University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2020, Vol. 6(3)

Successful pregnancy in Eisenmenger syndrome- a rare case report

SHWETHA S
Department of Obstetrics and Gynaecology, MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract : Eisenmenger syndrome is one of the very few heart diseases where pregnancy is absolutely contraindicated. Termination should ideally be offered to these patients before 10 weeks of gestation, as it carries a 50 percent maternal mortality rate, especially in the post partum period. Here I will be discussing a case of Eisenmenger syndrome in pregnancy who delivered successfully at our hospital. The patient had been an unevaluated case of heart disease since childhood, diagnosed for the first time as Eisenmenger syndrome at 19 years of age during a health camp. She only had occasional breathlessness on mild exertion for the past 2 years. She had been on irregular treatment with tablet bosentan and tablet sildenafil since then. On diagnosing pregnancy she was adviced termination but she refused. At 34 weeks she came to our hospital with breathessness for 5 days . She was managed symptomatically and stabilized. Fetus showed evidence of intra uterine growth restriction. Emergency caesarean section was performed in view of fetal distress. Postoperatively and after postpartum period she was well with a healthy baby.

Keyword : Eisenmenger syndrome, pregnancy, bosentan, sildenafil

INTRODUCTION:

Eisenmenger syndrome consists of pulmonary hypertension with reversed or bidirectional shunt at atrial, ventricular or aorto pulmonary level . Pregnancy should ideally be avoided in a patient with Eisenmenger syndrome because it carries a 50% risk of sudden death, especially few days post partum.

CASE:

22 year old Mrs.X, primigravida , belonging to socio economic class IV,with her last menstrual period on 26-10-13 and expected due date on 3-8-14, was a known case of a large atrial septal defect (ASD) with severe pulmonary hypertension (PHT) (Eisenmenger syndrome). She got admitted in intensive care unit on 2-7-14, at 35 weeks of gestation, with complaints of severe breathlessness for 5 days. At 4 years of age, patient was incidentally diagnosed to have heart disease during a routine school health check up . She was told to have a hole in the heart. She did not go for further evaluation Or treatment for the same, owing to financial constraints. At 19 years of age, in February 2012, during a health camp conducted by a private hospital, she had complained of occassional breathlessness for the past two years . She had no history of chest pain, syncope, epistaxis or hemoptysis in the past. Having given a history of 'hole in the heart' during childhood, she was admitted and evaluated for heart disease .

Echocardiography taken on February 2012 showed a large ostium secundum(OS) type of ASD ,bidirectional shunt , dilated right atrium(RA) and right ventricle(RV) ,grade 3 tricuspid regurgitation(TR) with severe PHT. Patient was started on tablet sildenafil citrate 25mg once a day . SILDENAFIL citrate (commonly called as viagra) is a phosphodiesterase 5 (PDE 5) inhibitor which degrades cyclic guanosine monophosphate (cGMP) .It is found in high concentrations in pulmonary arteries and corpora cavernosum. It was initially developed for treating erectile dysfunction. Normally, endothelium derived nitric oxide stimulates intracellular soluble guanyl cyclase , resulting in increased levels of cGMP, which then act to mediate smooth muscle relaxation. Sildenafil inhibits the degradation of cGMP by PDE 5 and prolongs the action of cGMP. Hence it acts by causing vasodilatation of pulmonary vascular bed. It belongs to Food and Drug Administration(FDA) category B medication. No reports of teratogenicity has been reported with its use in pregnancy. She was counselled regarding her condition, asked to continue her medications and then discharged.

In november 2012 ,she was admitted in the same private hospital for follow up and evaluation. Patient still complained of occasional exertional dyspnea. Echocardiography taken then, showed a large OS type of ASD, large left to right shunt,moderate PHT with normal biventricular systolic function . Patient was switched over to tablet bosentan 62.5mg twice a day and then discharged at request. BOSENTAN (generic name) is a competitive antagonist of endothelin 1 at the endothelin A and endothelin B receptors, thereby decreases the pulmonary vascular resistance. It is used in the treatment of pulmonary artery hypertension in patients with New York Heart Association (NYHA) class II-IV symptoms. It improves exercise capacity and decreases the rate of clinical worsening.

The side effects of this drug include hepatotoxicity and anemia. It belongs to FDA category X in pregnancy, as it is known to cause teratogenicity in animal studies. Hence, if possible ,it should be avoided in pregnant women in first trimester. The patient had no further consultation with cardiologist until pregnancy was diagnosed. At 21 years of age , pregnancy was diagnosed at 40 days of amenorrhea. She gave history of irregular treatment with tablet bosentan for the past one year .She hd no history of previous miscarriages. She was adviced to terminate the pregnancy but she refused. She did not go for regular consultations with the cardiologist . She was started on tablet sildenafil, tablet bosentan and tablet furosemide from the day her pregnancy was diagnosed . (Furosemide is loop diuretic used in the treatment of hypertension ,congestive heart failure and edema due to renal or hepatic failure where diuresis is required) .Patient took tablet bosentan irregularly, as she could not afford it. Her first trimester was uneventful . She came at 21 weeks for antenatal check up. Ultrasonography of the fetus showed a normal fetus corresponding to 20 weeks with no congenital anomalies. She came for monthly check up thereafter . She was adviced bed rest, iron and calcium tablets along with tablet bosentan , tablet sildenafil and tablet furosemide. At 26 weeks, she had an episode of breathlessness for which she was adviced admission . But patient had refused as she got symptomatically better .

Ultrasonography showed fetus corresponding to 24 weeks, interval growth was suggestive of intra uterine growth restriction, with normal liquor and grade 3 placenta. She was adviced admission at a tertiary care hospital until delivery , which the patient refused . On 30/6/14, at 34 weeks of gestation, patient developed severe breathlessness. Echocardiography showed a large OS type of ASD (2.5cm) with left to right shunt ,moderate to severe TR with severe PHT. Due to financial constraints, she did not get admitted in the private hospital where she had regular antenatal checkup and she came to our hospital for delivery. On admission, she was cyanosed,dyspneic and tachypneic. Oxygen saturation was 77percent at room air and 90 percent with nasal oxygen .She belonged to New York Heart Association(NYHA) class IV. Her investigations were as follows-arterial blood gas (ABG) pH -7.44,PaCO2- 31 mmHg,PaO2- 56mmHg , Hb-13.9mg/dl, PCV-40%. She was started on corticosteroids(Injection betamethasone 12mg intramuscular two doses given 24 hours apart) for fetal lung maturity. Cardiologist's opinion was obtained. Echocardiography was done and she was adviced to continue tablet bosentan and tablet sildenafil . Tablet spironolactone 25mg once a day (spironolactone is a potassium sparing diuretic used to treat edema and fluid retention due to heart failure, cirrhosis or nephrotic syndrome) and injection furosemide 20 mg intravenous once a day were added.USG obstetrics showed the fetus corresponding to 32 weeks of gestation(growth restricted) with normal liquor and normal doppler parameters. Patient was stabilized and was under observation in Intensive Care Unit.

A normal delivery had been planned for her and decision was made to wait till spontaneous onset of labour.On day 4 after admission, she suddenly developed an episode of syncope. Oxygen saturation(SpO2) was 50 percent at room air and 76 percent with continuous positive airway pressure(CPAP). She revived spontaneously but oxygen saturation was constantly around 75percent with CPAP. 6 hours after the episode, cardiotocograph started showing variable decelerations . 7 hours later, cardiotocograph showed persistent bradycardia with prolonged decelarations. Decision was made to deliver the baby by caesarean section, as her cervix was unfavourable for normal vaginal delivery and fetus showed signs of hypoxia.She was taken up for emergency caesarean section on 7-7-14, in view of fetal distress. Infective endocarditis prophylaxis, injection ampicillin 2g and injection gentamycin 80 mg intravenous, was given. Combined regional anaesthesia was chosen for this patient,as there can be diminished cardiac output with positive pressure ventilation and the use of halogenated compounds in general anaesthesia. Preterm boy of 2 kilogram ,with growth restriction and no signs of teratogenicity was delivered .Concurrent sterilization was done .Postoperatively, she was started on tablet furosemide 20mg once a day and tablet spironolactone 25mg once a day along with other drugs. Thromboprophylaxis ,injection low molecular weight heparin 40 mg once a day subcutaneously ,was given. Echocardiography taken on post operative day 10 showed a large ASD(30 mm) with bidirectional shunt,D shaped Left ventricle, dilated RA and RV with severe pulmonary hypertension.She was discharged on post operative day 14 . Patient was on tablet furosemide 20 mg once a day and tablet spironolactone 25 mg once day . 6 weeks after delivery she was well with a healthy baby.

DISCUSSION:A review of the literature on this subject revealed 115 reported cases[4]. In pregnancy, Eisenmenger syndrome is associated with approximately 30% to 70 % risk of sudden deaths. Eighty percent of these deaths occur between second and thirtieth postnatal day. *A high incidence of maternal death was associated with hypovolemia, thromboembolic phenomena and preeclampsia*. Cesarean section and other operations were associated with extremely high maternal mortality . Thirty-four per cent of all vaginal deliveries, seventy five percent of caesarean section, and 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) .[4] 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. 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 anaesthesia and can lead to arrhythmias, myocardial ischemia, ventricular failure and sudden death. There is increased incidence of pre eclampsia in these patients. The 15 year survival rate in non pregnant women is over 75 percent . Post operative fluid shifts associated with cesarean delivery pose an even greater risk, with mortality rates increasing to 70 percent. Fetal complications expected in such patients are preterm deliveries, miscarriages and intrauterine growth restriction. If maternal oxygen saturation is chronically less than 85% during pregnancy, babies usually die in utero before reaching a viable gestation. Incidence of congenital heart defects in babies is approximately 5%. ***In cyanotic heart disease of any etiology, fetal outcome correlates well with maternal haematocrit and successful pregnancy is unlikely with a haematocrit greater than 65*** percent . Third trimester fetal surveillance with ultrasound and antepartum testing is important, because at least 30% of the fetuses will be growth restricted.

Multidisciplinary team approach in a tertiary referral unit,equipped with both cardiology and anaesthetic staff, experienced in the management of adult complex congenital heart disease remains the gold standard of care for these patients. Ideally, a patient with Eisenmenger syndrome should be adviced termination of pregnancy in the first trimester, along with sterilization, after counselling about the risks(Our patient presented very late in

pregnancy, hence, therapeutic termination was not an option) If she decides to continue her pregnancy, heparin should be initiated early for thromboprohylaxis, 5000-10,000 units subcutaneously twice daily. Drugs used for pulmonary hypertension include nifedipine, sildenafil and bosentan. Early in third trimester, patient should be admitted in hospital. She should be adviced complete bedrest with oxygen therapy[upto fractional concentration of oxygen in inspired air of 0.4] . Frequent monitoring with pulse oximetry is recommended. Spontaneous onset of labour is preferred, to avoid the risk of caesarean section. During labour, oxygen flow is increased to 5-6 litres /minute. Oxygen saturation should be monitored continuosly. Arterial blood gas and blood pressure monitoring is a must. Inhaled nitric oxide or intravenous prostacyclin can be used in labour. These cause vasodilatation, improved oxygenation and prevents thromboembolism. Epidural analgesia is given at the onset of active phase of labour . Delivery should preferably be conducted in the left lateral position . Second stage of labour should be cut short using vacuum or forceps. In the post partum period, thromboprohylaxis should be continued. Rapid fluid shifts are possible after delivery, therefore close monitoring for the first 72 hours is a must. Pulmonary edema can develop in patients who don't have brisk spontaneous diuresis.

Hence ,controlled diuresis is adviced in the post partum period. Permanent sterilization is adviced as contraception in these patients. Depot progesterone may be considered as a temporary method of contraception to avoid anaesthesia and surgical risks in the immediate post partum period. There is very little experience with the combined use of bosentan and sildenafil in patients with pulmonary hypertension due to congenital heart disease in pregnancy. Despite its association with teratogenic effects in animal studies, bosentan was not associated with fetal malformations. So far, there has been only one case reported in Irish journal, where a 27 weeks pregnancy with Eisenmenger syndrome was managed with Bosentan and sildenafil.[1]

CONCLUSION: Eisenmenger syndrome is one of the diseases where pregnancy is absolutely contraindicated, as it carries a high maternal mortality along with a high incidence of fetal wastage. Hence , termination of pregnancy with sterilization should be insisted in all patients. In developing countries like India, we still encounter patients with severe heart disease, who have poor compliance to treatment .These patients may refuse termination and come to the hospital with complications in advanced pregnancy . Management of such cases should always be a multidisciplinary approach. Drugs like bosentan, sildenafil and other pulmonary vasodilators can be used to decrease the pulmonary arterial pressure and improve the maternal and fetal outcome in these patients.

REFERENCES:

1.Eisenmenger's syndrome in a 27 week pregnancy--management with bosentan and sildenafil. Molelekwa V, Akhter P, McKenna P, Bowen M, Walsh K. Ir Med J. 2005 Mar;98(3):87-8

2.Management of Eisenmenger syndrome in pregnancy with sildenafil and L-arginine. Journal of obstetrics and gynecology 2004 May;103(5 Pt 2):1118-20. Lacassie HJ1, Germain AM, Valdés G, Fernández MS, Allamand F, López H

3.Successful pregnancy outcome in Eisenmenger syndrome published in journal of obstetrics and gynecology of India feb 2012 62(1), 68-69 Partha Mukhopadhyav, Popli Bhattacharya, and Nilofur Begum

4. Eisenmenger's Syndrome and Pregnancy. Obstetrical & Gynecological Survey:October 1979 Gleicher, Norbert; Midwall, Jay; Hochberger, David; Jaffin, Herbert

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities