.

Conclusion:

Termination of pregnancy is the safer option but when pregnancy continued, an expert pregnancy heart team can support the patient and optimize better outcome both for the mother and baby. The successful outcome in this case was due to meticulous preoperative planning, the use of epidural anesthesia to control hemodynamics and close post operative monitoring in Coronary care unit. A multidisciplinary approach is essential in managing high risk pregnancies complicated by congenital heart disease particularly in those with Eisenmenger syndrome, where the risk of maternal and neonatal mortality is high.

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