



RISKING LIFE TO HAVE A CHILD: THE WOMAN WITH EISENMENGER SYNDROME

Gynaecology

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ABSTRACT

Managing a pregnant woman with Eisenmenger syndrome is an Obstetric Challenge due to high maternal mortality. A 36 year old G4 known Eisenmenger Syndrome with previous 3 MTPs, was referred to our emergency services at 32+3 weeks of gestation (NYHA IV). She was married at 32 years of age and underwent first trimester MTPs 3 times. She was divorced due to her medical condition and was re-married a year ago and determined to have child.

Echo showed high muscular VSD measuring 1.6 cms with right to left shunt; RVSP was 50 mm Hg. She was managed in ICU with continuous Oxygen therapy and Sildenafil 25mg Furosemide 40 mg once daily. She had spontaneous preterm precipitate labour on Day 3 of admission, an alive preterm male baby of 2.2 kgs was born. She received inj Enoxaparin from 2nd postnatal day along with continuous Oxygen therapy till 34 th day in ICU.

KEYWORDS

Eisenmenger's syndrome, Preterm labour, Sildenafil, Continuous oxygentherapy, Extended hospital stay.

Background:

Eisenmengers Syndrome (ES) is defined as pulmonary hypertension as a result of uncorrected left to right shunt of ventricular septal defect, atrial septal defect or patent ductus arteriosus, with subsequent shunt reversal and cyanosis. Maternal mortality is as high as 40% in pregnancy and approaches to 70% with caesarean section.¹

Pregnancy is contraindicated in pregnant women with ES due to the physiological changes that occur in Cardiovascular, Pulmonary systems and haematological changes that have an adverse affect. If Pregnancy occurs it is recommended to perform Medical termination of pregnancy (MTP) to save the life of the woman and there are no clear cut guidelines regarding management of pregnancy and delivery

Case :

A 36 year old 4th gravida with previous 3 MTPs, known Eisenmenger Syndrome was referred to our emergency services at 32+3 weeks of gestation with breathlessness at rest and further management from a district hospital. On examination she was poorly built and nourished with a respiratory rate of 22 per minute SPO₂ was 85% on room air. Her pulse rate was 74 per minute regular and BP was 130/70 mm Hg. She had grade III clubbing and there was no cyanosis or icterus. On auscultation her respiratory system showed bilateral basal crepitations and CVS revealed systolic murmur in mitral area and a loud P2 in pulmonary area. Her cardiac functional status was NYHA IV.

Per abdominal examination revealed gravid uterus of 28 weeks size with single live fetus in cephalic presentation with reduced liquor. USG showed biometry corresponding to the gestational age and AFI was 6 cms. NST was reactive. She was admitted to Obstetric ICU and was immediate measures for stabilisation were started and monitored continuously. Cardiology evaluation was undertaken. Echo cardiogram showed high muscular VSD measuring 1.6 cms with right to left shunt; RVSP was 50 mm Hg and there was mild TR. ECG showed sinus rhythm with right ventricular strain pattern. Cardiologist advised to continue tab Sildenafil and Diuretics which she was taking since 5 years and advised Caesarean section. Her Xray chest revealed cardiomegaly. Her haemogram, LFT and RFT were normal. She was kept under continuous Oxygen therapy and received Sildenafil 25 mgs once daily and Lasix 40 mg once daily and was given antenatal corticosteroids for fetal lung maturity.

Her past history was that she was diagnosed as VSD at 5 years of age and was advised surgery but did not undergo surgery due to fear. At 15 years of age she suffered from haemoptysis and on complete cardiac evaluation she was told to have Eisenmenger syndrome and told that it was inoperable. She was married at 32 years of age and underwent first trimester MTP 3 times as advised by the Physicians. She was divorced due to her medical condition and was re-married a year ago and determined to have child.

She had spontaneous preterm precipitate labour on Day 3 of admission and was managed under caudal analgesia. She was delivered by outlet forceps to cut short the second stage of labour. An alive preterm male baby was born with APGAR of 8/10 at 5 min weighed 2.2 kg. She had mild PPH due to retained and adherent membranes and placental bits and minor cervical lacerations. These complications were duly recognised and managed with gentle evacuation and cervical laceration suturing. She received antibiotic prophylaxis. Baby was screened for CHD by ECHO and was normal.

She was kept on continuous Oxygen therapy and started on Enoxaparin from 2nd postnatal day and continued on Sildenafil and furosemide. Her tachypnoea settled after a week and she breastfed the baby. Her Oxygen saturation improved to 92 % at room air (Fig) after 2 weeks but fluctuated again between 80 to 85% due to URI. She was kept in ICU for 4 weeks and cardiac evaluation was sought again. She was advised to undergo cardiac catheterisation and patch closure of VSD but she differed. She was discharged on postnatal day 34 with advice to continue enoxaparin till 6 weeks postpartum. On follow up visit at 7 weeks (2 weeks after discharge) she and her baby were healthy and her functional status was NYHA II.

DISCUSSION:

Eisenmenger syndrome accounts for 8% of congenital heart diseases. The high mortality in Eisenmengers syndrome during pregnancy is due to the physiological changes of pregnancy of a decrease in systemic vascular resistance and increase in blood volume which cause right heart failure. With the fall in peripheral vascular resistance, the right to left shunt gets exacerbated and the pre-existing hypoxia worsens¹. Hence it is advised to defer pregnancy and first trimester MTP is advocated to save the life of the women with Eisenmengers syndrome. The present woman underwent MTP 3 times as per the advice of the Cardiologist. But not having a child has affected her family life and social life.

Mortality is also due to the hypercoagulable state of the pregnancy resulting in increased risk of thrombosis, pulmonary infarction and thromboembolism. Death usually occurs in the third trimester of pregnancy and in the first postpartum week and hence extended observation is necessary². The present woman was hospitalised for more than 4 weeks after delivery. The use of high flow oxygen is advocated by many and the administration of anticoagulation is still controversial. The prevalence of pulmonary arterial thrombosis is reported to be 21% and anticoagulation is advised in those at higher risk for the same³. Prophylactic anticoagulation to prevent thrombosis and thus death was advocated in few cases⁴. Cases are on record where omission of peripartum prophylactic anticoagulation resulted in development of Pulmonary thrombosis⁵. Maternal death was reported as late as 4 weeks. The present woman received enoxaparin for 6 weeks and is alive and healthy. The Pregnancy outcome includes

spontaneous abortion in 30% and preterm delivery in 60% apart from developing IUGR and IUFD . The fetus developing congenital heart defect is reported in 5% of the cases and in this case the neonate was normal and ECHO did not reveal any CHD. However she had precipitate preterm labour. In a large multicentric study, cause specific mortality in Eisenmenger syndrome was identified in 74% and the most common cause was heart failure followed by sepsis and thromboembolism.⁶ Though this patient underwent 3 MTPs because of the Eisenmengers syndromne , she tolerated the 4th pregnancy she tolerated pregnancy up to 32 weeks because of compliance to therapy (Sildenafil) and her RVSP was maintained at 50 mmHg.

CONCLUSION:

Continuous Oxygen therapy along with Sildenafil and anticoagulation in the postpartum period may have prevented thromboembolism and thus mortality in this woman with Eisenmenger Syndrome.

Figure legend;

Woman of Eisenmenger syndrome on continuous oxygen therapy and monitoring with the infant



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