Case Report

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Ebstein's Disease: About Two Cases Discovered Following a Rhythm Disorder

Z. BENNOUI[1], M. ABDELBAKI[2]

[1]*Department of Cardiology, Tipaza Hospital, ALGERIA.*

[2]*EHS Maouche Amokrane, ALGERIA.*

***Corresponding Author**

M.ABDELBAKI, Department of Cardiology, EHS Maouche, Algeria.

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Abstract

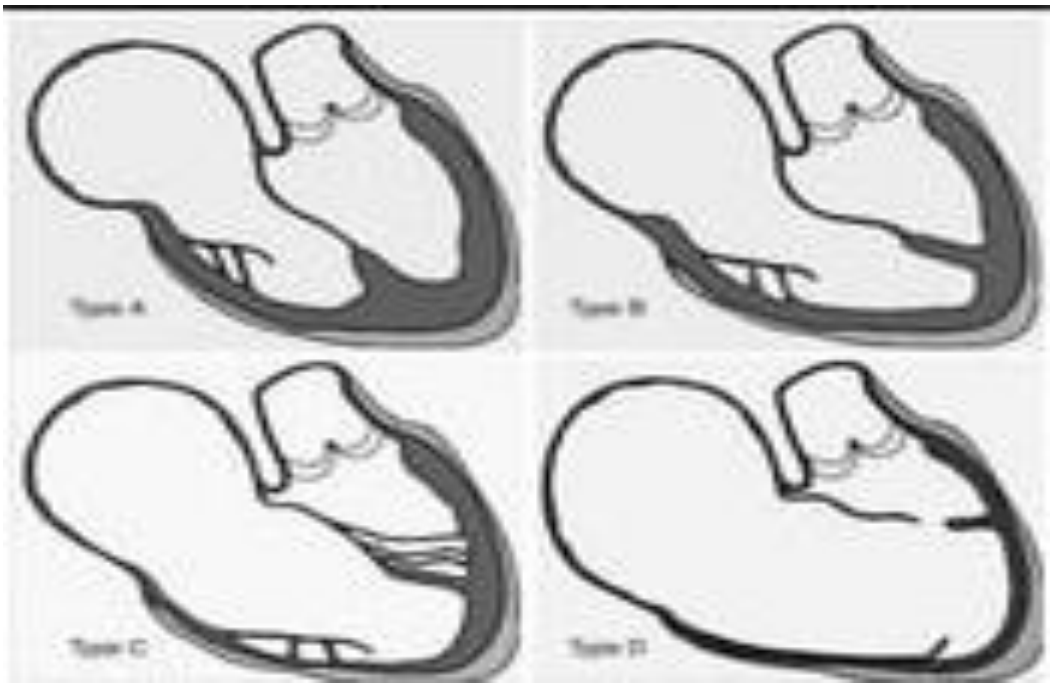
Ebstein's disease is a rare congenital heart disease. It is characterized by the low insertion of one or two tricuspid leaflets leading to atrialization of the right ventricle. The clinical forms vary, ranging from neonatal anasarca to right heart failure and arrhythmia in adolescents and adults. It can remain asymptomatic for a long time, and the discovery is fortuitous following a preoperative assessment for extracardiac surgery or other reasons. We report two clinical cases of Ebstein's disease, different in their circumstances of discovery: in the first case, the disease was discovered following an assessment of right bundle branch block with late left extrasystoles in a 20-year-old patient, and the second case, during an assessment of palpitations in a 13-year-old patient. Through these cases, we will review the circumstances of discovery of the disease by recalling the different types of cardiographic echo and therapeutic management.

Keywords: Ebstein's disease, Circumstance of discovery, Echocardiographic types, Heart failure, Rhythm disorders

Introduction

Ebstein's disease is an incomplete resorption of myocardial tissue which occurs in the septal and posterior leaflet of the ventricle septum, it never reaches the tricuspid ring. This abnormal arrangement displaces the tricuspid annulus apically, causing atrialization of a portion of the right ventricle which becomes hypoplastic and dysfunctional [1,2]. The clinical picture depends on the age of discovery and the magnitude of the anatomical defect, pulmonary blood flow, concomitant septo-atrial defect, development of arrhythmias and degree of RV dysfunction [3-6]. The most frequently identified ECG findings are: HAD, first degree BAV (25%), BBD (75%) due to fibrosis of the right branch of the ICH bundle, and ventricular preexcitation (25%). due to an incomplete tricuspid ring. Carpentier's classification makes it possible to distinguish four anatomical and functional types (A, B, C, D) by taking into consideration the tricuspid insertion, the size of the atrialized RV, and the function of the remaining RV [9]. Patients require regular clinical and echocardiographic monitoringin order to make the surgical indication at the appropriate time.This is indicated in the event of the occurrence of heart failure, arrhythmia or echocardiographic changes. It consists of a repair or replacement of the tricuspid valve, associated or not with a total or partial cavopulmonary anastomosis [11-13].

Carpentier classification [3]



Type A: Small contractile atrialized chamber with a large and mobile anterior valve, good ventricular volume

Type B: Significant displacement of the septal and posterior valves, large atrialized chamber of normal contractility or not with an anterior valve of reduced movement, small right ventricle

Type C: Reduced movements of the anterior valve which appears as a fibrous remnant, well-demarcated commissures, stenotic tricuspid orifice with hypoplastic septal and posterior valves, very reduced right ventricle and large and non-contractile atrialized chamber, sometimes aneurysmal. We can find atrial muscular bands which join the ventricular wall responsible for conduction disorders.

Type D: "tricuspid sac", the valves are adherent to the ventricular wall, the atrialized chamber is difficult to identify, the wall of the thin ventricle is not very contractile.

First Case

13-year-old patient, with no particular history, consulted the cardiology emergency room for palpitations, which had been present for 3 months, aggravated by the efforts made while playing school sports. Clinical examination: Audible heart sounds, systolic murmur of 2/6th of tricuspid insufficiency, absence of added noises. The ECG shows RSR at 90 cpm, PR at 120 ms, fine QRS, absence of rhythm disturbances.

Chest x-ray: Normal.

Cardiac Doppler echo is in favor of Ebstein's disease type B of the Carpentier classification with low insertion of the septal tricuspid valve, atrialization of part of the RV; good function of the remaining RV; IT grade II; good LV function; absence of left valve disease; absence of other associated malformations.

48-hour Holter ECG: RSR, average HR 85cpm. Several episodes of intermittent ventricular preexcitation.

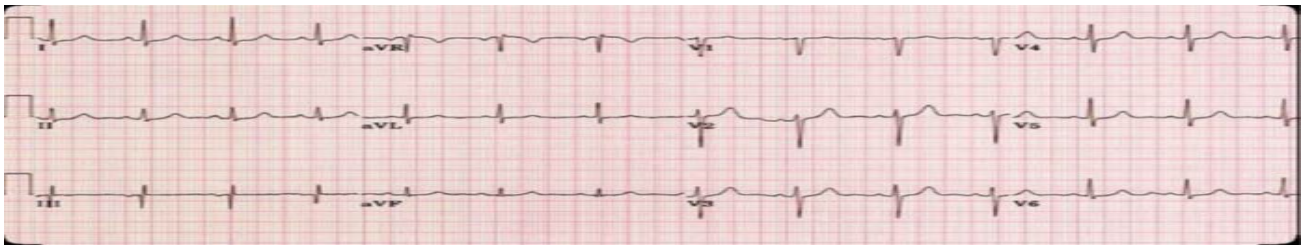
Second Case

Patient aged 20, referred for cardiovascular assessment in the context of employment, following a pathological ECG (BBD according to the referring doctor). The clinical examination and chest x-ray were unremarkable.

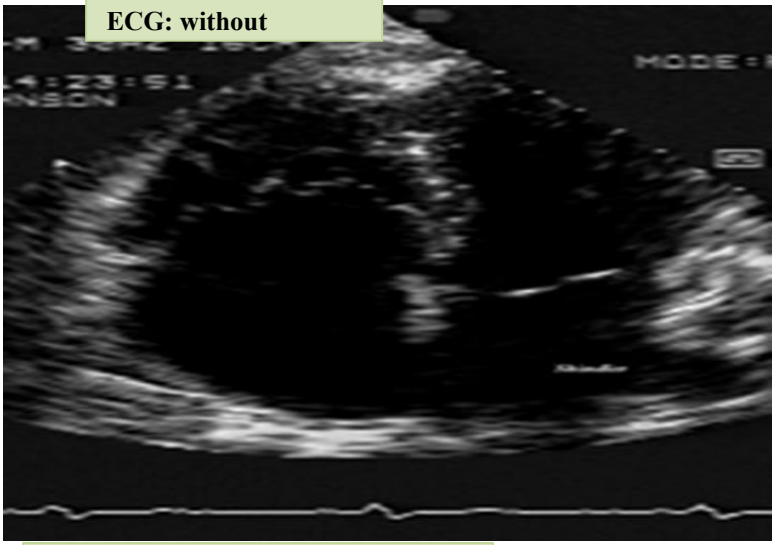
ECGregisters in RSR at 90 cpm, PR at 100 ms with positive delta wave lateral and anteroseptal, and negative inferiorly, in relation to a left lateral accessory pathway.

Echocardiographydemonstrated an Ebstein type B anomaly, without tricuspid leak or other associated anomalies.

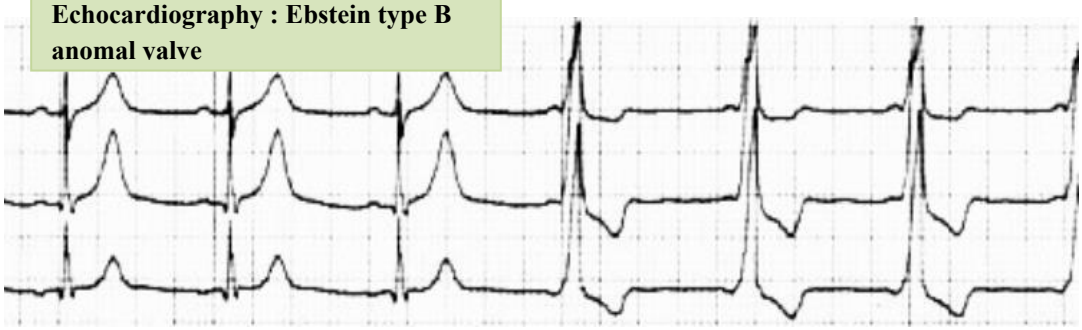
Characteristics of Patient 1



ECG: without

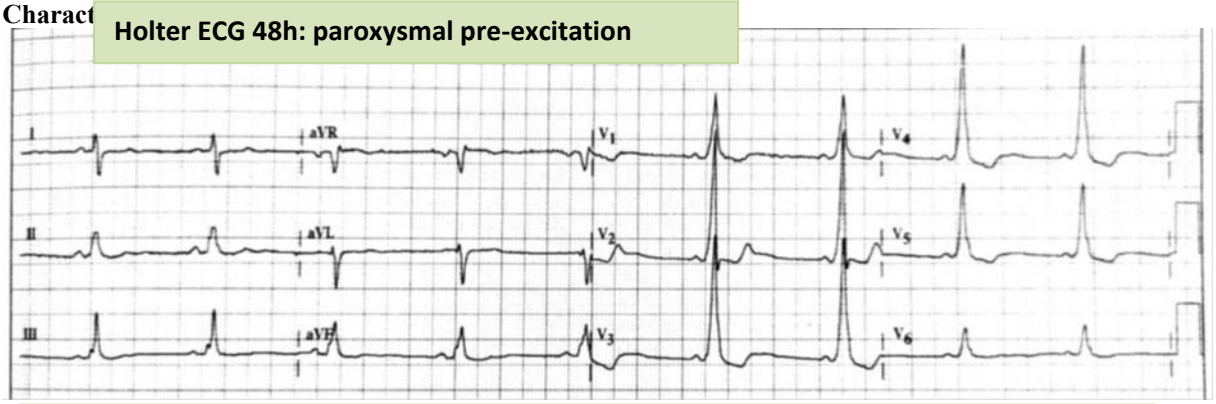


Echocardiography : Ebstein type B anomal valve

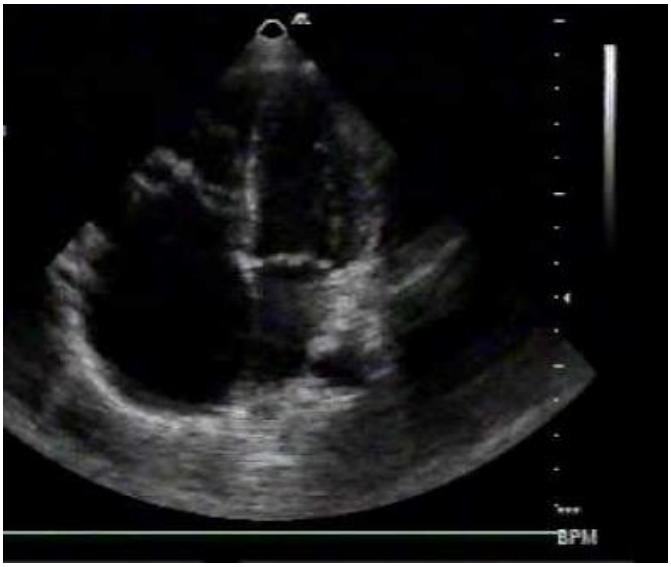


Charact

Holter ECG 48h: paroxysmal pre-excitation



ECG: lateral and anteroseptal delta wave: left lateral accessory beam



Echocardiography: Ebstein type B anomaly

Discussion

Patients may present with several clinical presentations depending on the type of disease [3-6]. These may be hemodynamic, embolic, infectious or rhythmic complications. Arrhythmias have been described to occur in patients throughout their life, linked to congenital and acquired arrhythmogenic substrates [8]. These arrhythmias are in the majority of cases of the tachycardia type, most of these tachycardias are based on accessory pathways located along the abnormal atrioventricular valve (30%). Several types are found such as ectopic atrial tachycardia, supraventricular extrasystoles, atrial fibrillation, atrial flutter and ventricular tachyarrhythmias [8]. The occurrence of sudden death in patients with the disease is mainly linked to atrial fibrillation in the presence of WPW syndrome. Antiarrhythmic drugs are often insufficient to control or avoid recurrences, particularly in the long term [3,4]. These drugs are potentially arrhythmogenic, leaving room for interventional electrophysiology. In congenital heart diseases with rhythm disorders, ablative techniques are rarely used compared to their use in healthy hearts, due to anatomical modifications [3,4].

As for the practice of sport, given that the disease affects young people, competitive sport is strongly discouraged in severe forms due to the risk of occurrence of arrhythmias and sudden death [11,15].

Conclusion

Ebstein's disease is a rare congenital heart disease. The clinical and anatomical forms are variable. It can remain asymptomatic for a long time, and must be sought in the event of an arrhythmia and particularly during ventricular pre-excitation, for optimal early management before the onset of right ventricular dysfunction.

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