ECRONICON OPEN ACCESS

EC CLINICAL AND MEDICAL CASE REPORTS

Case Report

Sudden Cardiac Death in a Teenager with Ebstein's Anomaly

Azza Makram Abdou*

Pediatric Cardiology Consultant, Egypt

***Corresponding Author:** Azza Makram Abdou, Pediatric Cardiology Consultant, Egypt.

Received: July 16, 2025; **Published:** August 12, 2025

Abstract

Ebstein's anomaly is a rare congenital heart disease with a wide range of clinical and morphological presentations in pediatric and adult age groups. This paper will present a teenager with mild form of Ebstein's anomaly presented with sudden death to highlight the cause of mortality in non-operated form of the anomaly [2].

Keywords: *Ebstein's Anomaly; Cardiac Death; Teenager*

Abbreviations

WPW: Wolf Parkinsonian White; SVT: Supra-Ventricular Tachycardia; CMRI: Cardiac Magnetic Resonance Image; GLS: Global Longitudinal Strain

Introduction

Ebstein's anomaly represents around 1% of the total congenital heart diseases. It occurs due to failure of proper delamination of the tricuspid valve leaflets resulting in apical displacement of the septal leaflet more than 8 mm/m^2 and sail like anterior tricuspid valve leaflet. Associated with congenital anomalies, like PFO, ASD, VSD, pulmonary valve anomalies, PDA, l -TGA, and left ventricular non compaction [4].

Clinically, presentation depends on associated cardiac malformations and degree of electrophysiological abnormalities. Cyanosis specially in the presence of right to left shunting lesion and in the presence of right ventricular obstructing lesion, circular shunt in neonate one of the early complications which need wise NICU management. Heart failure specially in adult non treated patient or in sever form in pediatric age group, arrythmias due to intra-atrial or atrio-ventricular accessory pathway although all type of arrythmias could be happen, pulmonary hypertension and paradoxical emboli resulting in myocardial infarction or cerebrovascular accident and unfortunately sudden cardiac death [1].

Assessment of patients needs multimodality in addition to good clinical sense with many scores. Echo cardiography, cardiac MRI are the main stay in assessment in addition to electrophysiological study; exercise tests; good clinical and laboratory assessment with traditional tools like ECG, CHEST X-ray, exercise stress test, CBC, ABG, cardiac biomarkers [2].

Early intervention consider a key factor for proper post-surgical outcome. waiting for symptoms linked to poor prognosis, fragmented QRS one of the emerging markers of worsening cardiac function, but not the sole indicator for early surgical decision, trans-thoracic

echocardiography taking part in decision but validation is needed, CMR consider the gold stander in diagnosis, it offers volumetric and non-volumetric parameter, cardiac function, myocardial fibrosis; degree of tricuspid valve angle rotation, in addition to evaluation of extracardiac structures. Exercise stress test and biomarker for heart failure are used as conjunct tools linked to other tools and also to estimate the clinical situation of the patient. Although there is no standardize algorithm to optimize the time of operative intervention [3].

Management of cases ranging from medical follow up with treatment of associated rhythm disturbance to wide range of staged palliative surgery and corrective surgery, cone repair considers the only corrective surgery of the anomaly with repair of its associated congenital heart defect if present. Early mortality in postoperative cone repair is considered null, although patients need lifelong follow up to avoid long term complications specially heart failure and arrythmias. Stem cell therapy in tricuspid valve repair, use of sodium co-transporter 2 inhibitor, pulmonary vasodilator, epigenetic modulator, all these emerging tools may change the paradigm in management of Ebstein's anomaly. Heart translation still a treatment option in end stage cases not suitable for operative treatment [3].

Case Study

16 years old boy known case of Ebstein's anomaly presented to ER arrested.

Since early infancy, mother notice facial cyanosis with intense cry for which mother sought medical advice and echocardiography done. The child diagnosed as a case of Ebstein's anomaly. The child followed up regularly. At the age of 11 years the child had syncopal attack during football game, for which EEG done and shows normal finding then ECG requested shows WPW pattern for which beta blocker prescribed and echocardiography. Previous diagnosis confirmed as a mild form of Ebstein's anomaly with WPW arrythmia for follow up and betablocker had been prescribed. After 2 years there is frequent syncopal attacks and the patient was enrolled in a big study about Ebstein's anomaly in Cairo university. Full investigations have been done which include basic and advanced investigations (CBC, ECG, HOLTER, conventional ECHO, SRAIN ECHO, cardiac MRI assessment) followed by electrophysiological study and ablation of the accessory pathway. Study suggests follow up of the case after ablation of the accessory pathway as there is reduction in diastolic function proven by 3D ECHO strain RV-GLS10%, LV GLS13%, in spite of normal cardiac function in conventional echo and CMRI study. When the patient stabilized with beta-blocker after ablation of the accessory pathway mother neglected follow up at higher center for 3 years. In the previous year patients had some limitations of physical activity specially with more than ordinary effort, patient sought advice again and another CMRI requested in addition to ECG, Echo-cardiography, 6-minute walk test. All investigation supporting a mild form of Ebstein's anomaly with severe to moderate tricuspid regurge, minimal pericardial effusion, mild degree of cyanosis in 6-walk test O_2 saturation drop from 97% to 89% in the end of the test, WPW pattern in ECG. His physician supports medical follow up as before and postponing surgical options to next year. 2-weeks later the child goes in picnic with his friends and returned tired at night and want to take a rest in bed, in early morning mother notice that the child is collapsed in bed, can't talk with pale face and cold extremities and with sever tachyarrhythmia. Mother transfers the child to the nearest ER. Patient resuscitated with DC shock but the patient does not respond.

Summary of the case						
At 2-months-old	Cyanosis with intense cry only	Normal growth and development	Initial echocardiography Ebstein's anomaly	Aneurysmal PFO	Saturation of oxygen 97%	Follow up of patient
At 11 years old	Syncopal attack	Normal growth and development	EEG normal	ECG WPW	ECHO as before	Beta Blocker Added

At 13 years	Frequent syncopal attacks, tired with more than ordinary activity	Normal growth and development	ECG WPW	Electrophysiological study	MRI shows mild form of Ebstein's anomaly with 1:1 QP/QS RVEDV: LVEDV 1.2:1 3D-ECHO RA EDVI 112 mm^2 RVGLS 10%, LVGLS 13% by 3D strain ECHO	Ablation of ectopic focus Then follow up to assess cardiac functions and dimensions
At 16 years old	Tired with more than ordinary activity	Normal growth and development	ECHO- sever tricuspid regurge, minimal pericardial effusion	ECG WPW Mild cyanosis with 6-minute walk test	MRI normal cardiac function and dimensions moderate to severe tricuspid regurge, mild pericardial effusion. Prominent hepatic veins, no cardiac muscle fibrosis	Surgical consultation postponed to next year. Add antibiotic and anti-inflammatory bills
2 weeks latter		Child collapsed in bed with sever tachyarrhythmia and not responding to resuscitation in ER				

Table 1

Cardiac functions and dimensions in the last screening of the case						
Ventricle	EF%	EDV	ESV	SV	EDVI	ESVI
CMRI						
Left	58	144	60	84	75	31
Right	66	172	58	114	90	30
2D-Echo						
Left	65	FS 34%	ESPAP 28 mmhg	Severe TR	LVIDD 46 mm	

Table 2

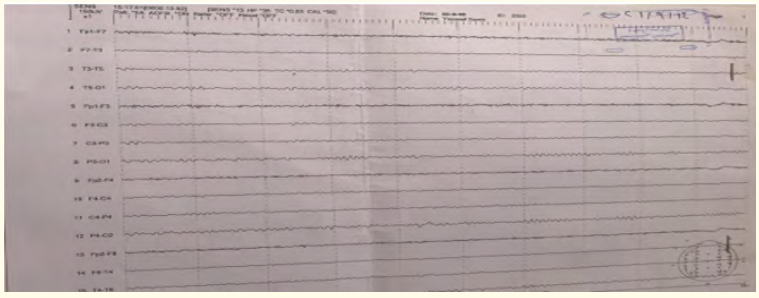

Figure 1: *EEG shows normal study.*

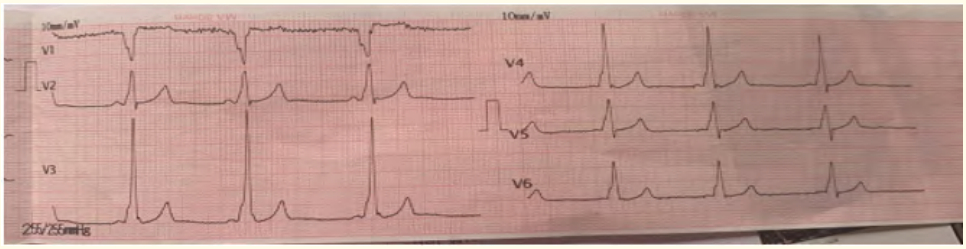

Figure 2: *ECG done 2 weeks prior to death shows wide QRS complex with delta wave during resting condition coincident with WPW syndrome.*

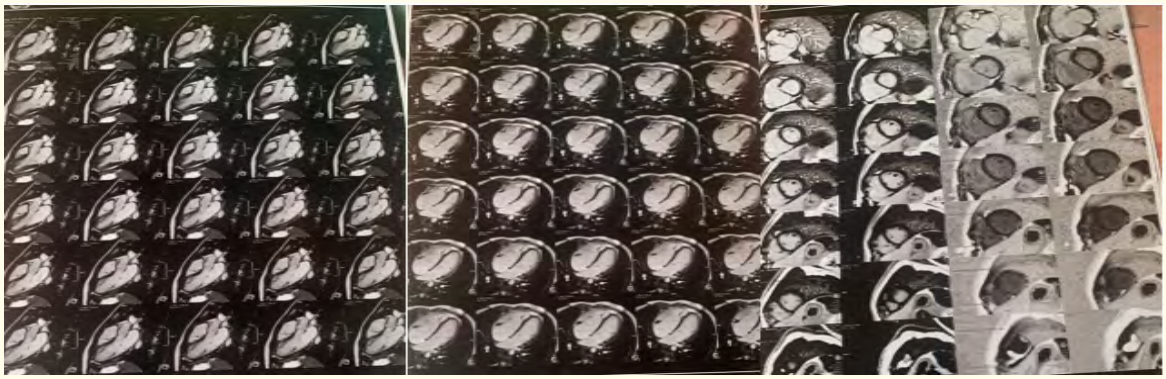

Figure 3-5: *Cardiac MRI show apical displacement of tricuspid valve hinge point with atralization of the right ventricle in different views and no evident cardiac muscle fibrosis in late gadolinium enhancement.*

Discussion

A national wide medical registry in Danish and Swedish had been done to study patient born from 1970 to 2017 and diagnosed as Ebstein's anomaly patient, this study includes 530 patient and 5.300 normal control person, each patient has 10 controls from same age and sex, this study cumulative mortality and hazard risk of mortality in those groups. Death due to cardiac cause is the most common among patient with Ebstein's anomaly, account for 86% in operated patients and 85% in non-operated patients, in corresponding age-related group was 14% in 2016 in Denmark. For more analysis the most common cause of death in operated patient is heart failure (20% of cardiac related death) and sudden cardiac death in non-operated cases (16% of cardiac related death). Early presentation magnifies the risk of cumulative mortality in both operated and non-operated cases [1].

In our case the patient diagnosed in infancy with simple form of the anomaly and associated PFO which closes partially with aneurysmal closure left 4 mm defect, deteriorated after syncopal attack due to accessory pathway, patient has background mild heart failure manifested by decrease global longitudinal strain in 3D-echo and this abnormality not assessed after ablation of the accessory pathway, also the patient in follow up had decrease activity in more than ordinary work, and when investigated the WPW pattern appear again in ECG after 3 years of ablation, in addition to minimal pericardial effusion and hepatic congestion which appear in cardiac MRI, sever tricuspid regurge which treated as infection in spite of normal lung and pleura in cardiac MRI study [1,3].

The patient was in true heart failure which aggravated with free activity during his picnic and ended by tachyarrhythmia as a subsequence of intraarterial or atrioventricular accessory pathway. Supraventricular arrhythmia is frequent in patient with Ebstein's anomaly. The impact of supraventricular arrhythmia itself or treatment with radiofrequency ablation is questionable and should be further investigated in this population. Patients often require more than one catheter ablation but eventually are likely to be free from arrhythmias. The imaging parameters assessed by echocardiography or cardiac magnetic resonance do not seem to be associated with ablation outcomes. It is believed that in Ebstein's patients SVA is a risk factor of sudden cardiac death, mostly because of the rapid conduction of electric impulses from the atria to the ventricle. However, the impact of SVA on overall survival, not just SCD, in this population needs to be better established [3].

Risk factor for poor outcome which include, HB > 15 mg/dl, hepato-renal impairment, increase N-terminal of atrial natriuretic peptide and it's relation to NYHA class, wide QRS complex corelated with decrease exercise tolerance also with frequency of syncopal attacks and tachyarrhythmias, associated cardiac defect, sever TR regurge, depressed myocardial function in echocardiography and CMRI in addition to fibrosis of the cardiac muscle. The interaction between all parameters and markers must be considered and keep in mind that decision making requires formal test [2].

Conclusion

In spite of 150 years passed since anomaly discovery many questions still not answered so national wide registry is needed for better understanding of this rare disease.

Bibliography

1. Connolly., *et al*. "Ebstein anomaly we should do better". *Journal of the American College of Cardiology* 81.25 (2023): 2431-2433.
2. Dib., *et al*. "Clinical and imaging predictors of poor prognosis in Ebstein anomaly". *CJC Pediatric and Congenital Heart Disease* (2025).
3. Di Cori., *et al*. "A twenty-year follow-up of adults with Ebstein anomaly with special focus on supraventricular arrhythmias, supraventricular arrhythmias and effectiveness of catheter ablation in 20-year follow-up of adults with Ebstein anomaly". *Journal of Clinical Medicine* 13.7 (2024): 2039.
4. Schultz., *et al*. "Heart failure in the adult Ebstein patient". *Heart Failure Reviews* 25.4 (2020): 623-632.

Volume 8 Issue 9 September 2025