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Case Series

Eisenmenger syndrome in pregnancy: experience from a tertiary care centre

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ABSTRACT

Eisenmenger syndrome first described by Victor Eissenmenger is the end stage of a long-standing congenital left -right shunt, characterised by reversal of shunt, pulmonary hypertension and heart failure. Pregnancy with its haemodynamic changes, in these women is contraindicated due to high risk of poor maternal and foetal outcome, as stated by WHO and is considered for termination. However, if pregnancy is continued, management requires intense monitoring and multidisciplinary approach in a resource intense centre. Present article describes the clinical presentation, management and outcome of three pregnant women with Eisenmenger syndrome in a tertiary care centre in Rajasthan. All three women presented in late pregnancy for the first time. Two had a known congenital defect but were non-compliant with their treatment and follow up. They were unaware of the risks of pregnancy in their condition. One woman was diagnosed for the first time in pregnancy. All women delivered by emergency Caesarean Delivery due to complications. However maternal and foetal outcome was good in all three women and they were discharged in stable condition. Counselling regarding the need for regular follow up and avoidance of pregnancy was done at the time of discharge. Eisenmenger syndrome poses a significant challenge in pregnancy with high risk of complications to both mother and foetus. Favourable outcome may be achieved with advanced cardio- obstetric team in a well-equipped centre. However, counselling regarding avoidance of pregnancy or regular antenatal care must be impressed upon in young women with congenital heart defects.

Keywords: Eissenmenger syndrome, Congenital heart disease, Pulmonary hypertension, Pregnancy with heart disease, Cyanosis

INTRODUCTION

Eisenmenger syndrome (ES) is a complex and advanced cardiac condition that arises from a long-standing congenital left-to-right cardiac shunt. This shunt leads to the development of pulmonary hypertension and ultimately causes a reversal of blood flow to a right-to-left direction, resulting in chronic hypoxemia, cyanosis, and heart failure.[1,2] First documented by Victor Eisenmenger in 1897, this condition represents the final, inoperable stage in the continuum of uncorrected shunt lesions and poses profound clinical challenges, particularly during pregnancy.[3,4]

Advances in the early diagnosis and surgical correction of congenital heart diseases (CHDs) have enabled a growing number of women with cardiac conditions to reach reproductive age. Consequently, pregnancy in women with CHD is becoming more common, and with dedicated management guidelines, many can undergo pregnancy with favourable outcomes.[5,6] However, pregnancy in women with Eisenmenger syndrome remains a rare but

extremely high-risk scenario. It is associated with severe complications such as worsened hypoxia, thromboembolism, arrhythmias, and progressive heart failure. For this reason, ES with associated pulmonary hypertension is classified under the modified World Health Organization (WHO) pregnancy risk class IV, where pregnancy is considered strictly contraindicated due to a very high risk of maternal mortality and foetal morbidity.[7,8]

The physiological adaptations of pregnancy, particularly the decrease in systemic vascular resistance, can exacerbate the right-to-left shunt, leading to a dangerous cycle of worsening cyanosis and ventricular dysfunction. This necessitates close monitoring, often with weekly or biweekly evaluations, especially during the third trimester.[7,8] Despite optimal care, reported maternal mortality rates remain alarmingly high, ranging from 30% to 50%, and may escalate to 65% in the context of caesarean delivery.[9,10]

Globally, the exact incidence of ES in pregnancy is not well-defined, with the literature predominantly consisting of case reports. In pregnant women with CHD, the reported incidence of ES is approximately 3%.[11] Data from India are limited, with two large studies from the north and south reporting incidences of 1:9086 and 1:6953 pregnancies, respectively.[11,12] State-level data from Rajasthan is unavailable. Given the region's higher baseline maternal mortality ratio and challenges in healthcare access, outcomes for such critical cases, if not managed promptly and effectively, could be significantly worse.

Therefore, this article aims to present the clinical presentation, multidisciplinary management, and maternal-foetal outcomes of three pregnant women with ES managed at a tertiary care centre in Rajasthan, and to highlight the importance of individualized, high-risk obstetric care in improving outcomes in this life-threatening condition.

CASE SERIES

Case 1

A 23-year-old gravida one presented to the obstetric emergency department at 37 weeks and 4 days of gestation. She had a history of surgical correction for both ventricular septal defect (VSD) and atrial septal defect (ASD) in 2016. Although asymptomatic for the initial four years post-surgery, she gradually developed dyspnoea. She had been prescribed tablet Bosentan for pulmonary hypertension but reported poor compliance with medication. On admission, the patient reported shortness of breath. Her room air oxygen saturation was 80%. Vital signs were pulse rate (PR) – 84 bpm, blood pressure (BP) – 110/70 mmHg, respiratory rate (RR) – 22/min, and saturation – 80% on room air. Abdominal examination revealed a 36-week-sized uterus with a cephalic presentation, relaxed tone, and positive foetal heart sounds. On vaginal exam, the cervical OS was closed. Auscultation revealed a pansystolic murmur. No signs of heart failure were observed. Echocardiography revealed a residual VSD with right-to-left shunt, moderate tricuspid regurgitation, and severe pulmonary arterial hypertension. An Emergency lower segment caesarean section was done after taking high risk consent in view of non-reassuring foetal heart rate and a healthy Female child of 2.43 kg was delivered. Her postpartum period was uneventful and she was discharged in satisfactory condition with oxygen saturation of 88% with room air on postoperative day 7 along with the baby. She was discharged on Tab Torsemide 10 mg and Tab Sildenafil 20 mg thrice daily. On follow up visit at 6 weeks and 12 weeks at postnatal clinic, she had no complains of dyspnoea, she was able to do household work, her Spo2 was 88% on room air and the baby was healthy. She was advised to avoid strenuous activities and exercises and counselling was done regarding contraception and avoidance of pregnancy.

Case 2

A 32-year-old woman, gravida 2, unbooked, presented at 31 weeks and 2 days gestation with vaginal bleeding and passage of clots. Fifteen days prior, she had haemoptysis and was diagnosed with polycythaemia vera, large VSD, and Eisenmenger syndrome. She was started on Ecosprin 75 mg once daily and referred to our centre. On physical examination, her BP – 136/72 mmHg, PR – 100 bpm, RR – 22/min and oxygen saturation 70% on room air. She showed signs of right ventricular hypertrophy and grade 3 digital clubbing. Her oxygen saturation improved to 95% on 4L oxygen. Abdominal exam showed a 30–32 week-sized relaxed uterus, cephalic presentation, and positive foetal heart rate. Vaginal exam revealed a closed os and presence of clots. Cardiovascular examination revealed-Pansystolic murmurs audible across all cardiac areas, loud P2 in the pulmonary area and Normal first and second heart sounds. Investigations showed Haemoglobin was elevated to 22.2 g/dL, suggestive of polycythaemia vera. Bedside ultrasound showed a single live foetus of 31 weeks with 4×4 cm retroplacental hematoma. Echocardiography confirmed a large 19 mm VSD with a right-to-left shunt and severe pulmonary hypertension. MRI was not done due to patient instability. Patient was shifted for Emergency lower segment caesarean section in view of Antepartum haemorrhage after taking high risk consent and an alive Female child of 980 grams was delivered. Liquor was blood stained and retroplacental clots of around 150 cc removed. Patient was shifted to Cardiac Care Unit. The patient developed postpartum haemorrhage which was managed medically. She received postpartum thromboprophylaxis and oxygen support and tab sildenafil 20 mg twice daily during postpartum period. Management focused on optimizing oxygenation, avoiding dehydration, and preventing thromboembolic events.

Patient was discharged on day 10 of postnatal day on Tab Sildenafil 20mg twice daily. The baby had 1 month NICU

stay due to prematurity but foetal Echocardiography demonstrated no congenital heart defect. On follow up visit at 6 weeks and 12 weeks at postnatal clinic, she had no dyspnoea, she was able to do household work, her Spo2 was 94% on room air and the baby was healthy. She was advised to continue Tab Sildenafil 20mg twice daily, avoid strenuous work, and to strictly avoid pregnancy.

Case 3

A 20-year-old primigravida presented at 36 weeks of gestation with complaints of breathlessness, fatigue, orthopnoea, and bilateral pedal oedema. She had been diagnosed in childhood with Patent Ductus Arteriosus (PDA)and pulmonary hypertension but was unaware of the risks during pregnancy. The patient appeared cyanotic with an oxygen saturation of 82% on room air. Vitals parameters were PR – 80 bpm, BP – 122/80 mmHg, RR – 25/minute. Echocardiography confirmed a patent ductus arteriosus with severe pulmonary hypertension and moderate atrial regurgitation. Ejection fraction was 55%, consistent with Eisenmenger physiology. The patient was managed with oxygen therapy, diuretics, and close foetal monitoring. A multidisciplinary team including a cardiologist, obstetrician, and anaesthetist was involved in her care. She was advised about the risks and complications of pregnancy. An emergency caesarean section was performed for non-reassuring foetal heart rate after taking high risk consent, resulting in the delivery of a healthy preterm male baby of 2.31 kg. The mother was managed postoperatively for heart failure but stabilized with medical therapy. On discharge she was prescribed Tab Sildenafil 20 mg thrice daily. On follow up visit at 6 weeks and 12 weeks at postnatal clinic, she had no complains of dyspnoea, she was able to do household work, her Spo2 was 90% (room air) and the baby was healthy. She was advised to avoid strenuous activities and exercises and counselling was done regarding contraception and avoidance of pregnancy.

DISCUSSION

Pregnancy complicated by ES is a poorly tolerated condition with high risk of stroke, left heart failure, endocarditis, polycythaemia and sudden death.[11] This case series describes the management of three pregnant women with ES at a tertiary care hospital in western India. Literature review shows 72 cases identified with ES in 11 studies globally.[13] In our institute, out of 1011 deliveries, there were 13 cases of cardiac disease including three with ES. All three patients presented late in gestation, reflecting significant gaps in antenatal surveillance and cardiac screening. Two women had previously diagnosed congenital heart defects (VSD and PDA) yet were not under regular cardiology follow-up, shedding light on the persistent issues of delayed diagnosis, lack of preconception counselling, insistence on continuing pregnancy in spite of risks and the unpredictable nature of disease progression during pregnancy as observed in other studies by Dachlan ,Liu and James.[2,9,14] All cases in our study were delivered by caesarean delivery (CD) due to maternal or foetal indications, aligning with global experience. In the studies by Dachlan et al and Liu et al, 88% and 95.2%women delivered by CD respectively.[2-9] Although vaginal delivery may be associated with lower risk of blood loss and thrombosis but CD may be favourable due to faster procedure in a more controllable environment and maternal condition and without the risk of cardiac failure with contractions of labour.[2,10,15,16]

Despite the high-risk nature of these pregnancies, maternal outcomes were favourable in all three cases. Each woman was discharged in stable condition, comparable to that seen in reports by Pandey and Alsomali.[11-16] Absence of maternal mortality was a positive deviation from the high fatality rates 38.8% and 26.2% respectively, documented in existing literature.[2-10] However, all women required ICU admission and intense monitoring categorising them as near- miss events .This further emphasises the necessity of a resource-intensive, multidisciplinary approach involving obstetricians, cardiologists, anaesthetists, and neonatologists, which may not be feasible in less-equipped healthcare settings.

Foetal outcomes reflected the severity of maternal illness. Two neonates were delivered preterm and required NICU care, with low birth weights and compromised APGAR scores. These outcomes reflect the detrimental impact of chronic maternal hypoxia and reduced uteroplacental perfusion on foetal development, as reported by Bédard and Liu.[8,9] Encouragingly, all neonates were eventually discharged in good health, with no congenital cardiac anomalies noted on echocardiography.

These findings reinforce the current recommendations which advocate against pregnancy in women with ES. The use of risk prediction models and comprehensive preconception counselling should become standard practice in managing women with congenital heart disease.[7]

CONCLUSION

This case series underscores the significant challenges associated with managing pregnancy in women with ES, while also demonstrating that positive maternal and neonatal outcomes are possible with timely, well-coordinated care. To enhance outcomes in similar cases, there is a critical need to raise awareness about the risks of pregnancy in women with congenital heart disease through pre conceptional counselling. Strengthening antenatal screening, and promoting early referral to tertiary care centres are essential steps and would be definitive in preserving lives of both mother and foetus in these precarious situations.

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