



DIAGNOSIS OF EISENMENGER SYNDROME IN PREGNANCY: THERAPEUTIC CHALLENGES FACED DURING MANAGEMENT OF A COUPLE WITH PRECIOUS CONCEPTION

Abhay Kumar*

Junior Resident, Department of Obstetrics and Gynecology, PGIMER, Chandigarh*Corresponding Author

Avir Sarkar

Junior Resident, Department of Obstetrics and Gynecology, PGIMER, Chandigarh

ABSTRACT

Eisenmenger Syndrome poses a hemodynamic overload and additional burden for the already compromised right ventricle. It causes right to left shunt thereby exaggerating central cyanosis, leading to various sequelae in the form of abortion, preterm labor and intrauterine fetal growth restriction. Vasodilators in the form of intravenous prostacyclin, phosphodiesterase inhibitors like sildenafil and tadalafil and inhaled nitric oxide during second stage of labor have been successfully used. Pregnancy is ideally contraindicated. Those continuing pregnancy must be adequately supervised by an experienced multidisciplinary team. Termination of pregnancy is done by Caesarean. Epidural analgesia is preferred.

KEYWORDS : Eisenmenger Syndrome, vasodilators, pregnancy, cyanosis

Introduction

The hemodynamic overload in pregnancy poses additional burden for the already compromised right ventricle. The fall in systemic vascular resistance increases the right to left shunt thereby exaggerating central cyanosis, leading to various sequelae in the form of abortion, preterm labor and intrauterine fetal growth restriction. Proposed therapies aim at reducing pulmonary vascular resistance, thereby stabilizing right ventricular function. Vasodilators in the form of intravenous prostacyclin, phosphodiesterase inhibitors like sildenafil and tadalafil and inhaled nitric oxide during second stage of labor have been successfully used [1].

Case Report

A 33-year old primigravida with a married life of 9 years, presented to a tertiary care hospital at 25 weeks gestation with shortness of breath for 2 weeks. On careful elicitation of history, she was found to have breathlessness on exertion for 3 years, which has exaggerated for the past 2 weeks. On examination, she was conscious and well oriented, had a blood pressure of 144/100 mm of mercury, pulse rate of 74/min, respiratory rate of 24/min and room air saturation was 84%. There was clubbing and central cyanosis. On respiratory examination bilateral vesicular breath sounds were present with basal crepitation in left lower lobe. Cardiac examination revealed a raised jugular venous pressure and a pansystolic murmur. Uterine height was corresponding to 20 weeks period of gestation.

Investigations include a hemoglobin of 15.4 grams/dl and a haematocrit of 67%. Renal and liver function tests were normal. Electrocardiogram showed a right axis deviation. Chest X-ray was done which showed gross cardiomegaly. Echocardiogram was suggestive of large inlet VSD with bidirectional shunt and severe pulmonary hypertension with an ejection fraction of 55-60%, suggestive of Eisenmenger syndrome (ES). Ultrasound examination showed single intrauterine live fetus with parameters corresponding to 22 weeks and a fetal weight of 490 grams. Amniotic fluid index was within normal limits. Considering high chances of maternal mortality with pregnancy continuation, she was counselled for termination of pregnancy. Patient and relatives did not consent for pregnancy termination owing to primary infertility of 9 years.

Immediate admission was advised and a cardiologist consultation was sought. She was started on oxygen, diuretics like furosemide and phosphodiesterase inhibitors like sildenafil. Since saturation was persistently low on room air, she was shifted to high dependency unit and started on positive pressure ventilation (PPV). Oxygen by face mask, diuretics and sildenafil were continued. After a prolonged stay of 28 days, her general condition improved gradually. At 29 weeks gestation, she went into spontaneous onset of labour and delivered vaginally a baby girl of 450 g with Apgar 1 and 6. Baby required neonatal intensive care unit support with artificial respiration in the form of CPAP. Despite several efforts, baby succumbed to death on fifth day of life owing to prematurity. Lactation suppression was given. It was due to the combined effort of the multidisciplinary team that she gradually improved and was weaned of PPV. Finally, she was discharged after 10 days with an advice of strict use of barrier

contraceptive. She is under regular follow-up from cardiologist side.

Discussion

The incidence of ES in pregnant women is approximately 3% [2]. The underlying etiology consists of congenital valvular diseases like ventricular septal defect (VSD), atrial septal defect (ASD) and patent ductus arteriosus (PDA). Index patient had VSD with Eisenmenger. Maternal mortality usually high in VSD (60%) as compared to ASD or PDA [3]. The major cause of death includes hypovolemia, thromboembolism and pre-eclampsia [4].

Patients with ES are vulnerable to hemodynamic changes as even a minor change in systemic vascular resistance (SVR) may increase right-to-left shunt. In pregnancy, systemic vascular resistance decreases which in turn increases the right-to-left shunting of blood, subsequently leading to reduced pulmonary perfusion and hypoxia and further hemodynamic deterioration of mother and baby [4]. ES is associated with high risk of maternal morbidity and mortality. Considering the high mortality rate of around 50%, pregnancy is contraindicated in ES [5]. If a woman gets pregnant and wishes to continue pregnancy, oxygen therapy, aggressive use of pulmonary vasodilator therapies and care by multidisciplinary team may help to minimize mortality [6]. Poor prognostic signs include a maternal haematocrit of >60%, an arterial oxygen saturation of less than 80%, repeated syncope and right ventricular hypertension. A gradually escalating pulmonary artery hypertension with increasing gestation deems a grave prognosis with maternal and fetal mortality of 28% and 7% respectively.

Management strategy consists of oxygen therapy, diuretics, vasodilators and anticoagulants [7]. Oxygen works as a pulmonary vasodilator which decreases blood flow across the right-to-left shunt and thereby improving oxygen saturation [8]. All said and done, majority of these pregnancies end up in a preterm delivery (54.9 to 86%). Similar was the case in our patient also. Despite all possible measures taken to give the best care in an HDU set-up, yet pregnancy could not be prolonged beyond 29 weeks gestation. As many as 25% foetuses are growth restricted and similar was the case with our patient too. Moreover, haematocrit of 67% was after all an early indicator of adverse pregnancy outcome in our patient.

Being classified into World Health Organization (WHO) category 4 cardiac disorder of pregnancy, an elective caesarean delivery remains the predominant mode of pregnancy termination after attainment of adequate fetal pulmonary maturity. Promotion of valsalva would increase the right to left shunt thereby increasing cyanosis. However, case reports do exist in literature where atraumatic vaginal deliveries were conducted under epidural analgesia with prophylactic forceps application to deliberately cut short second stage of labour.

Conclusion

Considering the high maternal mortality and morbidity, pregnancy is ideally contraindicated in a diagnosed case of Eisenmenger Syndrome. Those continuing pregnancy must be adequately supervised by an experienced multidisciplinary team, including a senior obstetrician,

cardiologist, anaesthetic and neonatologist. Termination of pregnancy is done by Caesarean. Epidural analgesia is preferred. A short labour and an atraumatic delivery under pudendal or epidural block are preferred.

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