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Ebstein Anomaly with Pregnancy: A Rare Case

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Abstract: ***Background: Ebstein anomaly is an uncommon, complex congenital malformation of the heart with an incident of one in 20000 live births, characterized by the typical displacement of septal and posterior leaflets of the tricuspid valve resulting in reduction in size of the right ventricle and atrialised of upper part of the right ventricle that behaves functionally as a part of the right atrium. These patients have severe tricuspid insufficiency which contributes to the enlargement of the right atria. Maternal mortality due to Ebstein is considered to be less than 1% in asymptomatic patients but may be as high as 5-15%, if aggravated by conditions like supraventricular arrhythmia, WPW syndrome or atrial fibrillation. Case Presentation: A 26-year-old G4P2A1L0 at 33 weeks 2 days gestation with a known with a known case of Ebstein anomaly was referred to Civil Hospital Ahmedabad in August 2020 with complain of breathlessness for further management as our institute is having well equipped cardiac care facilities. She was diagnosed as case of ebstein anomaly during 6-month amenorrhea and it was confirmed by ECHO during her treatment of cervical insufficiency. Antenatal steroid administration was done, and cardiology opinion was taken for termination of pregnancy, elective LSCS was done at 35 weeks and healthy baby weight 2.3 kg was delivered. Intrapartum and postpartum period was uneventfully. Conclusion: Due to varied clinical presentations, ebstein anomaly needed multidisciplinary approach during antenatal period to be diagnosed in term of any complications and treated accordingly.***

Keywords: Ebstein anomaly, feto-maternal outcome

1. Introduction

Ebstein anomaly (EA) is an uncommon, complex congenital malformation of the heart with an incident of one in 20000 live birth.

It is characterized by the typical displacement of the septal and posterior leaflets of the tricuspid valve resulting in reduction in size of the right ventricle and atrialization of the upper part of the right ventrical that behaves functionally as a part of the right atrium. These patients have severe tricuspid insufficiency which contricutes to the enlargement of the right atria.

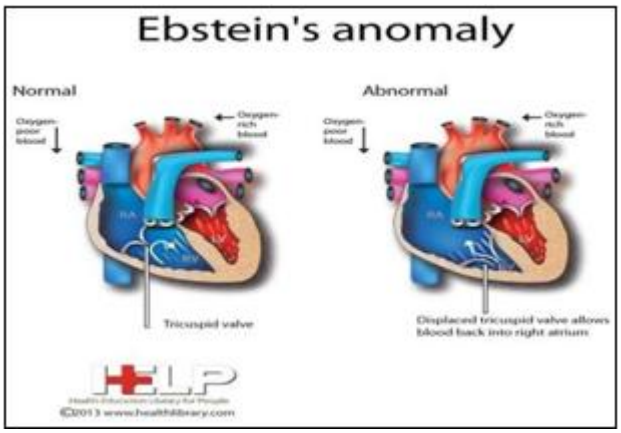
Patients can have a highly variable clinical course related to the anatomic abnormalities of Ebstein anomaly and their hemodynamic effects or associated structural and conduction system disease, like Atrial septal defect (ASD) (90%), pulmonary hypertension, ventricular and supraventricular tachycardia, ventricular septal defect, tricuspid atresia (30%), pulmonic stenosis and Wolf - Parkinson - White syndrome (WPW Syndrome) (up to 20% of patients).

Cyanosis, pulmonary and systemic emboli, congestive cardiac failure and sudden cardiac collapse are anticipated complications.

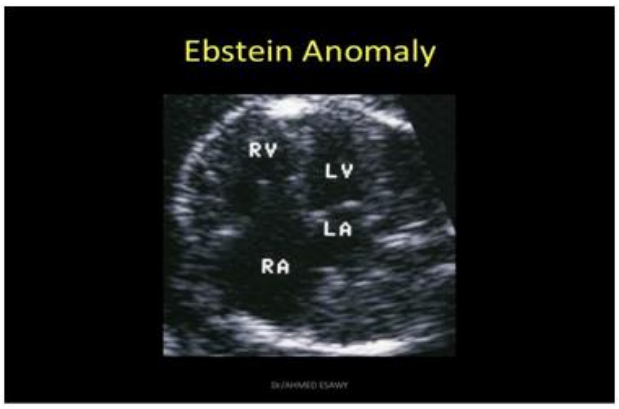
Risks of pregnancy co - relates well with the degree of tricuspid regurgitation, right ventricular function and presence of cyanosis.

Maternal mortality due to Ebstein anomaly is considered to be less than 1% in asymptomatic patients but may be as high as 5–15%, if aggravated by conditions like supraventricular arrhythmia, WPW syndrome or atrial fibrillation.

Due to its rarity and short life expectancy at birth, there are not many cases and varied clinical presentations associated with Ebstein anomaly during pregnancy; therefore, this case is presented to increase aware obstetricians.



Schematic representation of defect



2D echo of patient with ebstein anomaly

2. Case Presentation

A 26 year old G4P2A1L0 at 33 weeks 2 days gestation with a known case of Ebstein anomaly was referred to CIVIL HOSPITAL AHEMDABAD on August 2020 with c/o breathlessness* 12 hours for further management as our

institute is having well equipped cardiac care facilities. She was diagnosed as a case of ebstein anomaly during 6 month amenorrhea and it was confirmed by ECHO during her treatment for cervical insufficiency (os tightening stitches). All these years, she did not have any symptoms related to the condition and was not on any treatment. . On admission, no pallor, no icterus, no cyanosis, no oedema, no clubbing was seen. On examination of the respiratory system, no abnormality was detected. Her pulse rate was 102 beats per minute, regular and blood pressure was 110/80 *mm Hg* and her jugular venous pressure was normal. Her oxygen saturation in room air was 90%. On cardiovascular system examination, a split S1 was heard with audible S2, S3 and S4. Pan systolic murmur was also heard. On ECG, right axis deviation with normal sinus rhythm was seen. Echocardiography showed apical displacement of septal leaflet of tricuspid valve, elongated anterior tricuspid leaflet, dilated right atrium and right ventricle, moderate tricuspid regurgitation, small ASD and normal right ventricular function with left ventricular ejection fraction of 55%. Echo findings confirmed diagnosis of Ebstein anomaly. Other routine investigation reports were as follows:

Blood group: O positive, haemoglobin 12.5 *gm/dl*, TLC and DLC: WNL, coagulation profile: normal, liver function tests and kidney function tests: normal, and RBS: 102 *mg/dl*. All viral markers were negative and thyroid profile was normal. Ultrasound obstetrics doppler scan was done on the day of admission that showed single live intrauterine fetus with cephalic presentation; the gestational age was 27 weeks 4 days, placenta was posterior, liquor was adequate, and expected fetal weight was 1.7 *kg*. Umbilical artery Doppler showed a normal study.

No gross congenital anomaly in the fetus was observed. Antenatal Steroid administration was done. Cardiology opinion was taken and patient was advised to accept termination of pregnancy preferably by caesarean section to avoid stress of labor.

Elective LSCS was done at 35 weeks under general anaesthesia with antibiotic coverage (Injection of ampicillin 1 gram and injection of gentamycin 80 *mg* intravenously) for bacterial endocarditis prophylaxis. Os tightening stitches were removed intra op. A healthy male baby weighing 2.3 *kg* was delivered. Patient kept in obstetric ICU for 24 hours for monitoring. After delivery, ECHO of baby was done, which was normal. Exchange transfusion of the baby was done in view of high bilirubin Post - operative cardiology opinion was taken and patient was started on tab metoprolol and toresemide. Patient was given higher antibiotics for 7 days post op. Suture removal was done on post operative day 15th and was advice to attend postnatal clinic and cardiology OPD after 1 month. On follow - up, patient was advised to use progesterone only pill to avoid pregnancy.

3. Discussion

In Ebstein's anomaly, there is compromised right ventricular size and function, further impaired by the increased blood volume and cardiac output during pregnancy. . Increased right atrial pressure and volume both worsen tricuspid regurgitation. Raised catecholamine with maternal hypoxaemia and stress levels in pregnancy further predispose the cases to arrhythmia. The haemodynamic problems seen during pregnancy depend on the severity of TR and the functional capacity of the RV. Heart failure, stroke, arrhythmias, paradoxical embolism can occur even in the asymptomatic patients.

With this anomaly, fertility is usually unaffected, even in women with cyanosis. According to WHO, women with Ebstein anomaly without cyanosis and heart failure are categorized in class II and usually tolerate pregnancy well, but symptomatic patients with cyanosis and/or heart failure should be treated before pregnancy or counseled against pregnancy. While pregnant patients with EA are usually acyanotic, those with interatrial shunting can develop shunt reversal and cyanosis in pregnancy. The presence of arrhythmia or cyanosis in the mother is associated with increased maternal and fetal risk, and needs closer maternal and fetal monitoring during pregnancy and delivery. Mild cyanosis is associated with increased premature deliveries, low birth weight and thromboembolic complications.

During intrapartum period, one should avoid all factors leading to congestive heart failure, cyanosis and arrhythmias Management of patients with Ebstein's anomaly during labor focuses on maintaining normal sinus rhythm, avoiding fluid overload and providing enough relief of pain to the patient by epidural analgesia which can be upgraded to anesthesia if cesarean section is indicated. Large doses of oxytocin have discernible vasodilating effects and should be administered cautiously. Methylergometrine and prostaglandins increase pulmonary vascular resistance and are generally avoided.

Oxytocin (5–10 *IU* intramuscularly) is commonly followed in obstetric protocols AMTSL.

4. Conclusion

Maternal and fetal prognosis is favorable in patients with Ebstein anomaly and NYHA class I. But it can be complicated with various major cardiac events.

Due to varied clinical presentations associated with Ebstein anomaly during pregnancy, such women should undergo close surveillance with multidisciplinary approach during the antenatal period to be diagnosed in terms of any complications and hence to be treated accordingly.

References

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