Journal Homepage: -www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI:10.21474/IJAR01/19650
DOI URL: http://dx.doi.org/10.21474/IJAR01/19650

RESEARCH ARTICLE

EBSTEIN'S ANOMALY: A RARE CARDIAC DISORDER UNVEILED BY SUDDEN ARRHYTHMIA IN A PREVIOUSLY STABLE ADULT

M. Eddahbi, W. ID El Mouden, A. Sebbar, W. Beladel, A.K. Elbaz, M. Berrajaa and M.El Minaoui
Cardiology Department University Hospital Agadir, Faculty of Medicine & Pharmacy IbnZohr University, Agadir, Morocco.

Manuscript Info

Manuscript History
Received: 15 August 2024
Final Accepted: 18 September 2024
Published: October 2024

Key words:-
Ebstein's Anomaly, Junctional Tachycardia, Adult Congenital Heart Disease

Abstract

Ebstein's anomaly (EA) is a rare congenital heart disease which occurs in approximately 1 in 200,000 live births and represents less than 1% of all cases of congenital heart disease(1). AE is a rare cardiopathy of the right ventricle that originates in an embryological peculiarity of the tricuspid valve responsible for over-apical implantation of the septal leaflet of the tricuspid valve (2). We report the case of a 47-year-old patient with asymptomatic EA, admitted with junctional tachycardiaat 200bpm and haemodynamic instability. Due to this instability, we administred a synchronous external electric shock at 150 joules after sedation. The patient immediately recovered regular sinus rhythm and achieved haemodynamic stability. Echocardiographic evaluation supported the of EA, showing apical displacement of the tricuspid valve and a mitro-tricuspid mismatch of 11.6 mm/m2, The septal leaflet exhibited reduced mobility, attached to the interventricular septum, and the right atrium was dilated due to right ventricular atrialization. The clinical presentation of EA varies from neonatal respiratory distress to right heart failure and rhythm disorders in adolescents and adults. Some cases exhibit exceptionally long survival. Management of EA depends on its anatomical form and clinical presentation (3). Patients with minor tricuspid valve displacement are often asymptomatic and do not require specific treatment, Surgery is indicated when the patient is symptomatic or when arrhythmias or echocardiographic changesare detected (4).This clinical presentation highlights the rarity of such cases in our practice.

Introduction:

Ebstein's anomaly (EA) is a rare congenital heart disease, affecting approximately 1 in 200,000 live births and accounting for less than 1% of all congenital heart conditions(1). EA is a right ventricular cardiopathy caused by an embryological abnormality of the tricuspid valve, resulting in the over-apical displacement of the septal leaflet (2). Echocardiography is the key tool for the diagnosis and anatomic classification of this malformation.

Case Report:

We report the case of a 47-year-old woman patient with asymptomatic EA, she presented to the emergency department with permanent palpitations that had been presenting brutally for 12 hours at rest, associated with rest

Corresponding Author: M. Eddahbi
Address:Cardiology Department University Hospital Agadir, Faculty of Medicine & Pharmacy IbnZohr University, Agadir, Morocco.

dyspnea.Clinical examination on admission revealed a conscious patient, haemodynamically and respiratorily unstable: hypotensive at BP= 85/48 mmHg at both MS, tachycardia at 200bpm, cold extremities, polypnoeic at FR= 25cpm, SaO2= 89% on ambient air increased to 95% under 5l/min, Signs of Respiratory Distress+: intercostal/sternal indrawing, perioral cyanosis, presence of digital hippocratism, on cardiovascular examination, well-perceived rapid heart sounds associated with a murmur of tricuspid insufficiency without signs of heart failure. The pleuropulmonary examination and the rest of the somatic examination were without particularity. The electrocardiogram(ECG) demonstrated junctional tachycardia at a rate of 200 bpm, right axis, electrical HVD, appearance of BBD, retrograde P wave with RP:120ms **(Figure 1)**.Due to this instability, we administred a synchronous external electric shock at 150 joules after sedation, which successfully restored sinus rhythm and stabilized her hemodynamics. ECG after reduction showed a regular sinus rhythm with ventricular rate 75bpm, right axis, HAD, HVD, constant PR, fine QRS at 90 ms, wave T- DIII AVF V1V2V3V4 **(Figure 2)**.After the patient was stabilised, a chest X-ray showed cardiomegaly, cardiothoracic index of 0.7 with a supradiaphragmatic peak, hyperconvexity of the right lower arch, and the lung parenchyma was without abnormality **(Figure 3)**.

Subsequent echocardiographic assessment confirmed EA, revealing apical displacement of the tricuspid valve along with a mitro-tricuspid mismatch of 11.6 mm/m² **(Figure 4)**. Notably, the septal leaflet exhibited reduced mobility and was adherent to the interventricular septum, while the right atrium displayed dilation due to right ventricular atrialization **(Figure 5)**. Biological work-up showed haemoglobin at 19g/dl, normal renal and hepatic function, negative infectious work-up, thyroid and metabolic work-up with no abnormalities. The clinical evolution of our patient was favourable with clinical and electrical stability under medical treatment.Electrophysiological exploration was considered but the patient refused any further treatment.This case underscores the rarity of such clinical presentations in our practice.

Discussion:

EA is a congenital valvular and ventricular dysplasia of the right-sided heart, frequently associated with left-sided heart anomalies (1). EA results from incomplete delamination of the tricuspid valve (TV) from the right ventricular (RV) endocardium, which occurs between the 7th and 12th week of intrauterine life (2).

AE is a rare congenital heart disease that affects approximately 1 in 200,000 live births and represents <1% of all cases of congenital heart disease(2). It was first described in 1866 by W.Ebstein and remained unknown for a long time, with only 3 cases published in the literature between 1866 and 1950(2).

The clinical manifestations are very variable, theclinical presentation of AE depends on the severity of the tricuspid anomaly. Patients with minor displacementof the TV may remain asymptomatic until adulthood (3).Severe TV deformities lead to foetal death inutero or neonatal congestive heart failure with rapidly fatalrespiratory distress.In late adolescent and adult forms, the evolution is related to dilatation and right ventricular dysfunction and the development of rhythm disorders. This patient presented with junctional tachycardiawith haemodynamicinstability(3). These are some of the signsof advanced EA (8). The decompensating factorof this congenital heart disease, which had previously been asymptomatic, was probablythe onset of a rapid rhythm disorder (4).

The diagnostic approach should include an electrocardiogram, chest X-ray, transthoracic echocardiography and cardiac MRI in selected cases. Transthoracic echocardiography (TTE) is usually the first diagnostic tool(4). Second-line imaging includes transoesophageal echocardiography (TOE) and cardiovascular magnetic resonance (CMR), the former with particular usefulness in TV assessment, the latter in right-sided chamber volumes calculation and myocardial characterization(4). CMR is superior to TTE in the detection of PL and extracardiac abnormalities, while TTE reveals small septal communications more frequently(5).

Echocardiography and CMR are both recommended forevaluating unoperated patients, informing on progressive disease and anatomies suitable of repair, as well as for monitoring operated patients, revealing surgical results and possible complications(5). The quantitative criterion for EA diagnosis is an apical displacement of the SL hinge point by at least 8mm/m2 from the anterior mitral leaflet insertion, assessed in four-chamber view. An absolute distance in atrioventricular valves offsetting of 15mm in children and 20mm in adults is also considered diagnostic(6).Multimodality imaging is pivotal not only for anatomical and functional assessment of TV and right-sided chambers but also for identifying associated lesions.

The management of EA varies with the anatomical form and clinical presentation. Patients with minor displacement of the tricuspid valve are often asymptomatic and do not require specific treatment(7). In these patients, regular clinical and ultrasound monitoring is required. They should be monitored for arrhythmia, right chamber dilatation or right ventricular systolic dysfunction. Surgery is indicated when the patient becomes symptomatic or when arrhythmia or echocardiographic changes occur (8-9). It consists of repair or replacement of the tricuspid valve, with or without total or partial cavo-pulmonary anastomosis(10). In the event of a supraventricular rhythm disorder, long-term anticoagulation with or without an antiarrhythmic agent should be instituted. In the case of uncontrolled AF or AF associated with an accessory bundle, radiofrequency ablation or cryo-ablation may be proposed(10).

Conclusion:

Despite its rarity, the complexity and clinical implications of EA underscore the need for further research (9). Early diagnosis and appropriate management are crucial to improving patient outcomes and quality of life (10). We report the case of a patient with anEA at the age of 47 years which highlightsthe rarity of such cases in our practice.

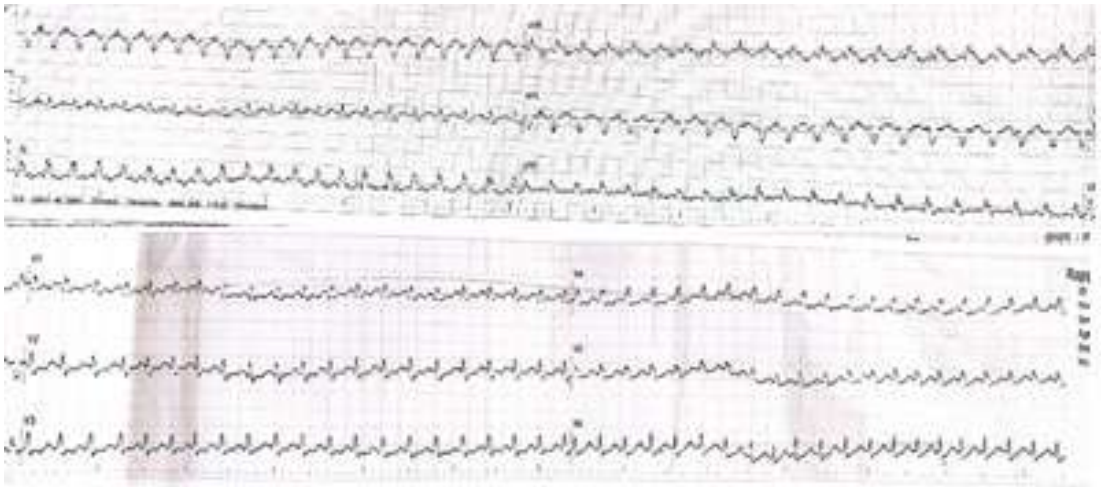

Figure 1:- ECG demonstrated junctional tachycardia at a rate of 200 bpm, right axis, electrical HVD, appearance of BBD, retrograde P wave with RP:120ms.

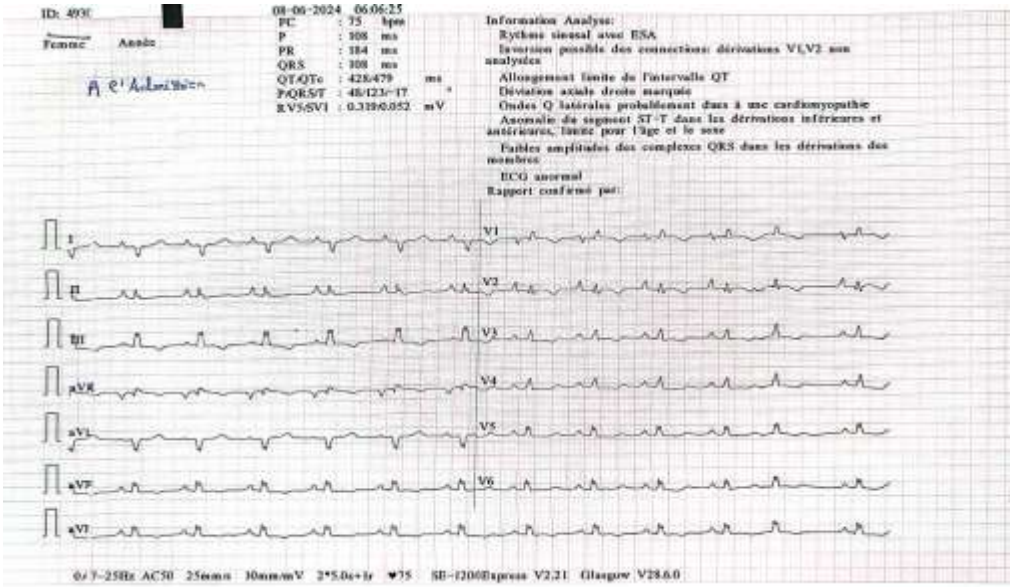

Figure 2:- ECG after reduction showed a regular sinus rhythm with ventricular rate 75bpm, right axis, HAD, HVD, constant PR, fine QRS at 90 ms, wave T- DIII AVF V1V2V3V4.

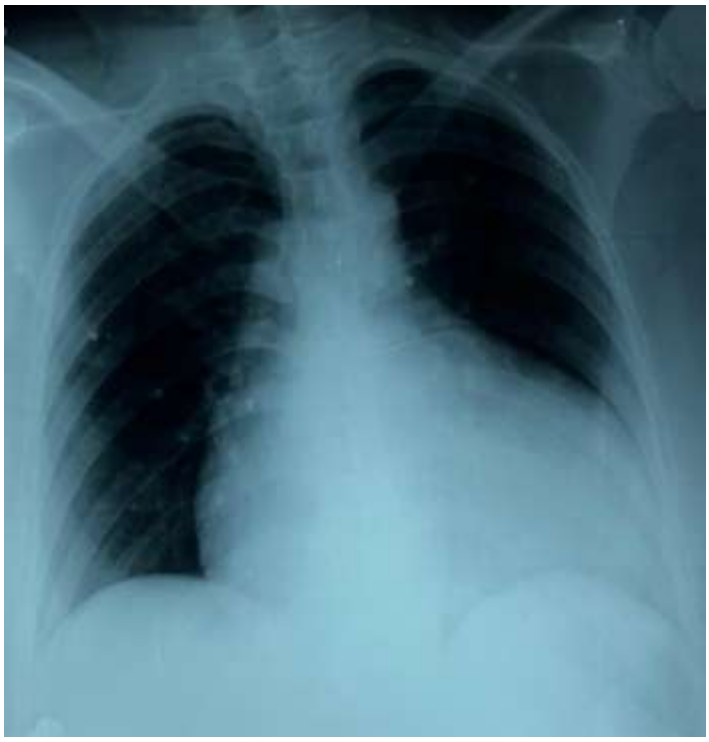

Figure 3:- Chest X-ray showed cardiomegaly, cardiothoracic index of 0.7 with a supradiaphragmatic peak, hyperconvexity of the right lower arch.

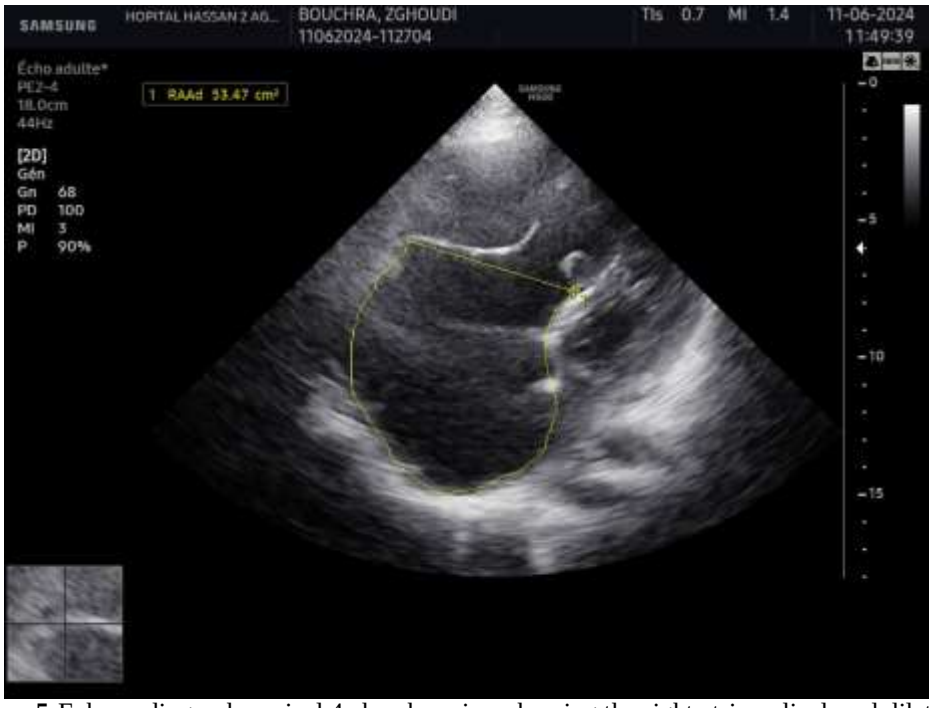

Figure 5: Echocardiography apical 4 chamber view showing the right atrium displayed dilation.

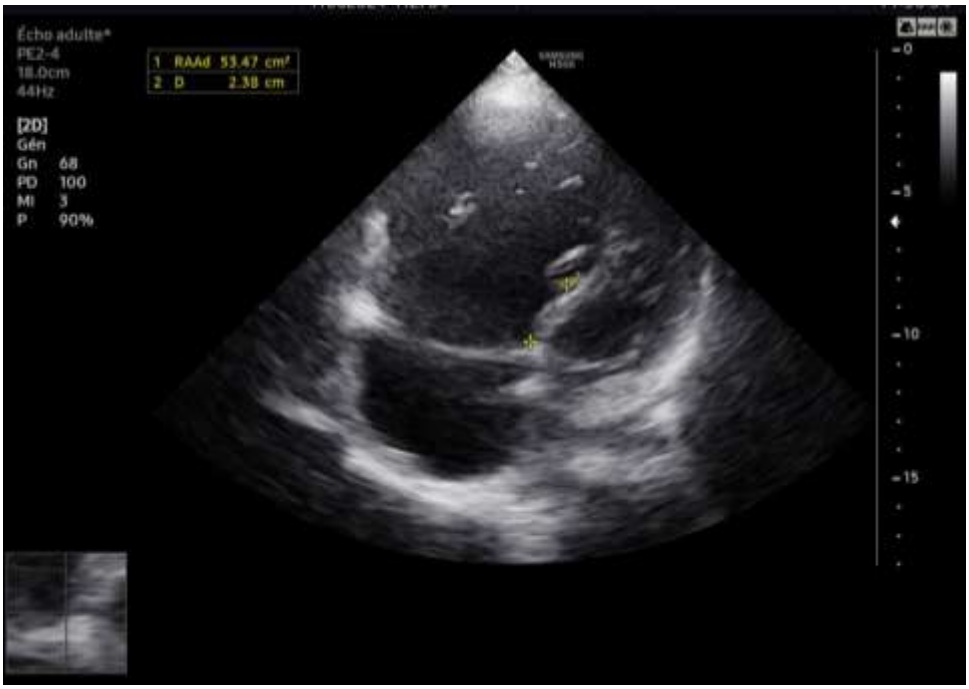

Figure 4: Echocardiography apical 4 chamber view showing apical displacement of the tricuspid valve along with a mitro-tricuspid mismatch of 11.6 mm/m².

References:-

1. Holst KA, Connolly HM, Dearani JA. Ebstein'sanomaly. *Methodist DebakeyCardiovasc J* 2019;
2. Yuan SM. Ebstein's anomaly: genetics, clinicalmanifestations, and management. *PediatrNeonatol*2017
3. Chauvaud S and Quarti A. Ebstein Anomaly inAdolescents and Young Adults. In :*TheTricuspid Valve in Congenital Heart Disease*,Eds: Giamberti A and Chessa M, Springer, Milan, 2014
4. Galea J, Ellul S, Schembri a, SchembriWismayer P, Callaja-Agius*J. Ebsteinanomaly :a review. Neonatal Netw*2014
5.Frescura C, Angelini A, Daliento L, Thiene G. Morphological aspects of Ebstein's anomaly in adults. ThoracCardiovasc Surg. 2000 Aug;48(4):203-8.
6. Adams JCL, Hudson R. A case of Ebstein'sanomaly surviving to age of 79. *Br Heart J*1956
7. Celermajer DS, Bull C, Till JA, Cullen S,Vassillikos VP, Sullivan ID *et al*. Ebstein'sanomaly: Presentation and outcome from fetus toadult. *J Am CollCardiol*1994
8. AttenhoferJost CH, Connolly HM, Dearani JA,Edwards WD, Danielson GK. Ebstein'sAnomaly. *Circulation* 2007
9. Chauvaud S, Carpentier A. Ebstein's anomaly: the Broussais approach. *Multimed ManCardiothoracSurg*2008
10. Malvindi PG, Viola N. Ebstein's anomaly:diagnosis and surgical treatment. *G ItalCardiol*(Rome) 2015.