



A Case Report of Ebstein's Anomaly First Time Detected in Third Trimester of Pregnancy

KEYWORDS

Ebstein's anomaly, pregnancy

Dr. Mangala Sonavani

HOD and Prof, Department of Medicine, GMCH Aurangabad.

Dr. Kusum Sikariya

JR III, Department of Medicine, GMCH Aurangabad.

Dr. Varun Nagori

Senior resident, Department of Medicine, GMCH Aurangabad.

Dr. G.A.Surwade

Asso. prof, Department of Medicine, GMCH Aurangabad.

Dr. Shailaja Rao

Asst. prof., Department of Medicine, GMCH Aurangabad.

ABSTRACT A 24 year old lady developed breathlessness on exertion at 28th gestational weeks of 3rd pregnancy. She had developed cyanosis at 7th month of 1st pregnancy, hence she was investigated and diagnosed as a case of Ebstein's anomaly on echocardiography.

INTRODUCTION

Ebstein's anomaly is a congenital malformation characterized by a downward displacement of the tricuspid valve into the right ventricle leading to atrialization of the right ventricle with a variable degree of malformation and displacement of the anterior leaflet.¹ Although the clinical manifestations are variable, some patients come to initial attention because of either progressive cyanosis from right to left atrial shunting, symptoms due to tricuspid regurgitation and RV dysfunction or paroxysmal atrial tachyarrhythmias with or without atrioventricular bypass tracts (WPW syndrome).²

CASE REPORT:

A 24 year old female multigravida residing at Soyegaon, Aurangabad with 28 weeks gestation came with complaints of breathlessness on exertion since 15 days. Patient was married in 2008. She delivered her first child in 2010. At the 7th month of gestation during the 1st pregnancy, she developed cyanosis and was investigated and diagnosed to have Ebstein's anomaly. She remained asymptomatic and delivered a female child vaginally at the 9th month. Her second delivery was also asymptomatic.

On examination she was conscious, oriented. She had cyanosis and clubbing. JVP was raised with prominent cv complexes. She had no oedema, lymphadenopathy. Her vitals were stable. Abdominal examination revealed a gravid uterus with fundal height corresponding to 28 weeks of gestation. Cardiovascular system examination showed apex beat in the 6th intercostal space in the midclavicular line, pansystolic murmur associated with a thrill in the tricuspid area, increasing on inspiration.

ECG showed p pulmonale and right bundle branch block. Echocardiography showed severe Ebstein's anomaly with severe tricuspid regurgitation, good LV systolic function, ejection fraction 75 percent and depressed RV systolic function. She was hospitalized and was treated with O₂ inhalation and diuretics.



Clubbing

DISCUSSION

First described by Wilhelm Ebstein in 1866. Ebstein's anomaly is a congenital malformation of the heart characterized by apical displacement of the septal and posterior tricuspid valve leaflets, leading to a part of the right ventricle becoming part of the right atrium with a variable degree of malformation and displacement of the anterior leaflet. This accounts for less than 1 percent cases or 1 per 2,00,000 live births of congenital heart diseases.³ True prevalence is unknown because mild forms often remain undiagnosed. Environmental factors implicated in etiology include maternal ingestion of lithium and benzodiazepine

and maternal history of miscarriage.⁴

A considerable proportion of these patients remain asymptomatic and reach child bearing age. Pregnancy is associated with an increase in stroke volume and heart rate and decrease in peripheral vascular resistance.⁵ In women with Ebstein's anomaly and in the presence of declining right ventricular function, these changes may be poorly tolerated leading to worsening of tricuspid regurgitation, increased right atrial pressure followed by increased right to left shunting.⁶ With good right ventricular function, pregnancy may be well tolerated.

In the present case study, the patient was diagnosed to have Ebstein anomaly in her first pregnancy when she developed cyanosis in the second trimester. She was asymptomatic and delivered successfully in both the earlier pregnancies. Several case reports emphasize the potential complications in pregnancy with Ebstein's anomaly⁽⁷⁻⁹⁾. In two studies on congenital heart diseases, 3 out of 5 women had cyanosis.

Patients with Ebstein's anomaly are liable to develop supraventricular and ventricular arrhythmias and Wolff-Parkinson-White syndrome which occur in upto 20 percent patients.¹⁰ Connolly et al reported in his study that though 14 percent of the women studied had one or more accessory conduction pathways at time of the pregnancy, they are not significant enough to hospital admission or adjunctive medical therapy.¹¹ In our case report patient had no arrhythmias which can be attributed to her relatively good right ventricular function.

Maternal cyanotic congenital heart diseases have been shown to be associated with prematurity and low birth weight and with infant's survival rate of 50-55%.¹² In our study patient had two successful spontaneous vaginal deliveries and infants born to the mother were healthy, full term.

Though pregnancies were well tolerated we advised permanent sterilization in view of her completed family status and unpredictable heart condition.

REFERENCE

1. Ruman U, Chowdhury TS, Nessam et al. A case of pregnancy with Ebstein's anomaly. *Birdem medical journal* 2015 vol.5. | 2. Harrison's Principles of Internal Medicine. 18th edition vol.2 pg 1927. | 3. Attenhofer Jost CH, Connolly HM, Edwards WD, Hayes D et al; Ebstein's anomaly- review of multifaceted congenital cardiac condition. *Swiss Med wly* 2005;135(19-20):269-281. | 4. Correa-Villasenor A, Ferencz C, Neill CA, Wilson PD, Boughman JA; Ebstein's malformation of the tricuspid valve: genetic and environmental factors. The Baltimore Washington infant study group. *Teratology* 1994;50(2):137-47. | 5. Gianopoulos JG. Cardiac disease in pregnancy. *Med Clin North Am*.1989;73(3):p639-651. | 6. Radford DJ, Graff RF, Neilson GH. Diagnosis and natural history of Ebstein's anomaly. *Br Heart J*.1985;54(5):p517-522. | 7. Whittemore R, Hobbins JC, Engle MA. Pregnancy and its outcome in women with or without surgical treatment of congenital heart disease. *Am J Cardiol*.1982;50(3):p641-651. | 8. Donnelly J. E, Brown J.M, Radford D.J. Pregnancy outcome and Ebstein's anomaly. *Br Heart J* 1991;66,p 368-371. | 9. Waickman LA, Skorton DJ, Varner MW, Ehmke DA, Goplerud CP-Ebstein's anomaly and pregnancy. *Am J Cardiol*.1984;15;53(2):p357-358. | 10. Till J, Celermajer DS and Deanfield JE-The natural History of arrhythmia in Ebstein's anomaly. *J Am Coll Cardiol*,19 Suppl A 1992, p 273A. | 11. Connolly HM, Warnes CA-Ebstein's anomaly: outcome of pregnancy. *J Am Coll Cardiol*.1994;23(5):1194-8. | 12. Beischer NA, Mackay EV-Obstetrics and the newborn(2nd edition) WB Saunders, Sydney 1986, pp-406-420. |