

Successful pregnancy in a patient with Ebstein's anomaly; a case report from a developing country

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Abstract

Ebstein anomaly is a rare malformation of the tricuspid valve of the heart. The malformed tricuspid valve may be incompetent, stenotic, or rarely, imperforate. Pregnant patients with Ebstein anomaly become a challenge for the physician in terms of management. We report a case in which patient delivered successfully with Ebstein anomaly. The only complication was breathlessness. Authors concluded that women with Ebstein anomaly can have successful outcomes of pregnancy with close monitoring.

Keywords: Ebstein anomaly; Pregnancy; cardiac malformation

Introduction

Ebstein anomaly (EA) is a rare cardiac congenital abnormality characterized by downward displacement of the posterior and septal leaflets of the tricuspid valve which results in atrialization of the right ventricle, enlargement of the right atrium and tricuspid regurgitation. Affected individuals experience a wide spectrum of clinical severity, ranging from heart failure in infants to asymptomatic adults and identification of the disease later in life. Other clinical presentations include cyanosis, arrhythmias and paradoxical emboli through atrial level shunts. Imaging modalities such as echocardiography and cardiac magnetic resonance are used for diagnosis. Appropriate surgical and medical management tailored to each patient's anatomy and haemodynamic status is necessary in order to ensure acceptable patient outcomes. Since most patients survive to childbearing age, understanding of the haemodynamic changes during pregnancy and careful planning of labour and delivery are paramount.¹ The woman can present with cyanosis and arrhythmias. We report a case with successful management of a pregnant patient with Ebstein anomaly. The case was first seen in February 2016 at Liaquat National Hospital, Karachi, Pakistan. Informed consent was taken from patient for publishing the case. Authors of the study have explored

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the local data and according to the best of our knowledge this is the first case of pregnancy with Ebstein anomaly reported from our country. Pakistan has different demographics in terms of genetic, race and economics. This case report provides significant information regarding Pakistani women who are managed for pregnancy with Ebstein anomaly.

Case Report

A 25 year old female, married for 2½ years, primigravida, known case of cardiac disease presented in consultant clinics for antenatal care in February 2016 at Liaquat National Hospital, Karachi, Pakistan. She had a past history of palpitations off and on for 10 years and occasional attacks of syncope every 3 to 4 months. Three years back she was diagnosed with Ebstein anomaly. Her mother was hypertensive. Antenatal history revealed spontaneous conception. She felt foetal movements at 5th month. Patient complained of palpitation on exertion on and off in third trimester.

On examination Blood Pressure was 120/70mm Hg, pulse was 180 beats/minute, temperature was 98°F and oxygen saturation was 98%. Chest and Nervous System examination was unremarkable. On cardiovascular examination, abnormal heart sounds were present in tricuspid area. On obstetrical abdominal examination, the height of fundus was 38cm, Presentation was cephalic, Lie was longitudinal and there were no uterine contractions at 32 weeks of gestation.



Figure-1: Showing Echocardiograph of patient in first pregnancy. Ebstein anomaly (mild type) is seen. Normal sized left ventricle with mild dysfunction. There was no free vegetation seen.

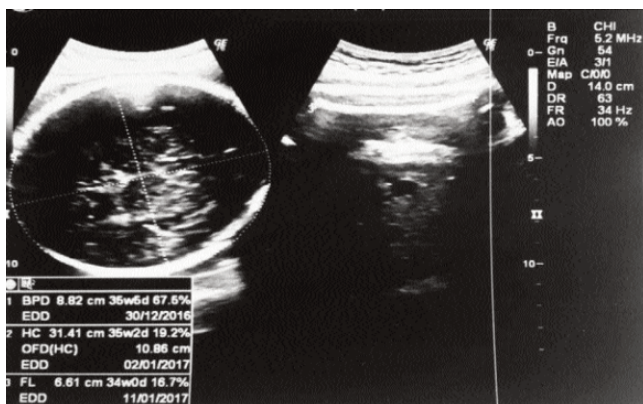


Figure-2: Showing the Ultrasound of patient. Single alive intrauterine foetus corresponding to gestational age of 34 weeks \pm 1 week. Umbilical artery doppler appear normal. Middle cerebral artery of PI falls at 40th percentile. Umbilical artery PI is at 60th percentile. CPR is at 15th percentile.

Cardiology review was done. Both maternal and foetal monitoring was performed by frequent antenatal visits with ECG report and echocardiography every 4 weeks. Foetal growth parameters were also monitored by regular growth scans and Doppler ultrasound for fetoplacental insufficiency. She was taking Bisoprolol 2.5mg daily. ECG reported sinus tachycardia. Ultrasound pelvis and Doppler of umbilical artery was done for foetal growth and to rule out intrauterine growth retardation (IUGR) Ultrasound showed single alive foetus with breech presentation, Amniotic Fluid Index was normal, placenta was lying posteriorly in upper segment at 36 weeks of gestation. No placental insufficiency was noted,

In view of persistent tachycardia and generally increasing breathlessness even on mild exertion, the cardiologist suggested elective caesarean section to avoid cardiac failure on exertion in normal vaginal delivery. On these grounds and recommendation of cardiologist, decision was taken for an elective lower segment caesarean section under spinal anaesthesia. An alive male foetus was delivered with APGAR score of 4/1 and 9/10 and weight of 2.5Kg. Vitamin K was given to the neonate and patient was shifted to the cardiac unit for monitoring. On second post operative day patients had two episodes of ventricular tachycardia which were treated with successful defibrillation. ECG and ECHO were repeated and patient was kept on symptomatic treatment. Pulse rate remained stable later on and she was discharged on 6th post operative day in a stable condition. Baby echocardiogram was done to rule out Ebstein anomaly. The Imaging of patient is

shown in Figure-1 and 2. On post natal follow up, after 4 weeks she was coping well with her baby and the cardiology review was also satisfactory.

In between the two pregnancies, patient had surgical intervention in Peshawar for Ebstein anomaly, Electrophysiologic resection was done by ablation of tricuspid leaflets which resulted in improvement of cardiac status on echocardiography.

Recently patient had her second delivery. She conceived after 10 months, again and reported late at 34 weeks of pregnancy. During this pregnancy she had similar symptoms, but no further deterioration of cardiac status. She delivered at 36 weeks of gestation by Caesarean section again. On third post-operative day one episode of breathlessness and tachycardia was experienced, controlled on Bisoprolol tablets. Baby was normal and was breastfed. Paediatric echocardiography was normal.

Discussion

It is suggested that before advising against starting or terminating pregnancy in Ebstein anomaly a careful assessment should be done since successful outcomes are possible.

Women with complex congenital defects, with or without surgical repair and/or residual defects, should be managed in tertiary care centers under a multidisciplinary team of physicians experienced in adult congenital heart disease and high risk obstetrics, who collaboratively participate in pregnancy planning, management and care through childbirth and postpartum.²

An international study reported successful pregnancy with Ebstein anomaly, they reported the only complication in their setting was breathlessness. They delivered the baby at 30 weeks of gestation through Caesarean section.³ Similarly in our study pregnancy was uneventful except for breathlessness and baby was delivered through Caesarean section at 36 weeks of gestation.

In another international study they reported a case of undiagnosed Ebstein anomaly which presented at 37th week of gestation with hypoxia, dyspnoea, central cyanosis and uterine contractions. The patient underwent emergent Caesarean section. Ebstein anomaly was later diagnosed by echocardiography.⁴ It proved that pregnancy with undiagnosed Ebstein anomaly can be difficult to manage.

A study from Bangladesh⁵ reported a case of Ebstein anomaly which presented at 34 weeks of gestation. The

woman underwent spontaneous pre-term labour and gave birth to a still born. She developed peripheral cyanosis during pregnancy which subsided few days after delivery. In contrast our patient was uneventful except for breathlessness at 36 weeks of gestation and the outcome was normal.

Conclusion

Women with Ebstein anomaly can have successful outcomes of pregnancy with close monitoring. They should be counseled accordingly.

Disclaimer: None to declare.

Conflict of Interest: Prof Haleema A Hashmi the academic head of obstetrics and gynaecology, gave

approval for submission of this case report. She is also a co-author in the case report.

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