



## Ebstein Anomaly in Neonate: A Case Report

Authors

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### Abstract

**Introduction:** *Ebstein's anomaly is a rare congenital heart disorder occurring in  $\approx 1$  per 210 000 live births and accounting for <1% of all cases of congenital heart disease. In ebstein anomaly there is downward displacement of the septal and posterior leaflets of tricuspid valve into the RV cavity, resulting in functional hypoplasia of RV.*

**Case Report:** *Full term appropriate for gestational age female child born by vaginal delivery, without any antenatal complications, APGAR at birth 8/10, did not require any resuscitation was admitted in NICU in view of cyanosis. Auscultation revealed murmur and saturation was 75-77% in all 4 limbs. Clinical features revealed persistent tachycardia, gallop rhythm, suggestive of congestive cardiac failure. The baby had multiple episodes of supra ventricular tachycardia which responded to anti-arrhythmic agents including intravenous adenosine, intravenous beta blocker, followed by oral beta blocker.*

*The chest x-ray showed cardiomegaly with oligemic lung fields. ECG had tall peaked P wave with RBBB. In 2 D echo there was right atrial dilatation with apical displacement of septal leaflet of tricuspid valve suggestive of ebstein anomaly. Cardiac CT confirmed ebstein anomaly. Anti-failure medication continued and baby was considered for surgical intervention on oral anti-arrhythmic medications.*

**Keywords:** *Congenital heart disease, ebstein's anomaly, supraventricular tachycardia*

### Introduction

Ebstein's anomaly is a rare congenital heart disease that corresponds to less than 1% of all congenital cardiac anomalies. In the general population, the incidence corresponds to 1/210,000 live births, with equal incidence in both sexes, male-to-female ratio being 1:1.<sup>[1,2]</sup> It is characterized by malformations and low deployment of septal and posterior leaflets of the tricuspid valve, showing an area of "atrialized" right ventricle<sup>[3]</sup>. This results in functional hypoplasia of RV. Tricuspid regurgitation is usually present, and can rarely obstruct the RVOT, resulting in dilatation and hypertrophy of RA. It's typical presentation in the neonatal period is

cyanotic newborn with cardiomegaly in x-ray<sup>[4]</sup>. Investigation done initially to diagnose include chest x-ray, ECG and echocardiography. However, the best test for confirmation of the diagnosis is echocardiography as it allow the quantification of severity and prognosis of malformation<sup>[5,6]</sup>.

Newborn present with cyanosis and CHF during the first few days of life in severe cases, and improvement coincides with reduction of the PVR. Conservative techniques are used for these neonates until the pulmonary pressure decline. Each patient's therapy is adapted according to the severity and degree of functional obstruction of the outlet of the right ventricle. Prostaglandins and nitric oxide infusion can be advantageous in more severe

patients<sup>[1]</sup>. The indications for surgical treatment of Ebstein's malformation are not clearly defined and the ideal surgical mode of management remains controversial<sup>[7]</sup>. Early surgical intervention within the first days of life showed good outcome after a modified Starnes operation. Here by reporting case of Ebstein anomaly in which early diagnosis lead to early intervention.

### Case Report

Full term appropriate for gestational age female child born by vaginal delivery, without any antenatal complications, APGAR at birth 8/10, did not require any resuscitation was admitted in NICU in view of cyanosis which appeared few hours after birth. Saturation in all 4 limbs were 75-77%. Baby was started on O<sub>2</sub> by prongs but saturation did not improved with O<sub>2</sub>. On auscultation systolic murmur was present with tachycardia and gallop rhythm, suggestive of heart failure. PGE1 infusion was started along with injection dobutamine and injection Lasix to control heart failure. Chest x-ray showed cardiomegaly with cardiothoracic ratio of 0.68. ECG had tall peaked P wave with RBBB pattern. 2 D echo was done, which showed right atrial dilatation with septal leaflet of tricuspid valve displaced apically by 8mm. There was 7.6mm ostium secundum atrial septal defect with left to right shunt across the shunt. Echocardiography findings were suggestive of ebstein anomaly. PGE1 infusion was stopped. As the pulmonary vascular resistance decreased, cyanosis improved and saturation was about 85-88%. during the nicu stay, baby had supraventricular tachycardia which responded to adenosine. Repeated episodes of arrhythmia occurred. When it did not responded to adenosine, iv b-blocker was given loading dose and then oral propranolol was given as maintenance dose. Arrhythmias were well controlled by propranolol. Cardiac CT was done, which confirmed ebstein anomaly showing tricuspid valve annulus 13mm below the level of mitral annulus, dilated right atrium with atrialization of right ventricle. Also there was pulmonary stenosis at infundibular, valvular and supra valvular levels with

infundibulum measuring 4mm and 8mm ostium secundum. As the baby had severe ebstein anomaly, was referred to cardiac surgery for further management.

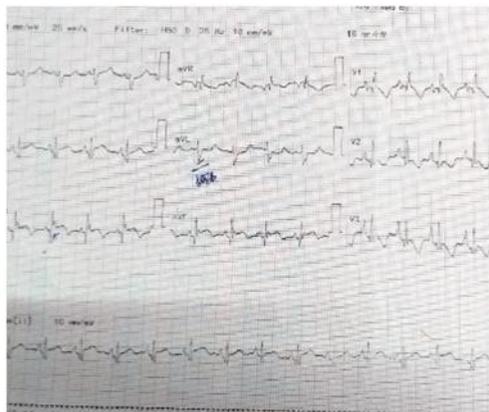
### Discussion

Ebstein's disease is a cardiac anomaly with variable clinical conditions that is characterized by apical displacement of the septal and posterior leaflets of the tricuspid valve. Symptomatic newborns may present with cyanosis and heart failure. Beyond infancy, patients display dyspnea, fatigue, palpitations, exercise intolerance. Diagnosis is difficult in milder cases and can go undetected for many years because patients initially may be asymptomatic. Ebstein have high propensity to develop arrhythmias. Arrhythmias are usually supraventricular and 10-30% of patients have pre-excitation. Ebstein's anomaly may be associated with some abnormality, including atrial septal defect in 90% of patients, pulmonary stenosis and pulmonary atresia. Occasionally in small percentage of cases ventricular septal defect can occur.<sup>[1]</sup>

In this case, the patient had cyanosis with signs of failure with fall in saturation after birth. Oxygen support was given. Oxygen therapy was given to promote the reduction of pulmonary vascular resistance by increasing arterial oxygen saturation and also due to its vasodilating action. These factors are essential in improving gas exchange and the disappearance of cyanosis. PGE1 infusion was started to keep PDA patent until 2 D echo was done. In the investigation of the present case, the chest X-ray showed cardiomegaly (figure 1) and ECG showed tall P waves with RBBB pattern (figure 2).



**Figure 1:** Chest x-ray showing Cardiomegaly

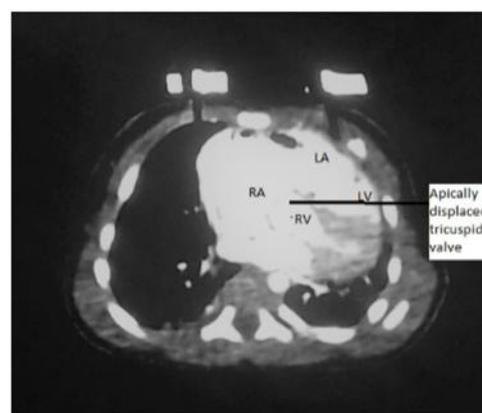


**Figure 2:** ECG showing tall P waves and RBBB pattern

These findings along with clinical picture lead to the suspicion of Ebstein's anomaly. The diagnosis was confirmed by echocardiography which showed right atrial dilatation with apical displacement of septal leaflet of tricuspid valve with ostium secundum atrial septal defect (figure 3). In Ebstein anomaly WPW pre-excitation is frequently associated and predisposes the patient to SVT. In this case patient had repeated episodes of SVT. When SVT did not revert back by adenosine, iv beta blocker (metoprolol) was given and SVT reverted. Patient was started on oral propranolol after which arrhythmia was controlled. Beta blocker has class II antiarrhythmic properties. It usually is administered as a loading dose of 100 to 500 mcg/kg iv over 1 minute followed by a continuous infusion of 50 to 100 mcg/kg per minute. It can be titrated upward by 50-mcg/kg per minute increments every 5 to 10 minutes to a maximum dose of 1,000 mcg/kg per minute until either there is control of the ventricular rate or hypotension develops<sup>[8,9]</sup>. Oral beta blocker such as propranolol is most appropriate for patients responding to esmolol or who have evidence of pre-excitation on ECG. The starting dose of propranolol is 0.25 mg/kg every 6 to 8 hours and can be increased slowly to a maximum of 4 mg/kg per dose with close monitoring of heart rate, heart size, and contractility.<sup>[10]</sup>



**Figure 3:** 2D ECHO showing downward displacement of tricuspid leaflet with atrialised right ventricle.



**Figure 4:** Cardiac CT showing apically displaced tricuspid valve leaflet.

The indications for surgical treatment of Ebstein's malformation are not clearly defined and the ideal surgical mode of management remains controversial.<sup>[8,9]</sup> Some of the indications include severe or progressive cyanosis, RVOT obstruction, progressive RV dilatation, atrial arrhythmias. In this case patient was having RVOT obstruction and recurrent arrhythmias, hence was shifted to cardiac surgery for further management.

### Conclusion

Early diagnosis of Ebstein's anomaly is important for the prognosis of the patient, because 50% of cases can be fatal, especially when symptoms occur during the neonatal period. In this patient early diagnosis led to early management and surgical intervention which prevented complication or neonatal death.

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