CASE REPORT
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Sixteen years old Boy with Ebstein's Anomaly

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ABSTRACT

Ebstein anomaly (EA), also known as Ebstein malformation, is a congenital heart defect that affects a small percentage of people. The tricuspid valve does not close properly in people with EA. A 16-years-old boy presented with six weeks history of palpitation. The symptom occurred during rest, 2–3 times per week, lasted up to 30 minutes at a time. After performing all investigations like blood test, ECG, Colour Doppler Echocardiography, chest x-ray etc, final diagnosis is confirmed as EA. Patient was not having any other history of communicable disease, asthma, tuberculosis etc.Patient was admitted to the hospital with the major complaints of difficulty in breathing and generalised weakness, and his situation was critical. After immediate symptomatic management patient's condition was improved and referred to pediatric cardiac surgery department of National Instiitute of Cardiovascular Diseases (NICVD) of Bangladesh for better managment

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INTRODUCTION

The tricuspid valve anomaly described by Ebstein in 1864 consists of apical displacement of the septal and posterior tricuspid leaflets. It results in an enlarged right atrium functionally integrated with the inlet region of the right ventricle ("atrialized" right ventricle). The outlet and trabecular portions of the right ventricle constitute an often hypoplastic functional ventricle. Ebstein anomaly (EA) occurs in 5 per 100000 live births, accounting for 0.5% of all cases of congenital heart disease.[1] Risk factors believed to be associated with the conditions are family history of EA, other congenital heart disease and maternal exposure to benzodiaze-pines or lithium.[1-3] More than 30% of patients with EA have associated cardiac defects Like pulmonary stenosis, Ventricular septal defect (VSD), Atrial septal defect (ASD), Tetra logy of Fallot, Patent ductus arteriosus etc.[1]

The Case

A 16-years-old boy admitted to the Department of Cardiology in North Bengal Medical College, Sirajganj, Bangladesh with the complaints of palpitations, difficulty in breathing and generalized weakness for six weeks. According to the statement of the patients, the symptoms occurred during rest, 2–3 times per week, lasted up to 30 minutes at a time. His respiratory rate and heart rate were 38 breaths per minute and 105 beats per minute respectively. There was no family history of congenital heart disease and taking sedatives during pregnancy. Relevant physical and biochemical examinations revealed no remarkable findings. An electrocardiogram

(ECG) revealed normal sinus rhythm and a Wolff–Parkinson– White pre-excitation pattern. Transthoracic echocardiography demonstrated the presence of Ebstein's anomaly (EA) of the tricuspid valve, with apical displacement of the valve and formation of an "atrialized" right ventricle (Figure 1A, arrow). The anterior tricuspid valve leaflet was elongated (Figure 1B, arrow), whereas the septal leaflet was rudimentary (Figure 1B, arrowhead). After diagnosis and proper emergency management, patient was referred to pediatric cardiac surgery department of National Instiitute of Cardiovascular Diseases (NICVD) of Bangladesh for specific management.

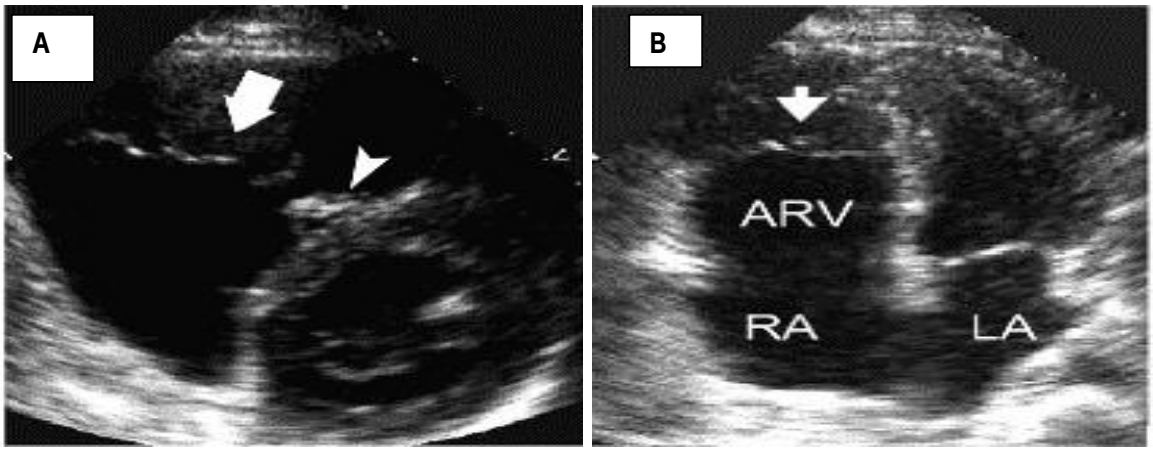

Figure 1: A: Echocardiogram showing elongated anterior tricuspid valve leaflet (arrow) and rudimentary septal leaflet (arrowhead). (RA= right atrium, LA= left atrium.) B: Echocardiogram showing Ebstein's anomaly (apical displacement) of the tricuspid valve (arrow) and formation of an "atrialized" right ventricle (ARV).

DISCUSSION

The clinical manifestations of Ebstein's Anomaly (EA) depend on the degree of tricuspid valve malformation and consequent regurgitation, and any associated cardiac defects.[2,3] Many patients first experience symptoms in adults, but the onset can occur after birth or in infancy or childhood.[4,5] Except for a grade 2/6 holosystolic tricuspid regurgitation murmur (best heard at the left sternal border with inspiratory accentuation), physical examination yielded unremarkable findings.[5]

In newborns, the anomaly often presents as cyanosis and, in the absence of surgical repair, is associated with a 20% mortality in the first year of life. In infants, it may present as congestive heart failure and in children as an incidental murmur. In adults, the anomaly commonly presents with arrhythmias. Factors associated with a worse outcome are young age at diagnosis, male sex, cardiothoracic ratio of more than 0.65 and the presence of cyanosis.[2] The cardiac abnormalities, such as pulmonary valve stenosis or atresia, atrial septal defect, or ventricular septal defect, might coexist with EA. Furthermore, many patients with EA have an additional electrical conduction route in the heart, which can contribute to supraventricular tachycardia (abnormally high heart rate).[6] Few studies reported 50% survival rate of the patients with EA and life expectancy is more in milder form of presentation. [6, 7]

The treatment of Ebstein's anomaly has to be modified to the individual patient. Patients with heart failure and little impairment in functional capacity can be managed medically. Atrial arrhythmias without evidence of pre-excitation can be treated pharmacologically, whereas percutaneous radiofrequency ablation is indicated in the presence of an accessory pathway. In general, surgical intervention with tricuspid valve repair or replacement is restricted

to patients with severe heart failure, cyanosis or intractable arrhythmias.

CONCLUSION

Electrocardiographic evidence of pre-excitation (delta wave on upstroke of QRS complex) is found in up to 0.25% of the general population. The patient should be advised to report any symptoms suggestive of tachyarrhythmia. Patients with Wolff–Parkinson–White syndrome should be referred to a specialist for electrophysiologic evaluation. The presence of pre-excitation and a tricuspid regurgitation murmur should raise the suspicion of Ebstein's anomaly.

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Conflicts of Interest: None.

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