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Successful Pregnancy in a Patient with Ebstein's Anomaly: A Rare Case Report

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Abstract:

Ebstein's anomaly, a rare congenital heart disease, occurring in 1 in 20,000 live births. The leaflets are placed deeper into the right ventricle instead of the normal position. Pregnancy however is well tolerated and the maternal risks of pregnancy co-relates well with the degree of tricuspid regurgitation, right ventricular function and presence of cyanosis. Aim is to emphasize the meticulous follow-up with fetal & maternal medicine specialist with multidisciplinary approach can give a successful outcome. Proper diagnosis and meticulous follow-up with good ANC have made this case, pregnancy with Ebstein's anomaly successful.

Key words: *Ebstein's anomaly, Multidisciplinary approach, Echocardiography, TR (Tricuspid regurgitation), EF (Ejection fraction), Pulmonary hypertension*

Introduction:

Ebstein's anomaly, a rare congenital heart disease, occurring in 1 in 20,000 live births.[1] It has an extremely variable natural history, depending on variety of underlying pathology. The leaflets are placed deeper into the right ventricle instead of the normal position & the defect most often causes the valve to work poorly, and blood may go the wrong way. Instead of flowing out to the lungs, blood flows back into the right atrium. The backup of blood flow can lead to heart enlargement and fluid buildup in the body. There may also be narrowing of the valve that leads to the lungs (pulmonary valve) causing pulmonary hypertension.

Pregnancy however is well tolerated and the maternal risks of pregnancy co-relates well with the degree of tricuspid regurgitation, right ventricular function and presence of cyanosis.

In this case Verbal consent has taken both from patient and her husband.

Objective: To disseminate information that careful F/U of high-risk pregnancy by Feto-maternal Medicine department, patient's consciousness & multidisciplinary approach can give a successful outcome.

Case Report:

A 27 years old female, G2, para 1, ALC 6 years, presented at 38 weeks of gestation with diagnosed case of Ebstein anomaly (EA) with Tricuspid regurgitation (TR-grade ll) with moderate pulmonary hypertension with history of one C/S with mild IUGR. According to statement of patient she was suffering from recurrent episodes of mild respiratory distress which developed just after one week of her marriage. She also gave the history of several episodes of fainting. With that complains, she first visited to local doctor then Mymensingh medical college (MMC) and referred to BSMMU and diagnosed as a case of EA and since then was on treatment.

Two years later she become pregnant and referred to BSMMMU from MMC and was under multidisciplinary approach (Feto-maternal Medicine, Neonatology and cardiology department). Throughout the pregnancy no cyanosis or features of heart failure developed but due to fetal distress C/S was done at 38 weeks and I hour after C/S severe respiratory distress developed and referred to CCU but 24 hours later she became quite normal with all parameter. She again became pregnant and this pregnancy was unplanned but on

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regular ANC with infrequent cheek-up by specialist. During her antenatal period, she was admitted twice in hospital for respiratory distress. She was managed conservatively both the times. She had mild dyspnoea and palpitations in the third trimester but she remained in NYHA class I throughout her pregnancy. There was no history of recurrent chest infections, cyanosis or heart failure in interval of two pregnancies and no significant family history. She was followed her by regular echocardiography. Physical examination revealed blood pressure of 124/80 mm Hg; heart rate of 80 bpm. Her oxygen saturation (SpO2) at room air was 98%. Liver was not palpable and there were no other signs of heart failure and cyanosis. Last Echocardiography showed TR (Grade II), moderate pulmonary hypertension, arterialization of right ventricle, EF - 72%, no left ventricular diastolic dysfunction. ECG demonstrated Sinus rhythm. Her CBC, TSH, glucose profile, serum Bilirubin, SGPT, SGOT, was normal but alkaline phosphatase was moderately raised. Fetal biophysical profile was good. Prophylaxis for bacterial endocarditis was given.

After consultation with Cardiologist, Anesthesiologist, Neonatologist with proper counseling LUCS with BLTL was done at 39 weeks under epidural anesthesia and epidural analgesia was maintained 72 hours postoperatively. Intra and postoperative period was uneventful. A male baby weighing 2.5 kg was delivered with APGAR score 6/10 in I min and 8/10 at 5 min. Baby managed and followed up by neonatologist and was good in all parameters.

Important investigation findings:

Name	Findings
Echo	Ebstein anomaly, Moderate TR (G II) with dilated Right atrium, Moderate pulmonary hypertension, Good biventricular function, EF-72%
ECG	Sinus Rhythm
USG	BPP & Doppler normal, no fetal anomaly but feature of mild IUGR
CTG	Reactive
CBC	Normal parameter
Glucose profile	Do
TSH	Do
Liver enzyme	Normal except slight rise in Alkaline phosphatase

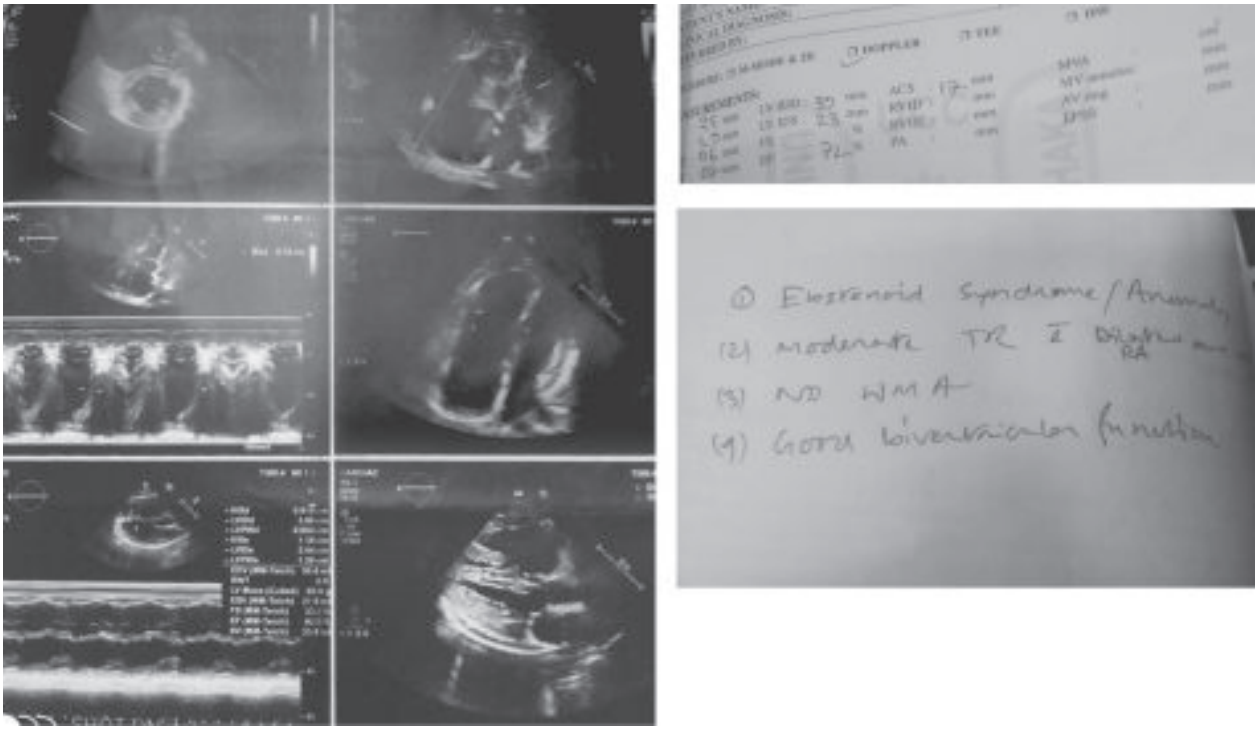

Fig-1 *Maternal Echocardiography*

Immediate Postnatal F/U:

Color – pink in room air, R/R-48 breath/min, H/R-142 b/min, CRT (Capillary refill time) -2 second, Abdomen-soft, nontender, No gross external anatomical defect found Bowel & Bladder- moved

Discussion:

An inter-atrial communication (Atrial Septal Defect or Patent Foramen Ovale) is present in 80-94% of patients of EA. Accessory pathways (Wolff-Parkinson-White syndrome) are commonly associated with EA (6-36%) and may lead to supra-ventricular tachycardia. First degree heart block is found in 50% of patients (may relate to right atrial dilatation and stretch).[2] Our patient did not have any signs of pre-excitation. In patients with Ebstein's anomaly these physiological changes may have appreciable adverse hemodynamic consequences. In the presence of impaired right ventricular size and function, increased blood volume may be poorly tolerated and result in worsening of tricuspid incompetence, raised right atrial pressure and increased right to left shunting.[3]

This anomaly does not have any effect on fertility, even in women with cyanosis.[4] According to the current guidelines, women with EA without cyanosis and heart failure are encountered to World Health Organization (WHO) risk class II and usually tolerate pregnancy well.[5]

In contrast, symptomatic patients with cyanosis and/or heart failure should be treated before pregnancy or counseled against pregnancy.

The management during labor should avoid all factors leading to congestive heart failure, cyanosis and arrhythmias.[6,7] To maintain normal sinus rhythm during labor, adequate pain relief in the form of epidural analgesia is helpful and can be upgraded to anesthesia if caesarean section is indicated.

The risk of congenital heart disease in offspring is reported in 4–6%, and familial EA in 0.6%.[6]

Conclusion::

In absence of severe maternal complications (cardiomegaly, cyanosis, arrythmia), pregnancy is well tolerated and in those with mild cardiac dysfunction as evaluated at echocardiography and a low NYHA class, maternal & fetal outcome is quite good. However EA may present with a multitude of problems and hence should be considered as high risk and cared for in a tertiary centre by a multidisciplinary team involving an obstetrician, cardiologist, anaesthetist and pediatric cardiologist

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