

CASE REPORT

Successful Pregnancy Outcome with Eisenmenger Syndrome

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Introduction

Pregnancy in the setting of pulmonary hypertension and Eisenmenger physiology is associated with a substantial maternal and fetal risk. Such patients are advised against pregnancy or to interrupt pregnancy before 10th gestational cut of week. Maternal mortality in the presence of Eisenmenger's syndrome (ES) is reported to be 30 to 50% [1, 2] and increases further with associated complications. When these patients decide to begin or continue a pregnancy against advice a coordinated multi-specialist care is mandatory. Overall fetal wastage is reported to be up to 75% [1]. We are presenting a woman with severe pulmonary hypertension due to Eisenmenger syndrome treated during pregnancy, delivery and postpartum.

Case Report

A patient Mrs. M. D. 20 years, PO+1, with past history of molar pregnancy 2 years back, attended our OPD at 10th week of gestation. Her LMP was on 12/11/2005. She had history of dyspnoea on exertion and occasional epistaxis and hemoptysis for last five years. She had an

echocardiography on 08/04/2004 which showed large non-restrictive perimembranous VSD with balanced shunt and significant pulmonary artery hypertension (PAH). During this period she was advised surgery but refused due to economic constraint. This time she was advised to terminate pregnancy but she refused. A cardiologic consultation was advised but she did not turn up. She again came back at 18th week of pregnancy and was admitted this time. USG on 31/03/2006 showed single live fetus of 18 weeks, 3 days (period of amenorrhea) 19 weeks, 5 days), no obvious congenital anomaly.

Echocardiography this time showed large VSD with bidirectional shunt, mean pulmonary artery pressure 50 mm of Hg. She was treated in consultation with cardiology department and advised bed rest, Iron and Calcium. Counseling about the prognosis was done. She was discharged on 21st week with repeated family request. She again got readmitted on 10/05/2006 at 25th week of gestation and same treatment was continued. USG on 27/06/2006 (33 weeks by LMP) showed 29 weeks, 6 days maturity (asymmetrical IUGR), grade-3 placenta, liquor adequate, EFW-1499gm, Doppler study within normal limit repeat.

Ecocardiography on 20/07/2006 showed an increased pulmonary artery pressure of 64 mm of Hg. Routine antenatal investigations were within normal limits Hb was 11.5 gm% on 12/05/2006 and 12 gm% on 08/07/2006. Hb and PCV were monitored repeatedly at 2 weeks interval to find early evidence of polycythemia. At 32nd week she had two bouts of hemoptysis and epistaxis on the same day,

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treated with antibiotics, sedation and diuretics. Patient became stable within two days. She was followed by weekly cardiotocography. She was normotensive during the antenatal period but had central cyanosis throughout the pregnancy.

Patient went into labor at 39th week on 13/08/2006. Cardiological review was done. She was treated with propped up position, oxygen inhalation, Inj. Pethidine 50 mg four hourly as analgesic. Inj. ampicillin 2 gm and Inj. gentamycin 80 mg were given as infective endocarditis prophylaxis in 1st stage and after delivery. Second stage was cut short by outlet forceps. A healthy baby weighing 2.4 kg was born. Patient was kept in hospital for two weeks with the continuation of same cardiac treatment and discharged in a good condition with a healthy baby. At 6th week follow up she was well with healthy baby, advised for barrier contraceptive and regular cardiac check up. At 3 month follow up she was well with no cardiological symptoms with healthy baby. She was advised for permanent sterilization after some time.

Discussions

The review of the world literature shows the very high maternal risk of this condition, which should be prevented by tubal sterilization or therapeutic abortion in very early pregnancy. A review of the literature on this subject revealed 115 reported cases, of which only 44 were felt to be adequately documented, representing 70 pregnancies. These formed the material for statistical evaluation. Fifty-two per cent of all patients died in connection with pregnancy, thirty and three-tenths per cent of all pregnancies resulted in maternal death. Maternal mortality in first, second, and third pregnancies was not significantly different. A high incidence of maternal death was associated with hypovolemia, thromboembolic phenomena and pre-eclampsia. Cesarean sections and other operations were associated with extremely high maternal mortality during pregnancy. Thirty-four per cent of all vaginal deliveries, three out of four cesarean sections, and only 1 out of 14 pregnancy interruptions (the only one by hysterotomy) resulted in maternal death. Abortions are significantly safer than any kind of delivery (*P* less than 0.05) [1].

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. Because of the high mortality associated with continuing pregnancy, medical termination of pregnancy is the preferred management for women with pulmonary hypertension of any etiology. For a patient who continues her pregnancy, hospitalization for the duration of pregnancy is often appropriate, in cyanotic heart disease of any etiology, fetal outcome correlates well with maternal haematocrit and successful pregnancy is unlikely with a haematocrit >65%. Third trimester fetal surveillance with ultrasound and ante-partum testing is important because at least 30% of the fetuses will be growth retarded. Kansaria [3] reported a case of ES in pregnancy where patient died 3 weeks after delivery.

Ganguly [4] reported three cases of ES in pregnancy in three years. Two of the 3 mothers and all 3 babies survived. One study shows that concomitant use of sildenafil and L-arginine for the management of pulmonary hypertension in pregnancy, combined with multidisciplinary care, permitted a good outcome for the mother and her infant [5]. Several studies done with heparin and vasodilators for the management of pregnancy with ES, revealed mixed response.

Conclusion

Although pregnancy should be discouraged in women with Eisenmenger's syndrome, we have demonstrated a case of successful pregnancy outcome in such a case.

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