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Case Report

Cardiac disease and pregnancy: hyper vigilance and extreme caution for optimal outcome

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ABSTRACT

Cardiac disease in pregnancy is a leading cause of maternal death in more so high-income countries. The armamentarium for winning this difficult battle involves shared decision-making with communication across the clinical team and the patient. There is limited clinical evidence concerning effective approaches to managing such complex care and moreover involvement of different specialists makes coordinated care challenging. Bicuspid aortic valve (BAV) is the most common congenital cardiac malformation, occurring in 1-2% of the population whereas a single ventricle is a rare congenital heart disease that accounts for less than 1% of all congenital heart diseases. We had two cases of pregnancy with bicuspid aortic valve in one case and the other with single ventricle. The involvement of multidisciplinary team involving cardiologist, cardiothoracic anaesthetist and fetal maternal medicine specialist resulted in good maternal and fetal outcome in both the cases.

Keywords: Bicuspid aortic valve, Multidisciplinary team, Single ventricle

INTRODUCTION

According to the most recent statistics from the World Health Organization (WHO), maternal mortality in developed economies is around 12 per 100 000 live births (0.012%) and 239 per 100 000 live births (0.2%) in emerging economies, with large disparities both between and within countries.¹ While leading causes such as haemorrhage and infection are declining, mortality due to maternal heart disease is increasing and in developed countries heart disease is now the leading cause of maternal mortality.²

CASE REPORT

Our first case was 23 years old Primi who was diagnosed to have bicuspid aortic valve with severe aortic stenosis

detected on evaluation for syncope. She was referred from peripheral hospital at 23 weeks period of gestation for cardiology and fetal maternal evaluation. On initial evaluation by cardiologist, she was found to have ejection fraction of 40 percent, concentric left ventricular hypertrophy and bicuspid aortic valve with peak systolic gradient of 128 mmHg and mean systolic gradient of 80 mmHg. Patient was planned for balloon aortic valvotomy, (BAV) and while under evaluation she developed tachycardia and breathlessness at 24 weeks POG. BAV was successfully done and after procedure gradient across the aortic valve reduced from 128 mmHg to 84 mmHg. At 37 weeks POG a joint decision for elective LSCS was taken by a team of obstetricians, cardiologists and anesthesiologists. Elective LSCS was performed under graded epidural and spinal anaesthesia with standby cardiac surgical team and cardiopulmonary bypass support for emergency aortic valve replacement

(AVR) in case of induction crash or arrest. The decision of graded epidural and spinal made intraoperative period smooth. Her post-operative period was uneventful and patient was discharged on postoperative day 8 with advice of 2 weekly follow up at cardiology centre.

Our second case was 27 years old Primi with complaints of easy fatigability at 31-week POG was referred for the first time to our centre for cardiology consultation and fetal maternal evaluation. She was found to have large VSD, severe pulmonary hypertension and single ventricle. On initial evaluation by cardiologist, she was found to have double inlet left ventricle, dextro transposition of great arteries, severe pulmonary artery hypertension and Eisenmengerisation. She was started on diuretics, Sildenafil and high flow oxygen with face mask to reduce the dynamic component of pulmonary artery hypertension.

Patient and next of kin were counselled about risk of maternal mortality associated with this condition. She was admitted and put on strict fetal-maternal surveillance. Fetal echo was also performed to rule out cardiac abnormality in fetus and fetal well-being was monitored by regular growth scan and doppler study. She was planned for elective LSCS in view of unfavorable cervix and prevailing cardiac condition. Elective LSCS was done at 35-week POG under low dose spinal and graded epidural anesthesia. Most important decision was the low dose spinal epidural which maintained her hemodynamics. The post op period was uneventful and patient was discharged on post op day 14 with advice of 2 weekly follow up at cardiology centre.

DISCUSSION

World Health organization divided pregnant women with cardiac disease into four classifications with class I having no detectable increased risk of maternal mortality and no/mild increase in morbidity and class IV having extremely high risk of maternal mortality or severe morbidity.

Cardiac disease in the pregnant patient can present challenges in cardiovascular and maternal-fetal management.³ Cardiovascular disease has been estimated to be present in 1% to 4% of pregnancies.⁴ Even in normal patients, pregnancy imposes some dramatic physiologic changes on the cardiovascular system. These include an increase in plasma volume by 50%, an increase in resting pulse by 17%, and an increase in cardiac output by 50%. After delivery, the heart rate normalizes within 10 days; by 3 months postpartum, stroke volume, cardiac output, and systemic vascular resistance return to the pre-pregnancy state.⁵

The outcomes in pregnant patients with bicuspid aortic valves (BAVs) are rarely reported despite the potentially critical nature of the condition. Bicuspid aortic valve (BAV) is the most common congenital cardiac

malformation, occurring in 1-2% of the population.⁶ The malformation involves both the valve and the aorta, and patients require lifelong surveillance.⁷ The critical complications of this condition are well described and include aortic stenosis, aortic regurgitation, infective endocarditis, and aortic dissection; of these isolated aortic stenosis is the most common associated disorder.^{8,9} Pregnant women with a severely stenotic BAV may experience cardiovascular deterioration, with 10-30% of this patient subset experience complications. Our patient also tolerated pregnancy due to balloon aortic valvotomy performed successfully in second trimester.

While cases of successful pregnancy and delivery in pregnancy with single ventricle have been reported in the United States and Japan and South Korea there have been no reported cases of pregnancy in Single functional ventricle patients in India so far.¹⁰⁻¹²

Pulmonary hypertension of any cause carries a high risk of maternal death even if pulmonary artery pressures are 50% of systemic blood pressure.^{13,14} The risk of death for a patient with Eisenmenger syndrome is 40-50%, unchanged in the last four decades. Patients should be advised against pregnancy.¹⁵ Maternal risk for such patients depends largely on ventricular function. Other risks are:

- Haemorrhage: impaired clotting factors and platelet function
- Paradoxical embolism: all cyanotic patients shunt right to left
- Heart failure: precipitated by the additional volume load of pregnancy on an already volume loaded ventricle
- Increasing cyanosis: caused by the vasodilation of pregnancy.

The major risks to the fetus are fetal loss, low birth weight, and prematurity related to maternal cyanosis and maternal hemodynamic decompensation. If pre-pregnancy maternal oxygen saturation is < 85%, the chance of a live birth is only 12%, compared with 92% if the oxygen saturation is > 90%.¹⁶ Prolonged admission for bed rest is an effective method of maintaining maternal oxygen saturation and hence fetal oxygenation. In our case also we hospitalized our patient for prolonged duration. Regular ultrasound assessment of fetal growth should be followed. We followed strict fetal surveillance of our patient by regular USG in the form of growth scan and dopplers and fetal echo was done to rule out cardiac abnormality in the fetus. For women with CHD, the prevalence of CHD among their offspring is approximately 10 times higher than that reported in the general population and it is therefore important that expert fetal cardiac scanning is undertaken.^{17,18} A high incidence of premature labour has also been noted in these patients which may contribute to morbidity and mortality.¹⁹ Maternal mortality in patients with cyanotic

congenital heart disease is also raised and has been associated with pulmonary hypertension and polycythemia.

The mode of delivery is controversial. Favorable outcomes reported following vaginal and caesarean birth using general, epidural and spinal anaesthesia. The literature is sparse in this sub-group of patients who present with advanced state of cardio-pulmonary decompensation. This suggests that each patient should be assessed by a combined obstetric, anaesthetic and cardiology team and a decision made on the mode of delivery.

Although spontaneous normal vaginal delivery is safest for the mother, in practice fetal problems often necessitate an elective section. Care must be taken to achieve meticulous haemostasis, maintain good hydration, and to avoid vasodilation (for example, to avoid high dose epidural anaesthesia) and air emboli through venous lines.

CONCLUSION

Certain cardiac conditions are a contraindication to pregnancy like our two cases one with bicuspid aortic valve and the other with single ventricle. It involves strict fetal and maternal surveillance with multidisciplinary team being involved together to tailor make management as per merit of each case.

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