(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 4, Issue 4, Page No: 1009-1012
July-August 2021

IJMSCR

Successful Pregnancy Outcome in a Patient with Eisenmenger's Syndrome

[1]Dr Sarita Agrawal, [2]Dr sefali shinde, [3]Dr Sagarika Majumdar, [4]Dr Pushpawati, [5]Dr Sarita Rajbhar, [6]Dr Taru

[1]HOD, [2]Senior Resident, [3,5]Assistant Professor, [4]Associate professor, [6]Junior resident
[1,2,3,5,6,7]Departoent of Obstetries & Gynecology, AlIMS Raipur
[4] Dr. Vasantrao Pavwar Medical College Hospital & Researeh Centre

***Corresponding Author:**
Dr. Sefali Shinde
Senior Resident department of gynaecology All India institute of medical science Raipur

Type of Publication: Original Research Paper
Conflicts of Interest: Nil

Abstract

Pregnancy with Eisenmenger syndrome is a high risk fetomaternal condition. Women with Eisenmenger syndrome are medically advised to avoid pregnancy. Maternal mortality associated with Eisenmenger syndrome is reported to be 30-50% and even up to 65% in those with Cesarean section. Therefore, pregnancy is not advisable in such patients. Co-ordinated multi specialist care is required in management of such patients. For a patient who continues her pregnancy, should be hospitalized as early as 20 weeks of gestation, depending on the cardiac condition. Herein, we report a 27-year-old gravid three parity one early neonatal death one, abortion one admitted to our hospital at 26 weeks of gestation. After a multidisciplinary approach in conjunction with obstetrician, physician and cardiologist, the patient was diagnosed to have Eisenmenger syndrome, NYHA grade II. She was managed conservatively and eventually delivered a preterm baby vaginally with a good outcome.

Keywords: Eisenmenger Syndrome, Pulmonary Artery Hypertension

INTRODUCTION

Clinical Diagnosis

A 27yrs gravid three parity one, early neonatal death one& abortion one came to our outpatient clinic for the first time at 26 weeks 3 days. Her past obstetric history revealed that during her first delivery at some private hospital 3 year back, she had an episode of breathlessness, cyanosis and cough after delivery of preterm baby with preterm prelabour rupture of membranes (PPROM). The baby died after 8 days of birth because of prematurity and neonatal jaundice. She was diagnosed with heart disease; however, her previous medical records could not be obtained. She had history of one spontaneous abortion in March 2019 at two and half month of gestation, and it was medically managed. No history of any cardiac symptoms in between the pregnancy. There was no history of heart disease in the family.

Differential diagnosis

In this pregnancy her first trimester was uneventful had no antenatal checkups. She first time present in our OBGY OPD at gestation age of 26 weeks 3 days with complaint of breathlessness on exertion no other complaint of chest pain and palpitations. On examination patient was of average built grade-1 pedal edema and clubbing was present. Her oxygen saturation (SPO2) was 88% at room air with respiratory rate-22/min and JVP was not raised. Thyroid and breast examination revealed no abnormality. On CVS examination, S1S2 was regular and a pansystolic murmur was heard with loud P2, bilateral vesicular breath sounds was heard throughout the lung field. Obstetric examination suggested 26 weeks gravid uterus, relaxed with cephalic presentation with fetal heart 130 beats/min. On

account of her history of cyanosis breathlessness in previous pregnancy and auscultation findings of murmur, cardiology opinion was taken and 2D echo was performed which revealed large VSD 23.2 mm, mild MR, mild TR, supra systemic pulmonary artery hypertension and diagnosis of Eisenmenger's syndrome with NYHA grade –II was made. Other investigations complete blood counts, liver function test and renal function tests were within normal limits. Her obstetric ultrasound report suggested normal growth scan with adequate liquor and normal Doppler with estimated fetal weight of 1. 7kg.Anomaly scan and fetal echo were normal. Venous Doppler was performed in view of right limb pedal edema which was normal not suggestive of any thrombosis. Patient was counseled about involved maternal and fetal risks and she was kept on conservative management with cardiology supervision. Patient was kept in propped up position with oxygen supplementation 10-12 liters of oxygen per minute to keep SPO2 >90%. Low molecular weight heparin (40 mg) subcuteneous once a day was started and dexamethasone 6mg IV 12 hourlies of total 4 doses for lung maturation was given. Other routine antenatal medication and CBC, electrolyte, fetal biophysical profile twice weekly performed. During the hospital stay her oxygen levels were fluctuating with episodes of breathlessness managed with regulating oxygen flow rates. At 30weeks in view of expected preterm delivery rescue dose of injection dexamethasone along with neuro protection dose of magnesium sulphate was given after checking of serum magnesium level.

Discussion of Management

At 31 weeks of gestation the patient went in spontaneous onset of preterm labour and deteriorated with fall in oxygen saturation to less than 86%. During labour she was kept in a propped-up position with high flow oxygen at 12 liters. Labour monitored with partograph and continuous intrapartum cardiotocography. Cardiologist, anesthesiologist and neonatologist were informed. Labour progressed well and she delivered a preterm alive male child of 1.720kg with APGAR score 8. Baby was shifted to NICU (Neonatal Intensive Care Unit) in view of prematurity. Injection Furesemide5 mg IV was given After delivery as per advised by anesthetist, oxytocin infusion was continued for one hour to prevent any possibility of PPH. Oxygen inhalation at 12 L/min was continued with continuation of antibiotics. Close monitoring done during postpartum period. Slow IV fluids at rate of 75ml/min were given for 24 hours. Baby was kept for observation for 10 days in NICU then shifted to mother side. Patient gradually improved and was discharged on day17 postpartum with the advice of strict use of barrier contraception to avoid future pregnancy and advice to attend cardiologist for follow up.

Pathological Discussion

ES is a triad of large anatomical cardiac defects, pulmonary arterial disease and cyanosis. The development of ES may accompany a variety of forms of CHD out of which commonest are VSD followed by ASD and PDA. The term was first clinically described by Viennese physician Victor Eisenmenger in 1897. Women with ES are advised to avoid pregnancy, or an early pregnant termination is preferably indicated within10th gestational week.[5] For a patient who choose to continue her pregnancy, should be advised early hospitalization in the second trimester itself, dependent on the cardiac condition.

Eisenmenger syndrome patients are particularly susceptible to hemodynamic changes induced by anesthesia or surgery, and even minor decrease in systemic vascular resistance (SVR) may increase the right-to-left shunting subsequently leading to a reduced pulmonary perfusion and hypoxia and further deterioration of mother and baby. Maternal mortality in the presence of ES is reported to be 30-50% and even up to 65% in those with Cesarean section.[6]

In a study by Yentis et al, the maternal mortality was 40% and fetal demise was 8%, and only 15% of infants were born at term. It is higher when associated with VSD (60%) than with ASD (44%) or with PDA (41.7%).[7] It has been seen in studies that 34% mortality is associated with vaginal delivery and a 75% mortality is associated with Cesarean section. It is reported that the most dangerous period is immediate postpartum, with 70% of deaths occurring on postpartum days 2-30 or died just at the time of delivery.[8]

Diagnosis of ES requires the presence of congenital heart disease (CHD). Sometimes, the diagnosis is not established until adulthood, after the development of symptoms or even overt features of pulmonary hypertension such as syncope, atrial or ventricular

Volume 4, Issue 4; July-August 2021; Page No 1009-1012

arrhythmias, cyanosis, and as late findings, both right and left heart failure.[9]

Evaluation for Eisenmenger syndrome should include physical examination followed by noninvasive assessment of cardiopulmonary anatomy and function failure by pulse oximetry, chest X ray, ECG, and imaging studies like echocardiography, CT angiography. The routine insertion of pulmonary artery catheters is contraindicated in Eisenmenger syndrome.These catheters provide no useful data, have a high potential for hemorrhagic and other complications, and do not improve maternal outcome(9).The most appropriate and cost effective study for patients suspected to have PAH is echocardiogram, with complete right heart catheterization for confirmation but in pregnancy invasive method is not recommended for evaluation(10).Close monitoring of pregnancy and supportive care is the key to success in ES.As the circulatory load peaks during 30-32 weeks of gestation, there is high chance of preterm labour ,PPROM(preterm prelabor rupture of membrane), or fetal growth retardation in the fetus . During episodes of dyspnoea, bed rest is recommended, and high concentrations of oxygen should be administered through a face mask.[4] Oxygen is a pulmonary vasodilator, which decreases the blood flow across the right to left shunt and thereby improves oxygen saturation.[11] The oxygen tension should be accessed through determination of serial arterial blood gases to detect changes in shunt flow.[4] Sildenafil is a phosphodiesterase type 5 inhibitor through its action on the NO/CGMP signal pathway, causes a selective fall in pulmonary vascular resistance and improves symptoms and exercise capacity of patients with pulmonary hypertension.[12] Oral sildenafil at a dose of up to 50 mg four times per day has been shown to improve both hemodynamic and oxygenation in patients with PAH(13).However, debates remain in the prophylactic anticoagulant therapy in the peripartum period. Admittedly, heparin use may prevent thromboembolic complications, but subsequent bleeding has been reported with significant blood loss and transient vital sign drop that warrants cessation of heparin and aggressive transfusion in postpartum(14) .Third trimester fetal surveillance with ultrasound and antepartum testing is important because at least 30% of the fetuses in these cases are having growth retardation.[15] In anticipated preterm birth baby, antenatal steroids for fetal lung maturity is warranted.

Mode of delivery in ES patients with PAH is controversial. Compared to cesarean delivery, although vaginal delivery is associated with less risk of haemorrhage, infection and venous thromboembolism, vaginal delivery leads to increased basal cardiac output and increased output with every uterine contraction, which would promote cardiac arrhythmia and worsen heart failure. Furthermore, ES patients with PAH should be delivered early due to serious disease condition with unfavorable cervix and labour induction is likely to be difficult in these patients.[3]

The goal of anaesthetist management is to maintain systemic vascular resistance in order to prevent an increase in right to left shunt. Slow-titrated epidural anaesthesia can be a safe mode of anaesthesia for Caesarean section in pregnancy with ES.[16]

If the cervix is favorable and vaginal delivery is planned, labor could also be induced with good pain control, usually with epidural anaesthesia. Labor are often conducted with the mother within the left lateral position to avoid inferior vena caval compression and maintain venous return. Bearing down (Valsalva maneuver) is often best avoided in patients with complex CHD, but the approach must be individualized.[17] The second stage can be cut short with forceps or vacuum extraction if necessary. Prolonged labor should be avoided.

In addition to fetal monitoring, maternal ECG monitoring should be performed to detect any arrhythmia during labor and therefore the puerperium. Intravenous lines, and if necessary central blood pressure catheters, facilitate the stabilization of hemodynamics by monitoring fluid shifts. Continuous pulse oximetry is beneficial, and in some circumstances (cyanotic heart disease) intra-arterial monitors may detect early changes in oxygen saturation and pressure in preference to automated vital sign monitoring. (18) The patient should be kept in bed for the first day after delivery and should be monitored continuously and then gradually mobilized. Studies suggest that the patient should stay in hospital for a period of 7-14 days after delivery under careful observation due to the continued risk of sudden death.[4]

Volume 4, Issue 4; July-August 2021; Page No 1009-1012

The distinctive feature in this case is that despite having an ES with associated large VSD and supra systemic PAH, our patient continued her pregnancy and delivered vaginally without any postpartum complications.

Final Diagnosis

Pregnancy should be preferably avoided in a woman with eisenmenger syndrome because of a high maternal mortality rate and poor outcome *of* baby. But it can be victorious as seen in our case wherein co-ordinated and experienced multi-specialty care and proper monitoring helped in avoiding any adverse maternal and fetal outcome.

REFERENCES

1. Wood P. The Eisenmenger syndrome or pulmonary hypertension with reversed central shunt. I. Br Med J 1958; 2(5098): 701-9. /;LO9`
2. Shinji Katsurahgi ,ChizukoKamiya.Maternal and fetal outcomes in pregnancy complicated with Eisenmenger syndrome.Taiwanese Journal of Obstetrics& Gynecology 58 (2019) 183e187
3. Duan et al. Pregnancy outcome in women with Eisenmenger's syndrome BMC Pregnancy and Childbirth (2016) 16:356
4. Mikael Bitsch, Christoffer Johansen, Alf Wennevold and Mogens Osler Eisenmenger's syndrome and pregnancy Eur. J. Obstet. Gynecol. Reprod. BioL, 28 (1988) 69-74
5. Mukhopadhyav P, Bhattacharya P, Begum N. Successful pregnancy outcome with Eisenmenger syndrome. *J ObstetGynaecol India*. 2012;62(1):68–69
6. Shi-Min Yuan Eisenmenger Syndrome in Pregnancy *Braz J CardiovascSurg 2016;31(4):325-9*
7. SubulBazmiet. al Pregnancy with Eisenmenger Syndrome International Journal of Obstetrics and Gynaecology Research (IJOGR) Vol. 2 (2015) No.2, pp. 151-154
8. Yentis SM, Steer PJ, Plaat F. Eisenmenger's syndrome in pregnancy: maternal and fetal mortality in the 1990s. Br J ObstetGynaecol. 1998;105(8):921-2.
9. Carole A. Warnes et al.ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary,, Volume 118, Issue 23, 2 December 2008, Pages 2395-2451.
10. Vongpatanasin W, Brickner ME, Hillis LD, Lange RA. The Eisenmenger syndrome in adults. Ann Intern Med. 1998 May 1;128(9):745-55. doi: 10.7326/0003-4819-128-9-199805010-00008. PMID: 9556469.
11. Srikanth JK, Gupta N, Chakrabarti S, Ish P. Eisenmenger syndrome with pregnancy – Double trouble. Indian J Med Spec 2020; 11:44-6.
12. Barnett and Machado. Sildenafil in the treatment of pulmonary hypertensionVascular. Health and Risk Management 2006:2(4) 411–422.
13. Reinalyn S Cartago. Pregnancy outcomes in patients with severe pulmonary hypertension and Eisenmenger syndrome treated with sildenafil monotherapy. Obstetric Medicine 2014, Vol. 7(1) 40–42.
14. Pitts JA, Crosby WM, Basta LL. Eisenmenger's syndrome in pregnancy. Am Heart J 1977; 93:321-325.
15. SetuRathod, Sunil KumarSamal. Successful Pregnancy Outcome in A Case of EisenmengerSyndrome.jcdr 2014 Oct, Vol-8(10): OD08-OD09.
16. SugataDasgupta, Soumi Das, BiswajitMajumdar& SM Basu (2016) Caesarean section in Eisenmenger's syndrome: anaesthetic management with titrated epidural and nebulisedalprostadil, Southern African Journal of Anaesthesia and Analgesia, 2016 ,22:2, 65-67,
17. MatthiasGreutmann and Petronella G. Pieper. Pregnancy in women with congenital heart disease. European Heart Journal (2015) 36, 2491–2499.
18. Carole A. Warnes, MDPregnancy and Delivery in Women with Congenital Heart Disease.Official Journal of the Japanese Circulation Society 2015
19. Basit H, Wallen TJ, Sergent BN. Eisenmenger Syndrome. [Updated 2020 Jun 30]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020.

Volume 4, Issue 4; July-August 2021; Page No 1009-1012