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Ebstein's Anomaly Associated with Atrial Septal Defect Revealed by Palpitations: A Case Report

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Abstract | **Case Report**

Background: Ebstein's anomaly is a rare congenital malformation of the tricuspid valve, frequently associated with atrial septal defect and atrial arrhythmias. Adult presentation is uncommon and often revealed by palpitations. ***Case summary:*** We report the case of a 21-year-old woman with no cardiovascular risk factors who presented with recurrent palpitations. Clinical examination revealed mild cyanosis and hypotension. Electrocardiography showed right atrial hypertrophy and complete right bundle branch block. Chest X-ray demonstrated massive cardiomegaly with a characteristic "rugby ball" appearance. Transthoracic echocardiography confirmed Ebstein's anomaly with severe apical displacement of the tricuspid valve and associated atrial septal defect. Holter monitoring revealed non-sustained atrial tachycardia. The patient improved clinically under beta-blocker and anticoagulant therapy with regular rhythmology follow-up. ***Conclusion:*** Ebstein's anomaly may remain asymptomatic until adulthood and may present with atrial arrhythmias. Echocardiography remains the cornerstone of diagnosis, and early management is essential to prevent complications.

Keywords: Ebstein's anomaly; atrial septal defect; congenital heart disease; atrial arrhythmia; palpitations.

INTRODUCTION

Ebstein's anomaly is a rare congenital heart disease characterized by apical displacement of the septal and posterior leaflets of the tricuspid valve, leading to atrialization of a portion of the right ventricle. It accounts for less than 1% of all congenital heart diseases, with an estimated prevalence of 1 per 20,000 live births [1,2]. First described by Wilhelm Ebstein in 1866, its clinical presentation varies widely, ranging from severe neonatal cyanosis to late adult onset arrhythmias and heart failure [3].

In adulthood, atrial arrhythmias represent the most frequent clinical manifestation, particularly atrial fibrillation, atrial flutter, and atrial tachycardia, due to atrial dilation, abnormal atrioventricular anatomy, and conduction system abnormalities [4–6]. Ebstein's anomaly is associated with interatrial communication in up to 80–94% of cases, increasing the risk of hypoxemia and paradoxical embolism [7]. We report a case of Ebstein's anomaly associated with atrial septal defect discovered during palpitations in a young adult woman.

CASE PRESENTATION

A 21-year-old woman with no cardiovascular risk factors was admitted for recurrent episodes of palpitations with sudden onset and termination. She had been diagnosed with Ebstein's anomaly at the age of 17 after atrial fibrillation episodes. On examination, the patient was conscious with mild perioral and extremity cyanosis. Blood pressure was 90/60 mmHg and heart rate was 100 beats/min. Cardiac auscultation revealed a right-sided gallop rhythm and systolic murmur suggestive of atrial septal defect. There were no signs of right heart failure.

Electrocardiography showed sinus rhythm with right atrial hypertrophy, complete right bundle branch block, and secondary repolarization abnormalities (Figure 1).

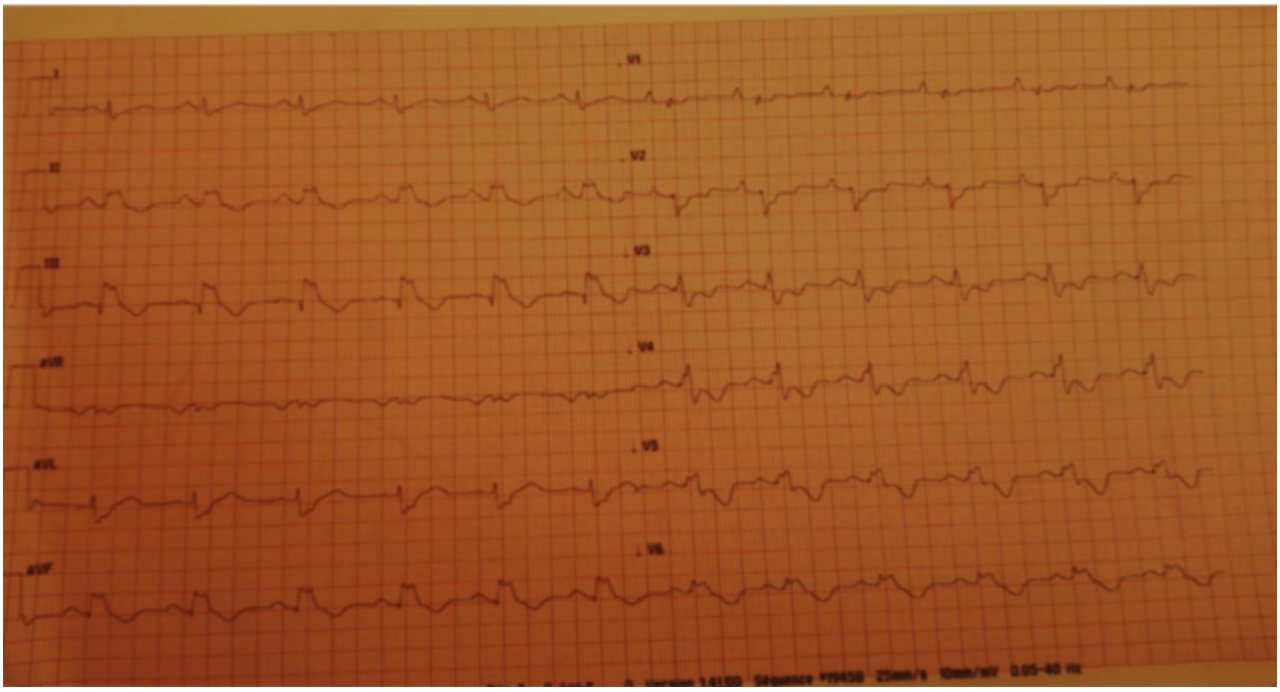

Figure 1: The EKG shows a regular sinus rhythm with axis in place, and appearance of right intra-atrial hypertrophy with intra-atrial conduction block with complete bundle branch block and secondary repolarization disorder a mean ventricular rate of 70 heart rate

Chest X-ray demonstrated massive cardiomegaly with a supradiaphragmatic apex and "rugby ball" configuration (Figure 2). Laboratory assessment revealed secondary polycythemia with hemoglobin of 17 g/dL and hematocrit of 51.7%.

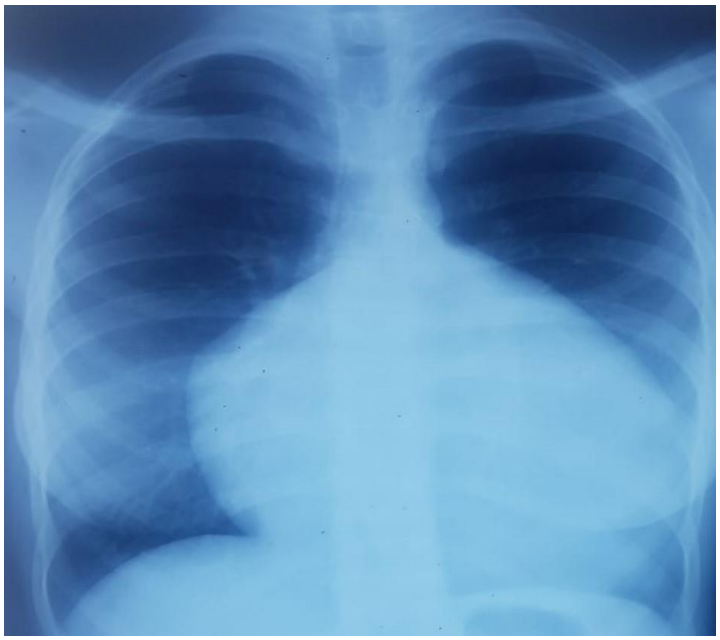

Figure 2: Chest X-ray shows massive cardiomegaly with supra-diaphragmatic point, and right overhang with rugby ball appearance

Transthoracic echocardiography confirmed Ebstein's anomaly with marked apical displacement of the posterior and septal tricuspid valve leaflets, resulting in a large atrialized right ventricle and severe right atrial dilation (combined surface area 75 cm^2). A secundum atrial septal defect with predominant left-to-right shunt was identified, with minimal tricuspid regurgitation (Figures 3–6). Holter monitoring revealed non-sustained atrial tachycardia with permanent complete right bundle branch block.

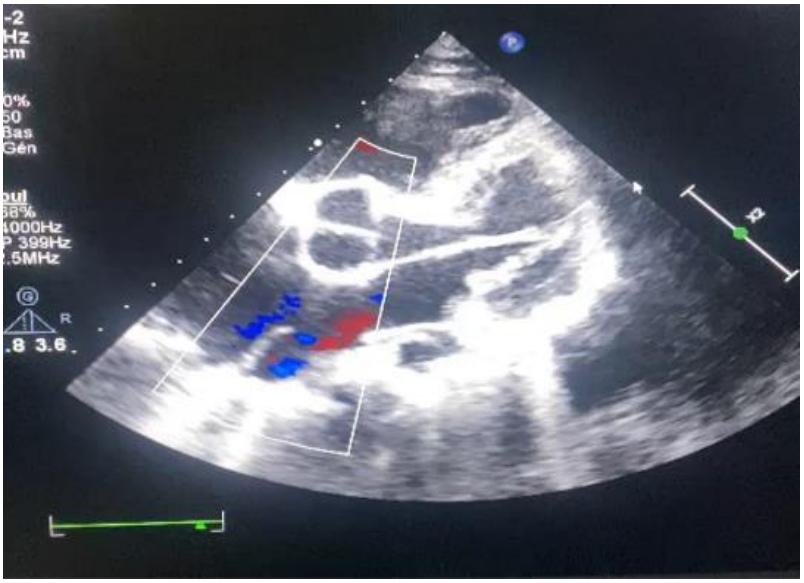

Figure 3: Parasternal short axis section on echocardiography showing the appearance of Ebsetin's disease with an interatrial communication with predominantly left-right shunt

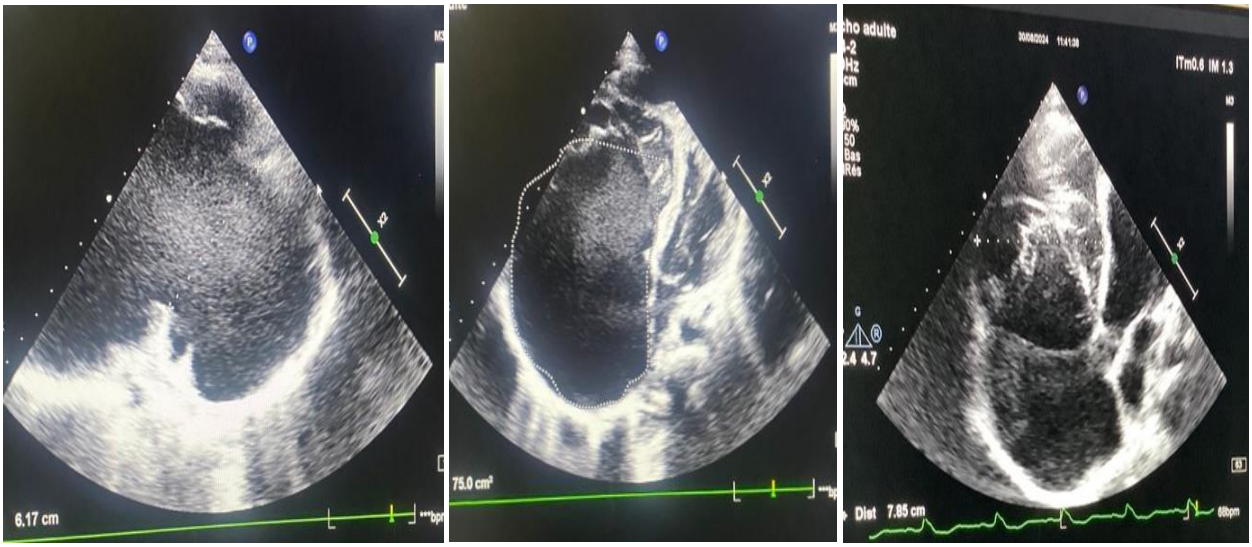

Figure 4+5+6= on transthoracic echocardiography Aspect of Ebstein's disease: The posterior tricuspid valve presents a clear defect of elimination with an insertion that is too low apically, to a lesser degree the septal tricuspid valve, large right atrium and atrialized chamber with surface area of 75cm², the remaining right ventricul is amputated of a large part of its admission chamber, good outflow chamber is trabuculated, it appears dilated with telediastolic diameter of the RV: 81mm

The patient was treated with beta-blockers and anticoagulation, with favorable clinical evolution and referral for regular rhythmology follow-up.

DISCUSSION

Ebstein's anomaly encompasses a wide spectrum of anatomical and clinical presentations. Adult patients frequently present with palpitations, dyspnea, cyanosis, or right heart failure, with supraventricular arrhythmias being the predominant manifestation [4,8]. The pathophysiology of atrial arrhythmias in Ebstein's anomaly relates to marked right atrial dilation, accessory pathways, atrial scarring, and abnormal atrioventricular junctions [5,6].

Atrial fibrillation has become the most frequent arrhythmia in adults with congenital heart disease, replacing re-entrant tachycardias observed in younger patients [9]. Interatrial communication is present in approximately 80–94% of patients with Ebstein's anomaly and contributes to hypoxemia, paradoxical embolism, and increased stroke risk [7,10].

Echocardiography remains the cornerstone of diagnosis, allowing accurate assessment of leaflet displacement, degree of atrialization, right ventricular function, tricuspid regurgitation severity, and associated anomalies [11]. Cardiac magnetic resonance imaging provides complementary anatomical and functional assessment and is increasingly recommended for preoperative evaluation [12].

Management is individualized and depends on symptom severity, arrhythmia burden, ventricular function, and cyanosis. Medical therapy includes rate or rhythm control and anticoagulation. Catheter ablation may be challenging due to abnormal anatomy but is feasible in specialized centers [13]. Surgical repair or valve replacement is reserved for symptomatic patients with progressive right ventricular dysfunction or refractory arrhythmias [14].

CONCLUSION

Ebstein's anomaly is a rare congenital heart disease that may remain asymptomatic until adulthood and present with atrial arrhythmias. Early recognition through echocardiography and multidisciplinary management are essential to prevent complications and improve long-term outcomes.

Figures

- **Figure 1.** Electrocardiogram showing sinus rhythm with right atrial hypertrophy and complete right bundle branch block.
- **Figure 2.** Chest X-ray demonstrating massive cardiomegaly with characteristic "rugby ball" appearance.
- **Figure 3.** Parasternal short-axis echocardiographic view showing Ebstein's anomaly associated with atrial septal defect and predominant left-to-right shunt.
- **Figures 4–5-6.** Transthoracic echocardiographic views demonstrating apical displacement of the tricuspid leaflets, enlarged right atrium, atrialized right ventricle, and reduced functional right ventricular cavity.

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