VOLUME-9, ISSUE-5, MAY -2020 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Gynaecology

PREGNANCY OUTCOME IN A WOMAN WITH EBSTEIN ANOMALY

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ABSTRACT Ebstein anomaly is a rare congenital heart disease most often asymptomatic and in women it may diagnosed during pregnancy for the first time because symptoms appear due haemodynamic changes of pregnancy. A 23 year old primigravida was diagnosed as Ebstein anomaly as she had symptoms of breathlessness and haemoptysis during first trimester of pregnancy and was referred for MTP. Cardiac evaluation revealed mild to moderate variant and pregnancy was continued and Obstetric and Cardiac care provided in a tertiary care Institute. There were no worsening of cardic condition and she delivered a healthy baby at term under epidural analgesia.

KEYWORDS : Pregnancy, Ebstein anomaly, NYHA Class II,

Introduction: Ebstein anomaly is a rare congenital heart disorder (CHD) reported to occur in less than 1% of all CHD. The life expectancy is reduced and this condition is usually diagnosed during pregnancy or when there is arrhythmias1. Maternal mortality increases up to 15% when associated with supraventricular arrythmia, WPW Syndrome and atrial fibrillation 2. We report here a case of Ebstein anomaly which was diagnosed during pregnancy due its rarity.

CASE:

A 23 year old primi gravida a known case of bronchial asthma since childhood was diagnosed to have congenital heart disease at 12 weeks of pregnancy. She was asymptomatic till then when she had breathlessness and hematemesis . she was confirmed to have Ebstein's anomaly by Echo. She had low placed tricuspid valve, atrialisation of right ventricle, moderate tricuspid regurgitation and there was no pulmonary hypertension. She was referred to our tertiary care centre for MTP. She is married for one year, non consanguineous marriage and there was no family history of heart disease or anyother medical disorders.

On Examination: she is well built , weight - 47 kg, no pallor, BP – 95 / 58 mm Hg ;CVS – S1,S2 Present pan systolic murmur present , RS- clear ;ECG Showed sinus rhythm right axis deviation, Cardiac evaluation by cardiologist confirmed Ebstein's anomaly, with moderate TR and a VSD of 3.5 mm with bidirectional shunt , normal LV function and opined to continue pregnancy as she was low cardiac risk. She had regular antenatal care at our Institute and received folic acid and other haematinics. Her investigations are shown in the table.

She was hospitalised at 38 wks. Her weight gain was 13 kg from the first trimester and symptoms did not worsen and she was NYHA class II throughout pregnancy. She was evaluated at term by cardiologist again and a decision was taken for vaginal delivery. She has spontaneous onset of labour at 39 ks 4 days and was managed in Obstetric ICU with propped up position and oxygen by mask and continuous CTG monitoring. Infective endocarditis prophylaxis was started and Oxygen saturation was 95 to 99% throughout labour. Labour analgesia was given with infusion of 0.0625% bupivacaine combined with 2 micrograms per ml of Fentanyl at the rate of 5 ml per hour in active phase of labour. She was on oxytocin for prolonged latent phase and progressed normally thereafter and delivered an alive male fetus 3.7 kg by Low forceps applied to cut short second stage. There was no hypotension or tachycardia throughout labour and there were no third stage complications. Her symptoms did not worsen and NYHA class II when was discharged on postnatal day 5.

Table: Investigations during antenatal Period

Date	Investigations	Results	Echo		Antenatal ultrasound
10/03/20	Hb	10.2gm%	Date	ECHO –Peripheral Health Centre	28/11/19
	Blood group	O positive	5/10/19	Ebstein's anomaly, TV placed high, atrialisation of right ventricle , moderate TR	SLIUG, CA+,18 wks, no GCM, placenta posterior. Fetal Echo –Normal
	HIV	Negative			
	HbsAg	Negative			
	VDRL	Negative			
10/01/20	GTT(75gm)	87/124/132	14/10/19	Small septal aneurysm. ECHO at Tertiary Care	
4/11/19	S. TSH	0.44 mIU/L			
20/4/2020	Hb	11 gm%			
			5/11/19	Ebstein's anomaly, low placed tricuspid valve, no PAH.Apical displacement of tricuspid valve, moderate TR,	13/12/2019 Fetal ECHO - NormaL
			14.12.2019		21/01/20 SLIUG, CA+, cephalic,26 wks, no GCM. Fetal ECHO-Normal
			4/04/20	Ebstein anomaly with VSD (3.5 mm) bidirectional.shunt	
				Ebstein's anomaly,mild TR VSD 3.1 mm with bidirectional shunt	12.3.20 Cephalic : Biometry 32wks CA + Placenta Posterior
			21.4.2020	Ebstein anomaly; TV displacement 2cm, RA/RV mildly dilated,, RVSP-34mmHg, PM VSD	
				Ebstein's anomaly, mild TR, VSD 3.1 mm with bidirectional shunt.	20/04/20 Cephalic Presentation

				3.1 mm present with bidirectional shunt, mild TR present.	BPD-38 wks, HC-39wks, AC-37 wks, FL-37 wks EFW: 3.3 Kg Placenta-anterior, AFI-9.

GTT: Glucose tolerance test; TV: tricuspid Valve; VSD: Ventricular Septal defect;RVSP: Right Ventricular Systolic Pressure; TR: Tricuspid regurgitation; SLIUG: single live intrauterine Pregnancy: AFI: Amniotic fluid index:

DISCUSSION:

Ebstein anomaly was described in 1866 by Wilhelm Ebstein. This was later found to be associated due to lithium consumption during first trimester of pregnancy 3. The anatomical defects described are anterior leaflet of tricuspid valve enlargement and adherence of other leaflets to the endocardium . Presence of ASD, Patent foramen Ovale and shunt between the right and leaft atria are reported in 50%. The present woman had apical displacement of tricuspid leaflet of 2 cm , right atria and ventricle mildly dilated, Perimembranous VSD of 3.1 mm, and bidirectional shunt. Clinically she was asymptomatic till the end of first trimester when she developed breathlessness and haematemesis. Pansystolic murmur was heard on auscultation. Appearance of new symptoms of vomiting and breathlessness during third trimester in pregnancy in a known case of CHD with asthma were described. She had central cyanosis, grade II blowing systolic murmur in tricuspid area and ECG showed sinus rhythm with premature atrial complexes and incomplete RBBB. The condition and cardiac abnormalities were diagnosed after Caesarean section due to persistent desaturation. Right to left shunt and severe tricuspid incompetence with atrialisation of right ventricle and patent foramen Ovale were the findings and surgical correction needed to be undertaken4 The present woman did not have cyanosis and her ECG showed only sinus rhythm and tolerated labour well under epidural analgesia. The severity of tricuspid regurgitation and functional capacity of the right ventricle are the determining factors for good outcome. Physiological changes of circulatory system such as increased heart rate, cardiac output and decreased peripheral resistance usually worsen tricuspid regurgitaion, precipitation of cardiac failure and result in right to left shunting.5 . The degree of displacement of tricuspid valve and symptoms do not correlate. Cyanosis is the poor prognostic factor as complications like pulmonary or systemic thrombosis, supraventricular arrythmias, infective endocarditis, cardiac failure are associated with it.6 Cyanosis also results in poor pregnancy outcomes such as miscarriage, preterm delivery, intrauterine growth restriction and intrauterine death.

Fabio V Lima and collegues reported acute myocardial infarction, cerebrovascular events, embolic events, cardiac failure, arrythmias, postpartum haemorrhage and increased rate of Caesarean section in a cohort of 82 women with Ebstein anomaly7. Connolly and Warnes reported 89% of women achieving vaginal delivery with only 11% requiring caesarean section.8The anaesthetic concerns for management of analgesia during labour in Ebstein anomaly include choosing an appropriate method to maintain preload and after load and prevent right to left shunting and avoidance of tachycardia. Decrease in sympathetic vascular resistance results in right to left shunt and hence in women with severe disease general anaesthesia should be advocated to prevent hypotension. Reduced peripheral vascular resistance associated with epiduaral or spinal can cause hypotension resulting in increase chances of right to left shunt9. This woman tolerated epiduaral analgesia with bupivacaine and Fentanyl well without any hypotension and with good fetal outcome.

Conclusion: Women with Ebstein anomaly with moderate tricuspid regurgitation and small VSD with bidirectional shunt can tolerate labour well with epiduaral analgesia .

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