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PREGNANCY AND EBSTEIN ANOMALY - CASE REPORT

Obstetrics & Gynaecology

Dr. Akshitha Meenakshi Veera Rajkumar MBBS - Senior House Officer, Vikram Hospital.

Dr. Ashwinilakshmi. T. S. MS (OG)- Junior Consultant, Vikram Hospital

Dr. Sunitha. V. MD., FRCOG – Medical Director, Vikram Hospital.

ABSTRACT

Background- Ebstein Anomaly is a rare and complex congenital cardiac disease characterized by displacement of the posterior and septal leaflets of the bicuspid valve towards the apex of the right ventricle resulting in atrialisation of the right ventricle, severe tricuspid regurgitation and right atrial enlargement. Prevalence rate is 0.3-0.5%. Average life expectancy at birth of a patient with Ebstein Anomaly is 25 - 30 years. Due to its rarity, this case is presented. **Case Presentation-** A 28 years old Primigravida at 22weeks of gestation, a known case of Ebstein anomaly with clubbing and cyanosis came to the OPD for further management. She was started on Beta Blockers and antenatal period was uneventful except for the fetus developing late onset IUGR. Elective LSCS was done at 36 weeks in view of IUGR and poor bishop's score and a low birth weight baby of about 2 kgs was delivered. Intrapartum and postpartum period were uneventful. **Conclusion-** Ebstein anomaly during pregnancy can have varied clinical presentation. Hence multidisciplinary approach with close surveillance during pregnancy is necessary to diagnose complications and treat them early.

KEYWORDS

Ebstein anomaly, cyanosis, fetal Echo, IUGR, epidural anasthesia

INTRODUCTION

Ebstein anomaly is a rare and complex congenital heart disease with varied clinical presentation. Prevalence rate is 0.3-0.5%.[1] It is characterized by dysplastic abnormalities of tricuspid valves[2] involving both basal and free attachments of the tricuspid valve leaflets (especially posterior and septal leaflets) causing it to displace downwards and causing elongation of the septal and anterior cusp resulting in tricuspid regurgitation. [3]This causes atrialisation of the proximal part of right ventricle where a part of the right ventricle becomes thin walled and poorly contractile along with right atrial enlargement.[4-5]

Around 50 % of the affected fetuses die in gestation or shortly after delivery due to the right ventricular failure, while other newborns may require early surgery.[6] However, in milder types of EA, symptoms (mostly arrhythmias) may not appear until maturity, or even during pregnancy.[7]

Patient with Ebstein anomaly can have varied clinical course due to its anatomical abnormalities and their hemodynamics. They can also have associated structural defects or conduction system diseases like ASD, VSD, SVT, VT, pulmonary hypertension, tricuspid atresia, pulmonary stenosis and WPW syndrome.[5]

Life expectancy at birth is usually 25-30 years due to complications such as pulmonary and systemic embolism, congestive cardiac failure, sudden cardiac collapse and development of arrhythmias.

Maternal mortality is high if the mother also has associated conditions like cardiac failure, SV arrhythmias, WPW syndrome or atrial fibrillation. But less than 1% if there are no other complications and is asymptomatic.

Pregnancy in a patient with Ebstein anomaly is rare as life expectancy at birth is short and varied clinical presentation, that is why the case is presented.

Case Presentation

28 years old primigravida at 22 weeks of gestation, a known case of Ebstein Anomaly since 5 months of age and hypothyroidism on medications for the past 3 months came to the OPD. During childhood, she was diagnosed as a case of cardiac anomaly. She was asymptomatic since childhood and was not on any treatment.

On her first visit, the general condition of the patient was good. She had mixed cyanosis, clubbing, No pallor, icterus or oedema were seen. On examination her pulse was 70 beats per minute, regular, blood pressure was 110/70 mmHg and her jugular venous pressure was normal. Her oxygen saturation in room air was 90%. The respiratory system was normal. On cardiovascular system examination, S1, S2 with audible click and pan systolic murmur was heard. Right axis deviation with normal sinus rhythm was noted in ECG.

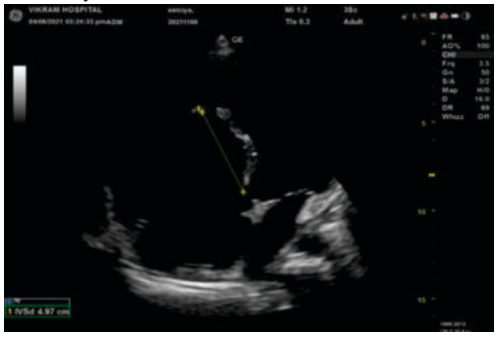

Figure 1: Two dimensional echocardiography showing ebstein anomaly

Echocardiography showed apical displacement of septal leaflet of tricuspid valve, elongated anterior tricuspid leaflet, dilated right atrium and right ventricle, severe tricuspid regurgitation, normal right ventricular function with left ventricular ejection fraction of 60%. Echo findings confirmed diagnosis of severe Ebstein anomaly with severe Goch score. Cardiologist started her on beta blockers and subsequent follow-up monitoring was advised.

Antenatal fetal ECHO revealed normal study. She was on regular follow-up once in 2 weeks. She was asymptomatic except for cyanosis, clubbing and saturation around 90%. She developed late onset IUGR at 32 weeks and was on follow up. Routine investigations reports were as follows -

Table 1: Investigation reports of the patient

Blood Group	O positive
Hemoglobin	12 g/dl
Coagulation profile	within normal limits
Total count, differential count	within normal limits

Renal function tests	within normal limits
Liver function tests	within normal limits
RBS	124 mg/dl
All viral markers	Negative

Serial monitoring of fetal growth done and on follow up her obstetric ultrasound scan showed a single live intrauterine fetus with cephalic presentation.

GA - 35 weeks + 1 day
Placenta – posterior
AFI - 9
EFW - 21st centile
AC – 29 weeks+ 6 days (IUGR)
Bishop score – 0

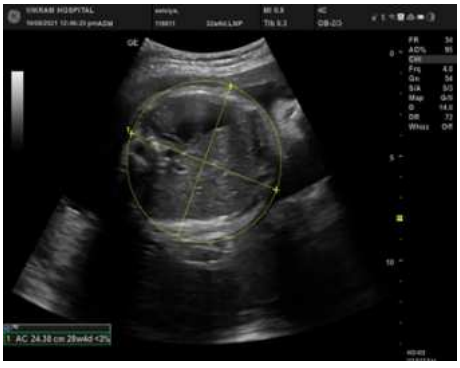

Figure 2: Obstetric ultrasonography revealing IUGR at 32 weeksNo gross congenital anomaly

Our team of obstetricians, anesthetist and Cardiologist decided for termination of pregnancy preferably by Cesarean section to avoid prolonged induction of labor in view of IUGR and unfavorable cervix. Elective LSCS was done at 36 weeks in view of IUGR and poor Bishop's score under epidural anesthesia and a low birth weight of weight 2 kg was delivered. Intra-operative and post-operative period were uneventful. Patient was discharged on 12th Post operative day after suture removal and advised for postnatal and cardiology follow-up. Contraception advice of IUCD given and patient complied.

DISCUSSION

Ebstein's anomaly (EA) presents a complex challenge, particularly in pregnant patients due to its impact on right ventricular function and the hemodynamic changes that occur during pregnancy. This discussion synthesizes current literature and the specific case presented to highlight the nuances of managing EA in pregnant patients and the implications for clinical practice.

Clinical Significance

The characteristic feature of ebstein anomaly is malformation of the tricuspid valve, leading to compromised right ventricular size and function. During pregnancy, the physiological increase in blood volume and cardiac output can exacerbate these issues. The increased right atrial pressure and volume further aggravate tricuspid regurgitation, while elevated catecholamine levels and maternal hypoxia can predispose patients to arrhythmias. [2] Consequently, even asymptomatic patients with EA may face significant risks, including heart failure, stroke, arrhythmias, and paradoxical embolism.

Impact of Pregnancy on EA

Pregnancy introduces several challenges for patients with EA. Increased blood volume and cardiac output during pregnancy can strain the already compromised right ventricle. This strain is compounded by elevated catecholamines and potential hypoxia, which increase the likelihood of arrhythmias. The severity of tricuspid regurgitation (TR) and the functional capacity of the right ventricle play crucial roles in determining the extent of hemodynamic disturbances experienced by these patients. [2]

Although fertility is generally unaffected in women with EA, pregnancy management is nuanced. The World Health Organization (WHO) classifies women with EA who do not exhibit cyanosis or heart failure as class II, indicating they usually tolerate pregnancy well. [8] However, symptomatic patients with cyanosis or heart failure should receive pre-pregnancy counseling and may require interventions before conception. Pregnant patients with EA are often acyanotic, but those with interatrial shunting risk shunt reversal and cyanosis during pregnancy. This necessitates rigorous monitoring and management to mitigate maternal and fetal risks. [2]

Management During Labor

The preferred mode of delivery for women with EA is vaginal, as this typically avoids the additional complications associated with cesarean sections. [8] During labor, it is essential to manage factors that could lead to congestive heart failure, cyanosis, and arrhythmias. Epidural analgesia is favored for its benefits, including minimal intravascular volume shifts, reduced catecholamine levels, and effective pain relief. In cases where cesarean delivery is necessary, epidural anesthesia can be upgraded to general anesthesia, ensuring effective pain control while managing hemodynamic stability. [1,12]

Intrathecal anesthesia, which can cause a sudden decrease in sympathetic vascular resistance, may complicate right-to-left shunts and is generally avoided. Managing labor effectively includes shortening the second stage to reduce intrathoracic pressure and the associated risk of exacerbating right-to-left shunts. [5,10]

Oxytocin, administered in standard doses (5–10 IU intramuscularly), is preferred due to its relatively benign effect on the cardiovascular system. Large doses of oxytocin, as well as methylergometrine and prostaglandins, are avoided due to their vasodilating effects and potential to increase pulmonary vascular resistance. [4,5]

Outcomes and Prognosis

In reviewing literature on pregnancies complicated by EA, Donnelly et al. observed that out of 42 pregnancies in 12 patients, most were well-tolerated, resulting in 36 live births. Mild dyspnea in the third trimester was common, with a few cases experiencing arrhythmias and right heart failure. [2] Similarly, Chopra et al. documented outcomes in 8 pregnancies among 4 patients with EA, noting occurrences of right heart failure and arrhythmias. [10]

Connolly and Warnes reported on 111 pregnancies in 44 women with EA, finding that 76% resulted in live births, with 89% delivered vaginally. They noted that cyanotic women had significantly lower birth weights compared to their acyanotic counterparts. [14] This underscores the impact of maternal cyanosis on fetal outcomes.

Pregnant women with EA face increased risks of major adverse cardiac events, including in-hospital death, myocardial infarction, cerebrovascular events, and cardiac complications during labor and delivery. [14] These patients also have a higher incidence of preterm delivery, postpartum hemorrhage, and cesarean sections.

CONCLUSION

Management of pregnancy in patients with Ebstein's anomaly requires a multidisciplinary approach, emphasizing careful monitoring and tailored interventions to address the unique hemodynamic challenges posed by EA. While many women with EA can have successful pregnancies, the risks associated with cyanosis, arrhythmias, and right heart failure necessitate a proactive management strategy to optimize outcomes for both mother and infant. Future research should focus on refining guidelines for the management of EA during pregnancy and improving our understanding of long-term outcomes for these patients.

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