

A Complex Case of Ebstein's Anamoly Presented in Adulthood

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Abstract- Ebstein anomaly (EA) is a rare congenital abnormality involving the tricuspid valve and the right ventricle. Ebstein anomaly occurs when an apical displacement of the tricuspid valve with tethering of leaflet attachments. Both the tricuspid valve and the right ventricle are dysplastic, with it frequently described as "atrialization of the right ventricle." Ebstein anomaly often results in pathological tricuspid regurgitation, right ventricular failure, and arrhythmias. Common clinical manifestations include exertional dyspnea, palpitations, and cyanosis.

Keywords: EA, Tricuspid valve

INTRODUCTION

Ebstein's anomaly is a disease of the entire right ventricle. It is a spectrum of abnormalities, characterized by apical displacement of the valve, anomalous distal attachment of the leaflets, size of the functional right ventricle and degree of tricuspid regurgitation, with alteration in the left ventricle as well. It occurs with a prevalence of about 0.3% to 0.7% among patients with congenital cardiac disease [1] and most cases occur sporadically with an equal distribution between males and females. The anatomical hallmark of this entity is the apical displacement of the attachments of septal and posterior leaflets of the tricuspid valve. The displaced tricuspid valve divides the right ventricle into two parts. The inlet portion is usually integrated functionally with the right atrium ("functional" atrialization) and the apico-trabecular and outlet portions constitute the functional right ventricle. The proximal atrialized ventricle has a thinner wall due to partial absence of myocardium and described as "anatomical" atrialization.

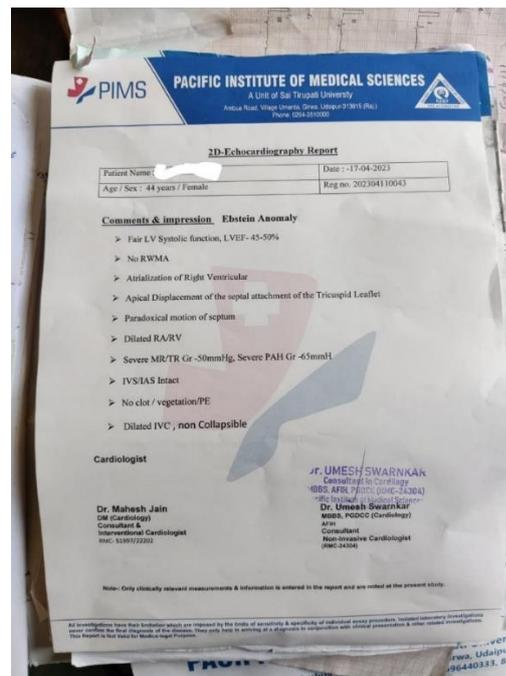
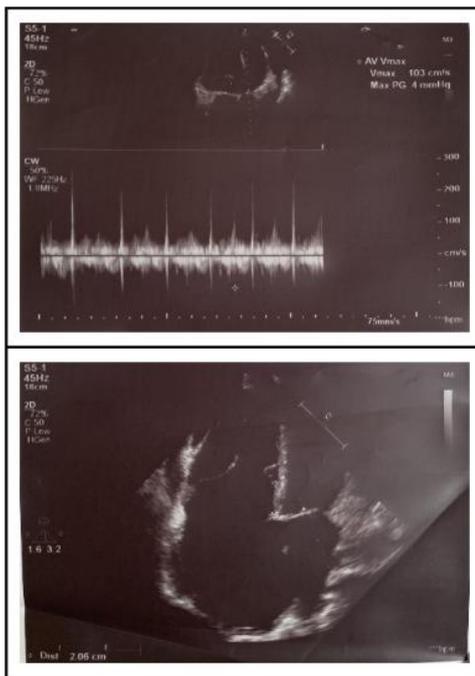
CASE REPORT

A 44-year-old woman followed for Gross ascites with recurrent episodes of heart failure exacerbations. She was admitted to the general medicine department for investigation of portal hypertension. On admission, the patient reported NYHA stage IV dyspnea, palpitations without orthopnea or chest pain. The clinical examination showed a conscious patient normotensive at 120/80 mm/hg, tachycardia at 110 beats per minute, saturation at 95% at free air, raised JVP, facial puffiness, heart sounds well perceived reguar, murmur of tricuspid regurgitation and mitral regurgitation, hepatomegaly, edema of the lower limbs.

Chest x-ray was suggestive of cardiomegaly with pulmonary hypovascularization.



ECHO shows a huge right atrium, atrialization of the right ventricle, severe tricuspid regurgitation, sever mitral regurgitation and mild pericardial effusion.



The patient was managed medically for pericardial effusion, and heart failure. A prophylactic anticoagulant was given as she had a high risk of thromboembolism. After four days of close monitoring and treatment, the patient improved clinically and her vitals were stable. She was discharged with diuretics and oral anticoagulants. She was called on follow-up after two weeks to discuss surgical treatment of EA.

On her follow-up, the patient was counseled about surgical treatment modalities but she wanted to get medical management only. She is continuing on her medications currently.

DISCUSSION

Ebstein's anomaly is a rare form of CHD. Long-term follow-up data for adults with this condition is limited due to the relatively low frequency of the condition and the variation of its anatomic and haemodynamic profile. Current international guidelines are restricted by the lack of available evidence and there are no randomised control studies to inform management strategies.

CONCLUSION

The approach to the management of patients with Ebstein anomaly is determined by the patient's age and clinical presentation including the presence of symptoms of heart failure secondary to tricuspid regurgitation and right heart failure, cyanosis, and right ventricular dilation or dysfunction. The components of management are monitoring, medical management (including temporizing relief of symptoms prior to surgery), management of arrhythmias, and surgical or catheter intervention. Many patients with Ebstein anomaly have no symptoms and require only monitoring while others are symptomatic and require supportive medical therapy and surgical intervention.

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