Ebstein’s Anomaly – A Rare Cause of Shortness of Breath in Pregnancy

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In October 2009, a 28-year-old woman, G1P0A0, presented at 24 weeks gestation, with a two week history of progressive shortness of breath. Prior to this admission, she had attended for routine antenatal care. She did not have any significant past medical history. On examination, she was not cyanosed. Her blood pressure was 80/40 mmHg, pulse was 100-120, irregularly, irregular and respiratory rate was 35 per minute. Her jugular venous pressure was raised to the ear lobes and she had mild peripheral oedema. Her chest was clear to auscultation. She had audible third and fourth heart sounds along with a 3/6 pansystolic murmur maximally heard in the third and fourth left intercostal spaces parasternally.

Her chest x-ray revealed severe cardiomegaly. Electrocardiogram showed atrial fibrillation incomplete right bundle branch block with right ventricular strain pattern. A CT pulmonary angiogram showed no evidence of pulmonary embolism. Her transsthoracic echocardiogram revealed normal left ventricle, dilated right ventricle with preserved contractility and severe tricuspid incompetence with a severely dilated right atrium. The findings were suggestive of a congenital Ebstein’s anomaly. She was admitted under the joint care of cardiology and obstetric medicine. She was treated with intravenous diuretics, digoxin and therapeutically anticoagulated with subcutaneous Low-Molecular Weight (LMW) heparin. An obstetric ultrasound showed normal foetal anatomy and normal foetal growth.

She was re-admitted to hospital at 28+6 weeks gestation with worsening shortness of breath. Antenatal corticosteroids were administered in the foetal interest. In view of deterioration in maternal symptoms, she was delivered by caesarean section, under general anaesthesia, at 32 weeks gestation. She had a male infant, weighing 1.9 kg, who was admitted to the neonatal intensive care unit for a few days. A transoesophageal echocardiogram during caesarean section showed severe tricuspid incompetence and severely dilated right atrium (Figure 1).

Postpartum, she remained in the intensive care unit for 3 days and was discharged home well 14 days later. She subsequently had a cardiac MRI which confirmed the diagnosis of Ebstein’s anomaly (Figure 2). On review in the clinic 10 weeks postpartum, her exercise tolerance had improved considerably and she was able to manage three flights of stairs fairly easily.

Possible prognostic factors in Ebstein’s anomaly have been associated in several studies and those features associated with poor outcome were:

- a. Other complicating cardiac lesion.
- b. Persistent cyanosis as a result of left atrial shunting.
- c. Diagnosis made in infancy.
- d. Severe cardiomegaly.
- e. Development of dyspnoea at rest.
- f. Right heart failure. 4-5

Pregnancy is well tolerated in women with the mild form of Ebstein’s anomaly (without cardiomegaly, severe cyanosis or arrhythmias).1

Nevertheless, it remains associated with an increased risk of miscarriage, prematurity or congenital heart disease compared to the general population.

The commonest cause of death in Ebstein’s anomaly are considered to be congestive cardiac failure and sudden collapse of undetermined cause. Paradoxical embolism is another reason for high mortality due to increased incidence of thromboembolic disease during pregnancy in these patients.2 Gasul et al reviewed 120 cases and concluded that 59% were dead by 20 years of age, 79% by 30 years and 87% by 40 years.3

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CONFLICT OF INTEREST
None

PATIENT’S CONSENT
Obtained

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Indications for surgical intervention in Ebstein’s malformation include New York Heart Association (NYHA) class III or IV, a decline in exercise tolerance, significant or progressive cyanosis, a cardiothoracic ratio greater than 0.65, right ventricular outflow tract obstruction, a history of paradoxical emboli and refractory atrial or ventricular tachyarrhythmias. 6-9

Our reported patient survived her pregnancy although she had two of the poor prognostic features, i.e. cardiomegaly and right heart failure. She underwent caesarean section at 32 weeks and delivered a healthy baby which was the outcome of an excellent and very close liaison between obstetricians, cardiologists and anaesthetists.

REFERENCES

1 Radford DJ, Graff RF, Neilson GH. Diagnosis and natural history of Ebstein’s anomaly. Br Heart J 1985; 54: 517-22.


