Title: Case series of pregnancy with Ebstein anomaly and associated maternofetal outcomes: Experiences from a tertiary care

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Introduction:

Ebstein anomaly is a rare congenital heart disease. It contributes to 0.3-0.5% of prevalence and 1% incidence among the congenital heart disease [1]. It's a dysplastic abnormality of tricuspid valve where there is a downward displacement and elongation of the septal and anterior cusp leading to tricuspid regurgitation and complete arterialisation of right ventricle, forming a hypoplastic, thin walled and reduced contractility of the right sided heart [2]. Patients will manifest with variable clinical features due to underlying anatomical and conduction system abnormality such as atrial septal defect (ASD), primary arterial hypertension, ventricular and supraventricular tachycardia and wolf -Parkinson- white syndrome (WPW). The pregnancy may aggravate the complications such as cyanosis, embolic events, congestive heart failure, pulmonary edema and cardiac collapse in Ebstein anomaly [3]. The survival rate in this anomaly is fairly good in asymptomatic patients and they reach old age without affecting their fertility.

The aim of this paper is to address the maternal and perinatal outcomes of women with Ebstein anomaly managed in our tertiary centre.

Case summary:

Case 1:

A 23- year-old G2L1 with a previous normal delivery, who had been diagnosed as Ebstein anomaly during her first pregnancy at 20 weeks of gestation was admitted to our labour ward at 39+4 of gestation in active phase of labour. Ebstein anomaly was detected by in her first pregnancy when she had palpitations aggravating on exercise and doing household chores that subsided after taking rest. She was not on any medications, and her previous antenatal, intrapartum and postpartum periods were uneventful. General physical examination revealed stable vitals and systolic

murmur with splitting of S1. In this pregnancy too, no remarkable cardiac events occurred. She set into spontaneous labour at term , augmented with oxytocin and a live boy, weighing 2.6kg was delivered vaginally with Apgar score of 10 at 1 minute. No postpartum complications were noted and she underwent puerperal sterilisation on postnatal day 3.

Case 2:

A 23-year-old primigravidae at 36+4 weeks was referred to us from elsewhere with severe pre-eclampsia with foetal growth restriction for further management. On physical examination, blood pressure recordings were 140/99 mmHg on antihypertensives with severe features. There was no tachypnea, tachycardia. Cardiopulmonary examination revealed grade 3 systolic murmur in tricuspid area, wide fixed splitting of S1, tapping of apex with no palpable thrills. Induction of labour started with transcervical foley, in active labour patient becomes breathlessness and started desaturating to 85% at room air, the grading of systolic murmur increased to 4 at tricuspid area and air entry reduced. Fine crepitations could be auscultated as well. Ebstein anomaly was diagnosed by electrocardiogram (ECHO) and after diuretics prescription her symptoms improved. Further, we continued the induction of labour under antibiotic coverage. She delivered vaginally a 1.01 kg female baby which was shifted to nursery for very low birth weight. On postpartum day 1 patient developed high grade fever with a tachycardia of 180 bpm and electrocardiogram (ECG) showed supraventricular tachycardia. She was managed medically for the same. We discharged her on postnatal day 6th on antihypertensives and diuretics with an advice to follow up in both postnatal clinic and cardiology department.

Case 3:

A 32-year-old, G2L1 with previous caesarean delivery for a non-recurrent indication, who was incidentally found to have Ebstein anomaly during pre-anaesthesia check-up and hence referred to our emergency facility for multidisciplinary treatment. She was at 40+1 weeks of gestation with gestational hypertension on medication and single live intrauterine fetus in transverse lie. She had remained asymptomatic in her antenatal period, on

examination vitals stable, BP of 134/88 mm of Hg and knee jerk normal, no clubbing or cyanosis were elicited. Chest auscultation revealed loud P2, ejection systolic murmur and splitting of S1. Elective LSCS with concurrent sterilisation was effected under spinal anaesthesia. She delivered a healthy male baby weighing 3.63kg, with a Apgar score of 9 at 10 minutes of birth and she withstood the procedure well. Postnatal blood pressure was was under control, cardiac status was optimal and got discharged on postoperative day 5.

Summary of all cases of Ebstein anomaly:

Table 1. Demographic details of three patients with Ebstein anomaly

Case	**Age**	**OI**	**Age at Diagnosis**	**ECG**	**ECHO**	**Functional assessment of cardiac system**	**Obstetric comorbidities**
1	22	G2L1	First pregnancy	RBBB	Displaced apicals to 25 mm, normal biventricular function, normal RVOT, mild RA dilatation	NYHA 2	-
2	23	G1	Intrapartum period	L-axis deviation, right atrial enlargem	Displaced apicals to 30 mm	NYHA1	Severe Preeclampsiawith Absent End

				ent, tall ‘P’ wave, short RR, -ve delta wave in lead III, avF, +ve delta wave in I and avL and accessory posterolateral pathway	ASD and mild PAH		Diastolic Flow
3	32	G2L1 Previous CS with transverse lie	In second pregnancy at term	R-axis deviation, RBBB	RA, RV dilated, mod-severe TR, RVSP=40mm of Hg,(mod. PAH) septal tricuspid leaflet displaced apically by 25mm	NYHA-I	GHTN

OI- obstetric induction, GHTN-gestational hypertension, RA-right atrium, RV-right ventricle, RBBB- right bundle branch block

Table 2. Maternal and perinatal outcomes in pregnancy with Ebstein anomaly

Case	Gestational age	Mode of delivery	Anaesthesia	Baby Weiwei (Kg)	Antepartum complications	Intrapartum complications	Postpartum complications	Follow up at 6 weeks post natal
1	39+2	VD	epidural	2.6	-	-	Atrial arrythmia	Uneventful
2	37	VD	epidural	1.07	-	Pulmonary edema	PSVT	Uneventful
3	40+1	Em. CS	spinal	3.6	-	-	-	Uneventful

VD-vaginal delivery, Em.CS-emergency caesarean section

Discussion:

Due to the advancement in cardiac facilities, we are achieving good pregnancy outcomes in both mother and neonates. As per the Pinelli and colleagues, the average life expectancy is 30-35 years among the survivors with Ebstein anomaly [4].

The echocardiographic detailing of Ebsteins anomaly reveals that there is a mild to extreme tethering of right atrial leaflet, associated tricuspid regurgitation occurs due to dysplasia, patent foramen ovale (67%), pulmonary atresia (33%), atrial septal defect and anomalous myocardial

band. However, VSD is not common [5]. In this paper, case 2 and 3 had ASD and pulmonary hypertension.

The changes in cardiovascular system during pregnancy, there is aggravation of tricuspid regurgitation which is due to increased stroke volume and right atrial pressure [6].

The pathophysiology behind the arrythmia may be attributed to worsening of tricuspid regurgitation, increased stress to meet the metabolic activity during pregnancy, raised level of catecholamines and maternal hypoxia [7]. Complications in Ebstein anomaly is directly proportional to the severity of the TR. In our first and second case the patient developed atrial arrythmias and supraventricular tachyarrhythmia respectively which were managed medically. It is of concern as arrythmias are more common in pregnancy with Ebstein anomaly. Arrythmias resistant to medical management can be treated by radiofrequency ablation.

Ebstein anomaly in pregnancy is considered as group II as per the modified WHO heart disease classification and tolerates pregnancy well. With this anomaly, patients should be followed up fortnightly in the early trimesters and weekly at third trimester by the team of cardiologist and obstetrician[8]. Multidisciplinary approach including obstetrician, anaesthesiologist, cardiologist, interventionist will help in close monitoring of the pregnancy and foetus and help in early treatment if complications occur. In our second case, patient developed pulmonary arterial hypertension which is another fatal complication of Ebstein anomaly [5].

In this study, case 2 and 3 had preeclampsia necessitating medications, which is commonly seen in underlying cyanosis and chronic hypoxia due to vascular endothelial damage. Nevertheless, in our cases there was no evidence of chronic hypoxia. In preeclampsia, there is an elevated systemic vascular resistance and patient may develop congestive cardiac failure, our second case had manifested with pulmonary oedema [5,8]. Serial ultrasonography must be performed to rule out foetal growth restriction due to chronic hypoxia. Case 1 and 3 were complicated by foetal growth restriction.

Maria Cristina et al reported that Ebstein anomaly is a heterogenetic condition proven by cytogenetic analysis, in which deletion of 1p36 and 8p23.1 chromosomes is seen [9]. An increased incidence of Ebstein anomaly by 6% occurs in offspring's of mothers with same anomaly as per Connolly et al [10]. In all our cases foetal echo was not performed as women presented in late pregnancy but postnatal cardiac evaluation of new-born appeared unremarkable. Few authors have reported right heart dysfunction in babies [11].

While considering labour and delivery in women with Ebstein anomaly, vaginal delivery is the optimal route of delivery in both acyanotic and cyanotic cases and caesarean delivery must be reserved for obstetric indications. Connolly and team reported that labour be closely monitored by involving multidisciplinary team comprising obstetrician, cardiologist, anaesthesiologist and neonatologist. Notable complications include maternal arrythmias, cyanosis and thromboembolism. Therefore, continuous cardiac monitoring, decreased preload and afterload and adequate analgesia should be the goal to prevent congestive cardiac failure during labour. Prophylactic anticoagulation and avoiding the dehydration are advised against thromboembolic events in puerperium and in women with cyanotic heart diseases. Valsalva manoeuvres do increase the intrathoracic pressure subsequently increase the right-to-left shunt, hence cutting short the second stage of labour is recommended [9]. In our study, case 1 and 3 delivered by vaginally and case 2 was delivered by LSCS for obstetric indication.

Conclusion:

Pregnancy with Ebstein anomaly is well tolerated in pregnancy. Cyanotic disease with oxygen saturation 80 percent needs counselling for medical termination of pregnancy as it is associated with higher maternal mortality. Close monitoring of pregnancy is essential with multidisciplinary approach, foetal echocardiography and serial growth scan is the key for an optimized maternal and perinatal outcome.

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