International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Eisenmenger Syndrome - The Worst Anticipated Nightmare in Pregnancy

Sanket Mahajan, Dhanesh Mhaskar

Resident Doctor, Department of Medicine, Krishna Institute of Medical Sciences University, Karad, Satara, Maharashtra, India.

Corresponding Author: Sanket Mahajan

Received: 05/09/2015 *Revised: 28/09/2015* *Accepted: 05/10/2015*

ABSTRACT

Maternal mortality in presence of Eisenmenger syndrome is reported to be 30% to 60% and increases further with associated complication. We report a case of Eisenmenger from our hospital who delivered a neonate without any complications to the mother and her baby. We will hereby discuss, in detail, about the pathophysiology of Eisenmenger Syndrome during each stage of pregnancy along with its complications and management in order to improve the maternal and foetal health in terms of prognosis.

Keywords: Eisenmenger Syndrome, Pregnancy, Maternal Mortality.

Key message: Our case report will help in understanding the course of disease throughout the different stages of pregnancy and its anticipated complications and their management and this in turn will lead to better prognosis of such a dreaded condition.

INTRODUCTION

Eisenmenger syndrome is defined as the development of pulmonary hypertension in response to a left-to-right cardiac shunt with consequent bidirectional or reversal (right-to-left) of shunt flow. Initially, left-to-right intracardiac shunting is associated with increased flow (and sometimes transmitted pressure) through the pulmonary vasculature. This results in pulmonary vascular remodelling and leads to pulmonary vascular disease. Eisenmenger's syndrome is defined as the process in which a left to right shunt caused by a congenital heart defect in the fetal heart causes increased flow through the pulmonary vasculature, causing pulmonary hypertension, [1] which in turn causes increased pressures in the right side of the heart and reversal of the shunt into a right-to-left shunt. The pulmonary arterial hypertension and associated elevation in right heart pressures result in reversal of the shunt with either right-to-left or bidirectional flow, which is called Eisenmenger syndrome.

Congenital heart defects that can lead to Eisenmenger syndrome include: atrial septal defects, ventricular septal defects, persistent arterial ducts, as well as more complex defects such as atrioventricular septal defects, truncus arteriosus, aortopulmonary window, complex pulmonary atresia, and the univentricular heart.

As a result of the right-to-left shunt, patients are chronically hypoxemic, hence cyanotic. Eisenmenger syndrome is associated with complications in many systems and is considered a multi-system disorder, mainly during pregnancy.

Maternal mortality is high. Fetal mortality is 55 to 75%. Deaths occur either during or within the first weeks after delivery. [2] Medical termination of pregnancy is preferred management for women with Eisenmenger syndrome who present early in pregnancy.

CASE REPORT

A 25 year old primigravida unregistered case presented to obstetric ward with history of 8 month old amenorrhea with NYHA grade 2 dyspnoea initially which progressed to NYHA grade 4 dyspnoea. She had history of syncope and edema feet for last three days. On examination she was tachypneic and had central cyanosis with SpO_2 of 70% on room air. There was no clubbing. Pulse rate was 100 per min. Blood pressure was 110/70 mmHg. Her JVP was raised. Obstetric examination revealed 8 month pregnancy and normal fetal heart sounds.

Medicine consult was sought where we found, on cardiovascular examination, atypical impulse in 5^{th} intercostal space, no precordial bulge, no pulmonary artery pulsation, apical impulse was tapping, no parasternal heave, palpable second heart sound and a systolic thrill was felt in left parasternal area. Patient was having systolic murmur in mitral, tricuspid, aortic and pulmonary areas of grade 3/4. Second heart sound was loud and had wide fixed splitting. Respiratory system revealed bilateral basal rales. CXR revealed cardiomegaly. ECG showed RVH. Suspecting ASD, a 2D-Echo was advised which was suggestive of large 18mm atrio-ventricular septal defect with biventricular flow noticed across VSD. A large 28mm ostium primum atrial septal defect (Figure 1) was noticed with predominantly right to left flow across atrial septal defect and estimated pulmonary artery systolic pressure of 102 mm of Hg.

Patient was started on continuous oxygen inhalation, digoxin and diuretics. A cardiology consult was sought where it was adviced to continue digoxin and diuretics. Patient was taken for elective, high risk caesarian section which turned out to be uneventful. She was advised further cardiac evaluation and intervention but due to financial restraints, she was managed conservatively. Patient delivered a healthy newborn without any intrapartum complications and was discharged after 39 days of hospital stay which was uneventful. Patient was followed up for 9 months after discharge from hospital where she was always totally asymptomatic and her child was always healthy. Patient was advised not to go for further pregnancies as that could be devastating in the future.

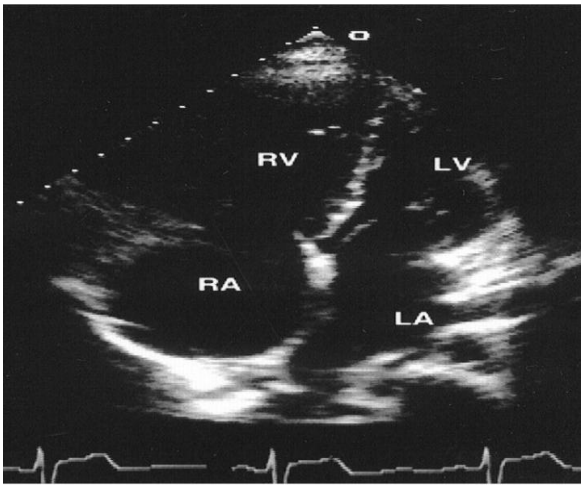

Figure 1: Diagram showing ASD in a patient with Eisenmenger syndrome

DISCUSSION

Effects of Pregnancy-Related Hemodynamic Changes: The hemodynamic changes of pregnancy are usually poorly tolerated in women with Eisenmenger syndrome. Most women with Eisenmenger syndrome are in a precariously balanced state and an important principle of care is to not disrupt this balance. In women with Eisenmenger syndrome and a low cardiac output state, the compromised right ventricle may not meet the demands of increasing blood volume and cardiac output associated with pregnancy. In addition, a fixed pulmonary

vascular resistance with a resulting inability to increase pulmonary blood flow may not accommodate an increase in cardiac output. Similarly, large fluctuations in blood volume both pre and post partum may not be tolerated by an already compromised cardiovascular system. The fall in peripheral vascular resistance that occurs during pregnancy can augment right-to-left shunting, worsening maternal hypoxemia and cyanosis. [3]

During pregnancy, the blood becomes more hypercoagulable and in the cyanotic patient the risk of deep venous thrombosis, pulmonary infarction, and/or paradoxic embolus and stroke increases.

Cardiac complications include supraventricular arrhythmias, ventricular arrhythmias, congestive heart failure, progressive valvular disease, sudden death. Non-cardiac complications include bleeding (pulmonary hemorrhage, gastrointestinal, cerebral), ischaemic complications (thromboembolic events, paradoxic emboli, air embolism), renal dysfunction, symptoms related to hyperviscosity (headache, dizziness, visual disturbances, altered mentation, tinnitus, fatigue), iron deficiency (due to inappropriate phlebotomy for secondary erythrocytosis), pulmonary arterial dilation, infections (endocarditis, cerebral abcess, pneumonia), gout, hypertrophic pulmonary osteoarthropathy.

Maternal Complications: Pregnancy is a cause of significant mortality in most published series of women with Eisenmenger syndrome. A systematic review of published studies from 1978-1996 examined maternal mortality rates in women with Eisenmenger syndrome and demonstrated mortality rates of 56%. [4] A more recent review suggested that mortality remains high. [5] Most complications occur near term and early (1st week) post-partum, and therefore extended post-partum hospital observation is suggested. Mortality is typically from heart failure, sudden death presumably due to arrhythmias, or thromboembolic events.

During pregnancy, it is important to watch for cardiac symptoms including increasing fatigue, worsening peripheral edema, palpitations, chest pain that could reflect right ventricular ischemia, and/or volume overload and presyncope/syncope with exertion reflecting a decrease in cardiac output. However, other complications as described can also occur, particularly thromboembolism.

Fetal Complications: Miscarriage is common in cyanotic women. Intrauterine growth restriction is seen in 30% of pregnancies as a result of maternal hypoxemia. Premature labour is found in 50-60% of instances and the high perinatal mortality rate (28%) is due mostly to prematurity. In one study of women with Eisenmenger syndrome, 47% delivered at term, 33% between 32 and 36 weeks, and 20% before 31 weeks of gestation. [4]

Management strategies:

Preconceptional Counselling: Based on the high mortality risk both during pregnancy and peripartum, women with Eisenmenger syndrome should be strongly advised against pregnancy. [4-6] Some women who are fully informed and understand the maternal and fetal risk and complications may still become pregnant, and unfortunately women may present pregnant without having received appropriate preconceptual counseling.

Currently many women with Eisenmenger syndrome are treated with pulmonary vasodilators. A discussion about contraceptive methods is imperative. [6] Progesterone-only formulations as depot injections and sub dermal implants are a reasonable option. Contraceptive pills containing estrogen (combined contraceptive pills) are contraindicated due to an increased risk of thromboembolism. [4] The insertion of intra-uterine contraceptive devices can be associated with vasovagal reactions, which can be devastating in women with pulmonary

hypertension. Moreover, the laparoscopic procedure carries risk in this population, as it requires insufflation of the abdomen with carbon dioxide, intermittent head down tilt and positive pressure ventilation, all of which reduce cardiac output and may be poorly tolerated. There is also a risk of air embolism, which may pass through the shunt to the brain (paradoxical embolism) in face of a right to left shunt. Transmission of congenital heart disease to offspring should be discussed. The risk of the fetus having structural cardiac defects varies between 3% and 50%, compared with the background risk of 1% for the general population. This risk will depend on the underlying cardiac lesion of the mother.

Ante-Partum Care: If a woman with Eisenmenger syndrome becomes pregnant, coordinated care should be established early, involving a congenital heart disease specialist, pulmonary hypertension specialist, high-risk obstetrician, and an obstetrical anaesthetist. Close cardiovascular monitoring, with specific attention to volume status, is essential throughout pregnancy and the peripartum period. Serial echocardiograms are important to assess the size and function of the right ventricle. Some experts use serial BNP-levels. Hypovolemia can lead to increased right to left shunting, reduced cardiac output and refractory hypoxemia. Similarly, volume overload should also be avoided as it cannot be accommodated by the compromised pulmonary vascular bed and/or right ventricle and can result in heart failure and increasing right to left shunt. While no pulmonary vasodilators are considered completely safe during pregnancy, there are case reports/case series in the literature describing the use of pulmonary vasodilators during pregnancy. [7-9] However, use of bosentan is not advised in pregnancy owing to the teratogenic effects seen in animal studies. Bed rest is must to reduce cardiac demands. Treatment for heart failure may be required prophylactically. These women are also vulnerable to thromboembolism. Thromboprophylaxis is essential at any time that the woman is relatively immobile. An appropriate anticoagulation plan should be devised with a hematologist/thrombosis expert when required. [5]

Fetal echocardiography can be offered to the expectant mother to screen for congenital heart defects. A fetal echocardiogram is done at approximately 20 weeks gestation.

Labour and Delivery: Labour and delivery must be planned carefully with a multidisciplinary team well in advance. Successful vaginal and cesarean deliveries have been reported. The decision regarding mode of delivery should be based on the individual patient and the local obstetrical experience. If vaginal delivery is chosen, good pain management is very important. Epidural anesthesia should be initiated early and local anaesthetic drugs should be given in small and incremental doses. In general, less local anaesthetic and more narcotic is preferred to decrease the likelihood of further decrease in peripheral vascular resistance. During labour, valsalva manoeuver should be avoided. To decrease maternal expulsive efforts during the second stage of labour, forceps or vacuum delivery is often utilized. Oxytocin, which induces vasodilation and arterial hypotension, should be avoided. Immediate post partum hemorrhage should be watched for and aggressively treated. Maternal monitoring will often include telemetry, pulse oximetry, and invasive blood pressure monitoring. Air/particulate filters for all intravenous lines are essential for women with Eisenmenger syndrome. Compression stockings or thromboguards are very important around the time of delivery, along with early ambulation.

Post-Partum Care: The mortality risk remains particularly high postpartum. Attention to volume status is important.

After discharge, close postpartum monitoring is necessary. Care should focus in particular on managing volume status. The risk of pregnancy related complications exists until 6 months post-partum at which time pregnancy related hemodynamic changes will have fully returned to baseline.

REFERENCES

1. Jensen AS, Iversen K, Vejlstrup NG, Hansen PB, Søndergaard L (April 2009). "[Eisenmenger syndrome]". Ugeskrift for Laeger (in Danish) **171**(15):1270–5.PMID 19416617.
2. Erwin Oechslin, Siegrun Mebus, Ingram Schulze-Neick et. al. The Adult Patient with Eisenmenger Syndrome: A Medical Update after Dana Point Part III: Specific Management and Surgical Aspects. Curr Cardiol Rev. 2010 November; 6(4): 363–372. [PMCID:PMC3083818]
3. Gleicher N, Midwall J, Hochberger D, Jaffin H. Eisenmenger's syndrome and pregnancy. Obstet Gynecol Surv 1979; 34:721-41.
4. Weiss BM, Zemp L, Seifert B, Hess OM. Outcome of pulmonary vascular disease in pregnancy: a systematic overview from 1978 through 1996. J Am Coll Cardiol 1998; 31:1650-7.
5. Bédard E, Dimopoulos K, Gatzoulis MA. Has there been any progress made on pregnancy outcomes among women with pulmonary arterial hypertension?Eur Heart J. 2009;30(3):256-65.
6. Thorne S, MacGregor A, Nelson-Piercy C. Risks of Contraception and Pregnancy in Heart Disease. Heart 2006; 92:1520-5.
7. Bendayan D, Hod M, Oron G, Sagie A, Eidelman L, Shitrit D, et al. Pregnancy outcome in patients with pulmonary arterial hypertension receiving prostacyclin therapy. Obstet Gynecol 2005; 106 (5Pt2):1206-10.
8. Geohas C, McLaughlin VV. Successful management of pregnancy in a patient with Eisenmenger syndrome with epoprostenol. Chest 2003; 124:1170-3.
9. Kiely DG, Condliffe R, Webster V, Mills GH, Wrench I, Gandhi SV, Selby K, Armstrong IJ, Martin L, Howarth ES, Bu'lock FA, Stewart P, Elliot CA. Improved survival in pregnancy and pulmonary hypertension using a multiprofessional approach. BJOG. 2010; 117(5):565-74.

How to cite this article: Mahajan S, Mhaskar D. Eisenmenger Syndrome - the worst anticipated nightmare in pregnancy. Int J Health Sci Res. 2015; 5(11):412-416.

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