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Ebstein's Anomaly between Valve Damage and Cardiac Arrhythmia

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Abstract **Case Report**

Ebstein's anomaly (E.A) is a congenital malformation that represents less than 1% of all congenital heart disease. They are predisposed to arrhythmias because of abnormal cardiac anatomy. We report the case of a 60-year-old female with a history of early infancy diagnosis of E.A, admitted for tachycardia with wide QRS complex. An electrophysiology study revealed an atrial rhythm and programmed atrial stimulation induced atrial tachycardia identical to the clinical tachycardia with multiple circuits. Radiofrequency ablation was not performed because of technical difficulties. At two months follow-up, the patient's cardiac condition worsened and echocardiography revealed worsening tricuspid regurgitation. She underwent surgical repair. The postoperative course was uneventful. The diagnosis of this anomaly is mainly made by transthoracic echocardiogram, which constitutes the cornerstone of imaging examinations. Diagnosis is made when there is an apical displacement of the tricuspid septal leaflets past 8mm/m². Cardiac arrhythmias are frequent with Ebstein malformation; sudden death and accessory pathway fast-conducting AV with anterograde and retrograde conduction are commonly described in most patients. While, wide-QRS tachycardia, ventricular tachycardia (VT), or flutter through the para-septal pathway may occur. The management of tachyarrhythmias associated with E.A remains a real challenge. Even if it can be difficult given the atrial dilatation, electrophysiological exploration with possible radiofrequency ablation of symptomatic supraventricular tachyarrhythmias remains necessary. Otherwise, supraventricular tachyarrhythmia in E.A, can also be ablated surgically.

Keywords: Ebstein's anomaly, tachycardia, Wide QRS complex.

1. INTRODUCTION

Ebstein's anomaly (E.A) is a congenital malformation that represents less than 1% of all congenital heart disease [1]. Patients with E.A are predisposed to arrhythmias because of abnormal cardiac anatomy. We report a case of wide complex tachycardia in a patient with this anomaly.

2. CASE REPORT

A 60-year-old Caucasian female with a history of early infancy diagnosis of E.A. Presented with dyspnea and palpitations. On admission, she was at New York Heart Association (NYHA) functional class II. SBP of 120/80 mm Hg and heart rate of 200 beats per minute. In auscultation, we found a holosystolic murmur at the low left sterna border. The 12-lead electrocardiogram (ECG) during tachycardia disclosed a wide QRS tachycardia and a left bundle branch block (LBB) morphology with late precordial transition and left axis deviation (Figure 1). The tachycardia was resolved spontaneously, and the control ECG tracings suggested a sinus rhythm with right bundle branch block (RBB), and no signs of ventricular preexcitation (Figure 2). Transthoracic echocardiogram (TTE) showed the presence of E.A, with an apical displacement of the septal and posterior leaflets (15mm/m2) with severe tricuspid regurgitation (TR) and a huge right atrium dilatation (area = 70cm2) (Figure 3). The systolic function of both ventricles was preserved. An electrophysiology study was performed revealing atrial tachycardia with 2:1 anterograde atrioventricular (AV) conduction, mimicking a sinus rhythm (Figure 4). Atrial stimulation stopped atrial tachycardia but several atrial tachycardias of different origins were induced by atrial stimulation. 1:1 atrial tachycardia developed with isoproterenol. Programmed atrial stimulation induced atrial tachycardia with 1:1 conduction identical to the clinical tachycardia (Figure

5). Programmed ventricular stimulation performed up to 3 extra stimuli in the control state and after isoproterenol was negative. After exercise test, the atrial rate increased somewhat with minimal degrees of AV block and ventricular rate elevation (1:1 conduction). The conclusion that was retained, is that the initial ECG represented an atrial tachycardia with LBB aberrancy conducted 1:1 and that the control ECG tracing which was thought to represent a sinus rhythm was atrial rhythm with 2:1 AV conduction.

The patient underwent cardiac magnetic resonance imaging (CMR), which objectified normal left ventricle (LV) and right ventricle (RV) systolic function, a huge right atrium, and apical displacement of the tricuspid valve leaflets (Figure 6).

Radiofrequency ablation was not performed because it was judged not feasible. Combination therapy with Digitalis and Amiodarone therapy was instituted to control heart rate and for medical conversion. At two months follow-up, dyspnea worsened to NYHA class IV despite a sinus rhythm and TTE showed a new aggravation of TR with lack of leaflet coaptation. She had repair surgery with a tricuspid valve and a combined total cavo pulmonary connection with a fenestrated RV exclusion technique. She remained hemodynamically stable during hospitalization and at discharge with an LV ejection fraction of 57%, and at later follow up she remains healthy with an improvement of NYHA class, and sinus rhythm on amiodarone.

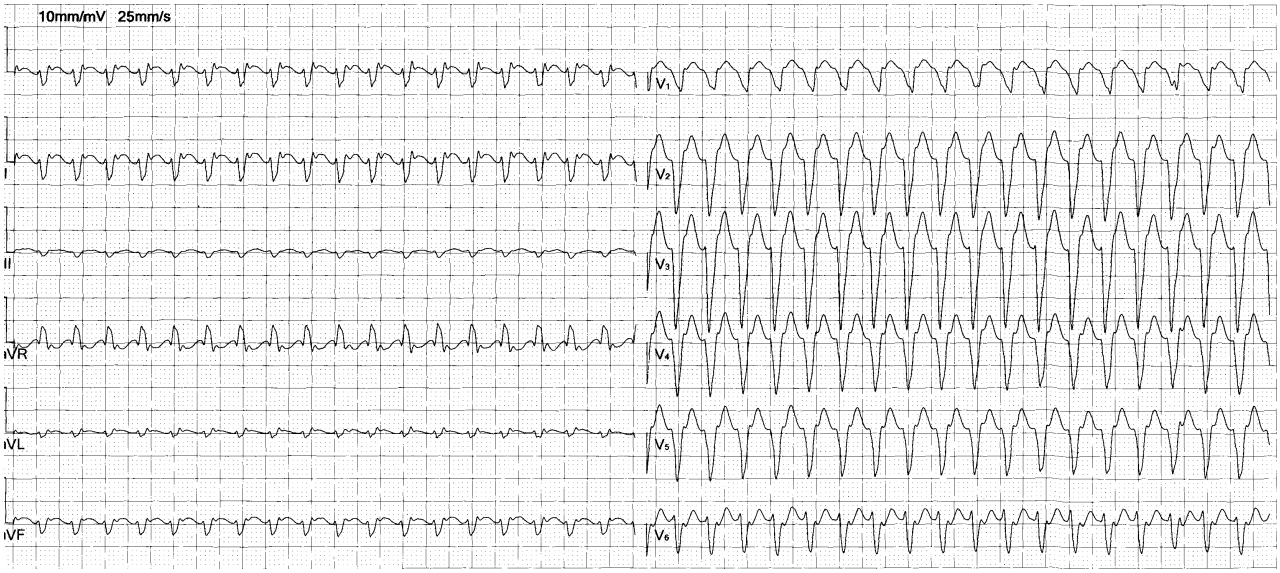

Figure 1: The 12-lead ECG during tachycardia: wide QRS tachycardia with left bundle branch (LBB) morphology and left axis deviation

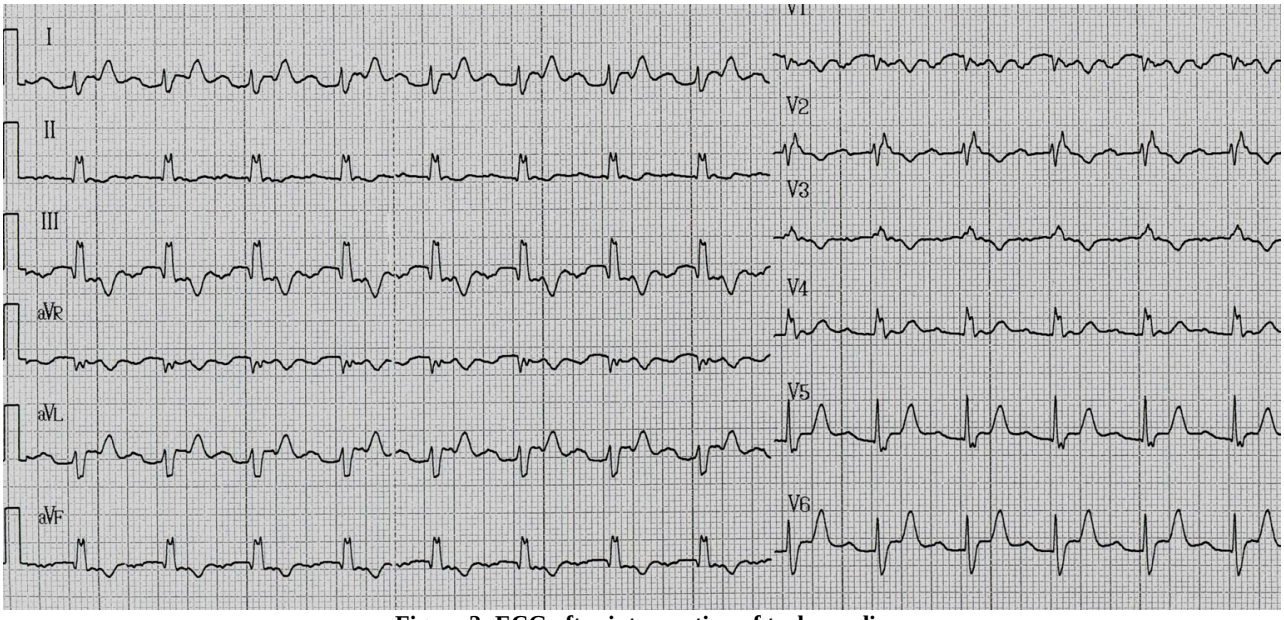

Figure 2: ECG after interruption of tachycardia

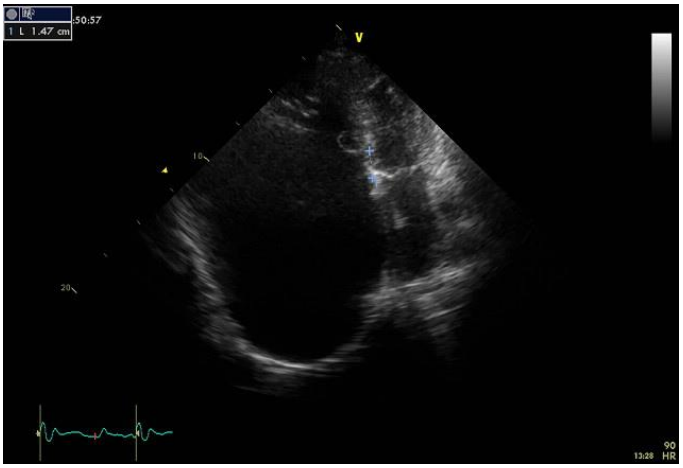

Figure 3: Transthoracic echocardiography; apical 4 chambers view; apical displacement of the septal and posterior leaflets of the valve (15mm/m2) and marked enlargement of the right atrium

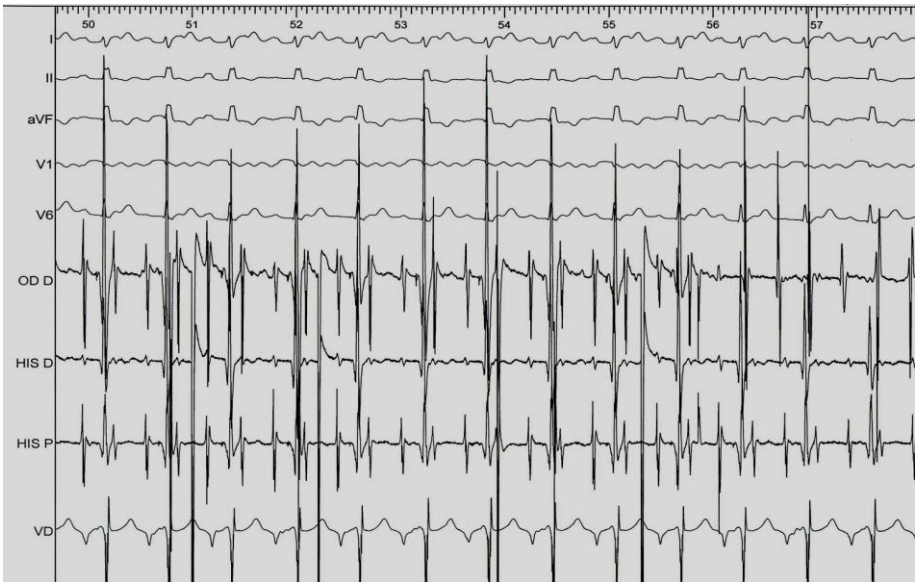

Figure 4: Intracardiac recordings indicated an atrial tachycardia with 2:1 conduction

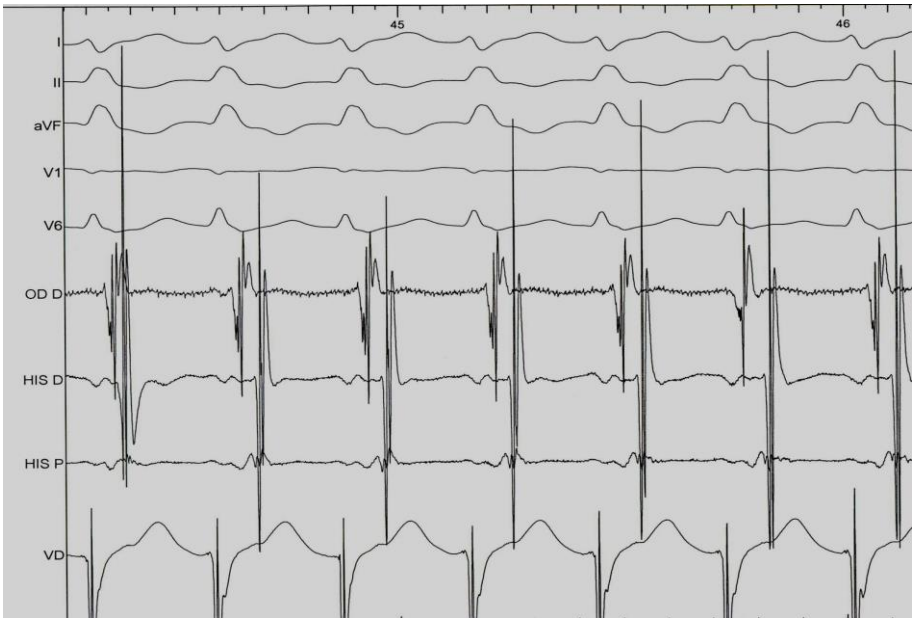

Figure 5: 1:1 atrial tachycardia during isoproterenol infusion

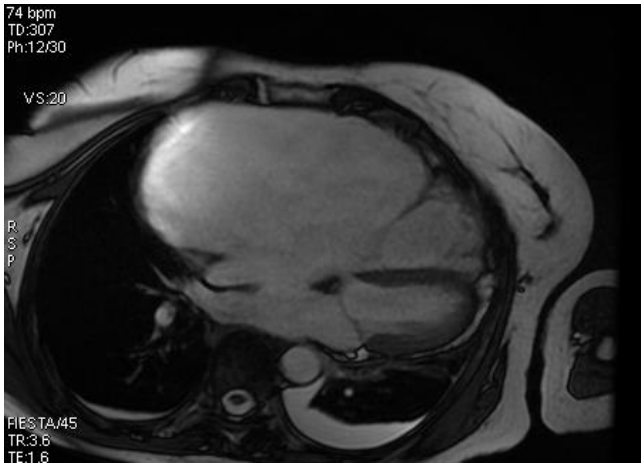

Figure 6: Cine CMR at end diastole in the long-axis 4-chamber View (3T scanner GE): Normal RV systolic function with large right atrial enlargement

3. DISCUSSION

Ebstein malformation is a rare congenital heart malformation. This anomaly is found in about one in 20,000 live births. It accounts for less than 1% of all congenital heart malformations [1]; only 5% of patients without surgical correction survive over 50 years of age [2]. Adults often present with worsening cyanosis, exercise limitation capacity, or cardiac arrhythmias [3, 4]. In this case, a 59-year-old patient presented with NYHA functional class II dyspnea and extensive complex tachycardia. E.A is characterized by septal or posterior leaflet of the tricuspid valve apical migration, causing a kind of "atrialization" of the R inflow tract and consequently, an RV of variable size but functional. This morphology causes different degrees of TR, leading to right atrium dilatation. The diagnosis of this anomaly is mainly made by TTE, which constitutes the cornerstone of imaging examinations. Diagnosis is made when there is an apical displacement of the tricuspid septal leaflets past 8mm/m² [5]. In our case, the apical displacement was 10 mm/m². Other abnormalities are observed, the leader of which atrial septal defects represent 80 to 94% [6, 7]. Other concomitant abnormalities described are atretic or bicuspid aortic valve abnormalities, hypoplastic or atretic pulmonary artery, coarctation, subaortic stenosis, mitral valve prolapse, ventricular septal defect and pulmonary artery stenosis. [8]. None of these abnormalities were observed in our patient.

Cardiac arrhythmias are frequent with Ebstein malformation; sudden death is relatively common since it is reported in 2.6% of cases [9]. And tricuspid valve repair does not prevent sudden death [10].

On the other hand, accessory pathway fast-conducting AV with anterograde and retrograde conduction are commonly described in most patients [11]. While, wide-QRS tachycardia, ventricular tachycardia (VT), or flutter through the para-septal pathway may occur. Also, atrial, tachycardia, flutter, and fibrillation could be seen [11, 12].

The apical migration of the septal leaflet of the tricuspid valve, and the loss of continuity between the central fibrous body and the septal AV ring leads to direct muscular connections, thus creating a potential anatomical substrate for accessory pathways and pre-excitation. The latter is described in 6% to 36% of patients with E.A [9, 11, 12]. But neither of these accessory pathways were revealed in our patient.

Atrial flutter and fibrillation are probably due to tricuspid valve insufficiency which leads to right atrial dilatation and promotes the onset of premature atrial contractions. On the other hand, it can be induced by right atrial wall abnormalities in post cardiac surgery or after incisional atrial tachycardia [11].

According to these results, most authors considered that palpitations and sudden death are largely induced by the supraventricular tachyarrhythmia in this anomaly. The implication of ventricular arrhythmias in sudden death in patients with E.A has not been defined. Even if many cases have been described during heart catheterization, or during catheter manipulation in the abnormal part of the RV. Electrophysiological studies showed that the atrialized RV could be particularly irritable [13]. The anatomical substratum for VT occurrence on E.A is supported by the presence of a histopathologically abnormal RV myocardium, a finding established in several studies [14]. But, in our case, the programmed ventricular pacing was negative. The management of tachyarrhythmias associated with E.A remains a real challenge [15]. Even if it can be difficult given the atrial dilatation, electrophysiological exploration with possible radiofrequency ablation of symptomatic supraventricular tachyarrhythmias remains necessary in these patients. The search for anatomical landmarks is difficult and targeting the AV junction with a catheter can be challenging [16]. In our case, radiofrequency ablation of the multiple ectopic atrial foci was judged not feasible because of right atrium dilatation and the severe TR leading to catheter ablation instability.

Otherwise, Supraventricular tachyarrhythmia in E.A can also be ablated surgically [17, 18]. That said, the results are less convincing in flutter or atrial fibrillation regarding right and intraoperative atrial scars, which promotes atrial arrhythmias and their recurrence which can reach 25%, but which can be reduced to 7% with associated drugs [16].

4. CONCLUSION

Regular wide QRS complexes tachycardia was suggestive of VT. ECG after interruption of tachycardia mimicked a sinus rhythm. The diagnosis of 1:1 atrial tachycardia was retrospectively made. The surgical correction of TR was associated with the clinical improvement of the patient and the arrhythmia was only treated with beta-blockers.

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