

SAFE MOTHERHOOD IN A THIRD GRAVIDA WITH EBSTEIN'S ANOMALY AND SEVERE PRE-ECLAMPSIA

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ABSTRACT

Ebstein's anomaly is a rare congenital cardiac abnormality that may be associated with dyspnoea and cardiac arrhythmias. Some of the patients reach reproductive age but the newborn may have prematurity and low birth weight. We are reporting a case of 30 year old lady with Ebstein's Anomaly, who had 3 pregnancies successfully and delivered normal healthy babies at full term. In two of the pregnancies there was hypertension in the third trimester, which could be corrected with medicines.

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INTRODUCTION

Ebstein's anomaly is a rare congenital cardiac abnormality that may be associated with cyanosis and arrhythmias. A considerable proportion of women with this disease reach childbearing age. This case is being reported to address the issue of pregnancy outcome and complications in cases of Ebstein's anomaly.

CASE REPORT

Mrs.G, a 30yr old woman, G₃ P₂ L₂, at term gestation with Ebstein's anomaly, was referred from a peripheral hospital, in view of anemia and severe pre-eclampsia for evaluation and management. On admission, patient was asymptomatic. There was no history of any breathlessness, chest pain, palpitation, syncopal attacks or cyanotic spells. She was diagnosed as a case of severe pre-eclampsia at 8 months of gestation and was on anti-hypertensives since then. She did not give any symptoms suggestive of imminent eclampsia. On verification of past records, she was diagnosed as a case of congenital heart disease, Ebsteins anomaly during 1st pregnancy, 6 yrs back coincidentally during her antenatal check up in our Institution. She had no cardiac symptoms at that time. She gives history of hypertension in the first pregnancy. She delivered a full term normal, healthy female child of wt 2.5kg. Four years later the second pregnancy resulted in a male child by full term, normal vaginal delivery. Both the children are healthy. No congenital disease was detected in them. Intrapartum and postpartum period of both pregnancies were uneventful. In the present pregnancy, on the day of admission, patient was conscious and coherent. No signs and symptoms of imminent eclampsia. Pallor and pedal oedema grade 2 were present. Vital data: Pulse rate:102/min, regular in rate and rhythm, normal in volume.

BP: 200/120 mm of Hg in the right arm

Respiratory Rate was 22/min,

On examination of Cardiovascular system, a systolic murmur was heard in the tricuspid area and also in the

mitral area, lungs were clear. Rest of the systemic examination was normal.

Obstetric examination revealed a term gestation in cephalic presentation with clinically less liquor, a good fetal heart rate and a relaxed uterus. Pelvic examination revealed that patient was not in labour.

- Investigations: Hb - 11.7gm%:
- Platelets - 1.8 lakhs/cu mm
- RBS - 80mg/dl
- Renal function tests and liver function tests - Normal
- Urine albumen 2+
- Ultrasound examination of the gravid uterus showed a single live fetus of 32 wks gestation with Amniotic Fluid Index of 5 with grade 2 placental maturity
- ECG was normal
- 2D Echocardiography - Ejection Fraction: 78%
- Tricuspid valve: Septal leaflet was apically displaced.
- Severe Tricuspid Regurgitation with mild Pulmonary Arterial
- Hypertension present
- Dilated Right and Left Atrium.
- Good Left Ventricular and Right Ventricular Function

Cardiologist categorized the patient as NYHA functional class I - low risk for vaginal delivery or cesarean section. Patient set into labour after 4 weeks spontaneously after stripping of membranes. Antibiotic Prophylaxis was given. She delivered an active term female baby of weight 2.3 kg with good Apgar score.

She had mild postpartum hemorrhage which was controlled by uterine massage and uterotonics. She was observed in Intensive Care Unit for 24 hrs and then rest of her postpartum period was uneventful. The albuminuria resolved within 10 days. Hypertension was controlled with metoprolol. She was discharged after 3 weeks. Baby was followed up for 6 months. No congenital heart disease was detected in the baby during follow up. She was advised

permanent sterilization in view of her heart condition and completed family status.

DISCUSSION

Ebstein's anomaly is a rare congenital malformation of the heart first described by Wilhelm Ebstein in 1866 [1]. It is characterized by apical displacement of the septal and posterior tricuspid valve leaflets, leading to atrialization of the right ventricle with a variable degree of malformation and displacement of the anterior leaflet. During ventricular systole, the atrialized part of the right ventricle contracts with the rest of the right ventricle, which causes a backward flow of blood into the right atrium, accentuating the effects of tricuspid regurgitation. The severity of regurgitation depends on the extent of leaflet displacement, ranging from mild regurgitation with minimally displaced tricuspid leaflets to severe regurgitation with extreme displacement.[2].

A considerable proportion of these patients remain asymptomatic and reach childbearing age. Pregnancy is associated with an increase in both stroke volume and heart rate and decrease in peripheral vascular resistance[3] In women with Ebstein's Anomaly, and in the presence of declining right ventricular function, these changes may be poorly tolerated leading to worsening of tricuspid regurgitation, increased right atrial pressure followed by increased right to left shunting[4] With good right ventricular function, pregnancy may be well tolerated.

In the present case study, the patient was diagnosed with Ebsteins Anomaly in her first pregnancy. She was asymptomatic and delivered successfully in all the three pregnancies. Though the pregnancies were well tolerated, the patient had severe preeclampsia in all the three pregnancies in contrast to a few published reports which suggest preeclampsia as infrequent in Ebsteins Anomaly[5] Several case reports emphasize the potential complications in pregnancy with Ebstein's Anomaly[6-8]. In two studies on congenital heart diseases, 3 out of 5 women had cyanosis. The largest series published so far by Donnelly et al only 2 cases (12 patients studied) had cyanosis.

Patients with Ebsteins anomaly are liable [10] to develop supraventricular and ventricular arrhythmias, and Wolff-Parkinson-White syndrome that can occur in up to 20% patients[9]. Connolly et al reported in his study that though 14% of the women studied had one or more accessory conduction pathways at the time of pregnancy, they were not significant enough to require hospital admission or adjunctive medical therapy[10] In our case report, patient neither had arrhythmia nor cyanosis which could be attributed to her good Right ventricular function.

Our patient had successful spontaneous vaginal delivery not once but thrice. This is in accordance with study by Connolly et al which suggests that no patients in their series required induction or cesarean section for maternal heart condition.[10]

Maternal cyanotic congenital heart diseases had been shown to be associated with prematurity and low birth weight and with infants survival rate of 50-55%[11]. This may be due to maternal hypoxemia leading to growth retardation in the fetus and premature labour. The three infants born to the mother in our study were healthy, full term with an average birth weight of 2.5 kg. They were followed for 2 years after birth. None of them had congenital heart disease. This correlates with the case reports that show the incidence to be low or negligible [12].

The natural history and progress of the disease is uncertain as reported in a case study where unnecessary termination was advised to a patient of Ebstein's anomaly [13]. Though pregnancy was well tolerated in our case, we advised permanent sterilization in view of her completed family status and unpredictable heart condition.

Here, we had described the case of a patient with Ebstein's anomaly who gave birth to 3 healthy unaffected full-term infants. The anomaly was diagnosed during her first pregnancy, was not associated with other cardiac anomalies, cyanosis or arrhythmias. The echocardiographic degree of severity of the condition was low (grade 1). All these are good prognostic factors. During the pregnancies, no arrhythmias, cyanosis or signs of cardiac failure were observed and the patient's NYHA functional class I remained unchanged. Our case is one of the rare cases of three successful vaginal deliveries in a woman with Ebstein's anomaly, it confirms the importance of proper evaluation and the prediction of pregnancy outcome based on baseline characteristics of congenital heart disease.

CONCLUSIONS

Pre conception counseling regarding cardiac evaluation and assessment is a must in patients with Ebsteins Anomaly who wish to pursue pregnancy. The risk of pregnancy should be individually assessed depending on the cardiac functional status. A multidisciplinary team approach including a cardiologist, obstetrician and a neonatologist is required for successful and safe outcome in patients with Ebstein anomaly. Though pregnancy is well tolerated and fetal outcome is good in most of the patients, restricting family size and contraceptive advice should be given to all the patients in view of the unpredictable course of the disease and its complications.

DECLARATIONS

The patient Mrs G, had given written consent for publication of the details. No Identifiable details of the patient are submitted with the article.

CONTRIBUTORSHIP

Dr Anupama is mainly responsible for the data collection and work-up of the patient. Dr Abhijeeth, Aditya Hari, Akshay had managed the patient under the guidance of the first Author. Dr Nisha and Dr Rajeswari prepared the manuscript. All Authors reviewed the manuscript and approved the final version.

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