Medical and Research Publications
International Open Access

Case Report **Journal of MAR Cardiology (Volume 3 Issue 5)**

Repair of Ebstein's anomaly in adult.

Ahmed, Basil[1], Alamri, Nora I[2], Alromaih, Norah Jamal[3], Hassan Mahmoud Hassan Mahmoud[4], Nashy, Medhat R[5]

1. Dhahran High School, Saudi Arabia.
2. Imam Abdulrahman University, Saudi Arabia.
3. Imam Abdulrahman University, Saudi Arabia.
4. King Fahd Hospital of University, Saudi Arabia.
5. King Fahd Hospital of University, Saudi Arabia.

Corresponding Author: Alamri Nora I, Imam Abdulrahman University, Saudi Arabia.

Received Date: October 25, 2021

Published date: November 01, 2021

DOI: 10.1027/marcy.2021.0160

Introduction:

Ebstein's anomaly (EA) is a rare form of congenital heart defects accounting for an incidence of one per 200,000 live births, this represents less than 1% of all congenital heart anomalies. (1) It was described in 1866, by Wilhelm Ebstein in a 19-year-old patient's autopsy complaining of palpitations and dyspnea.

We report a newly diagnosed 60-year-old patient with this condition, who presented with complaints of exertional dyspnea, swelling of legs and abdomen. Physical exam showed HR 100/min, engorged systemic veins, pedal edema, and massive ascites. Chest x-ray showed cardiomegaly and bilateral

pleural effusion. ECG of the patient revealed atrial flutter. Echocardiography revealed displacement of the septal and posterior leaflets of the tricuspid valve with severe tricuspid regurgitation, massive right atrial enlargement and small RV almost filled with calcium, with severely reduced function. Pt was operated upon with bicaval cannulation, and cardioplegic arrest. The RV was debulked of calcium taking care to not cause a VSD or inferior wall tear. The Tricuspid valve was repositioned and repaired with an annuloplasty ring. Post operatively hemodynamics improved with no TR. Pt was discharged home on the 4th post operative day. Pt was placed on diuretics, beta blockers and oral anticoagulation. 6 months post operative, he attends the clinic, with no ascites and much improved exercise tolerance.

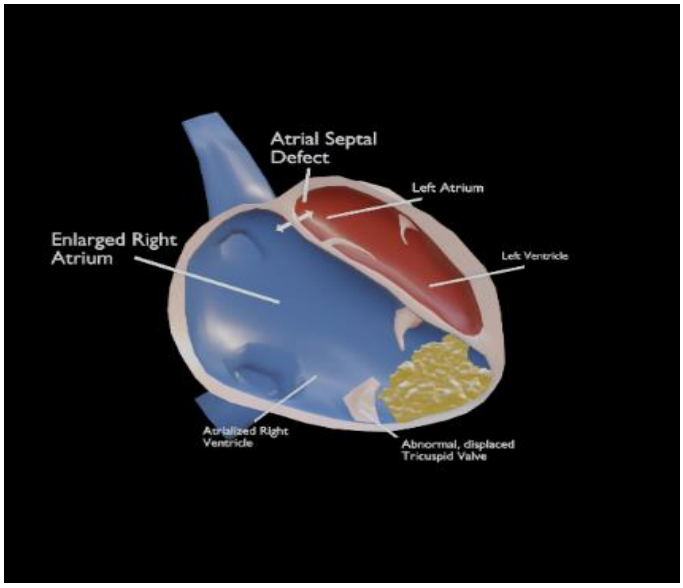

Figure 1: Depiction of anatomy in our patient

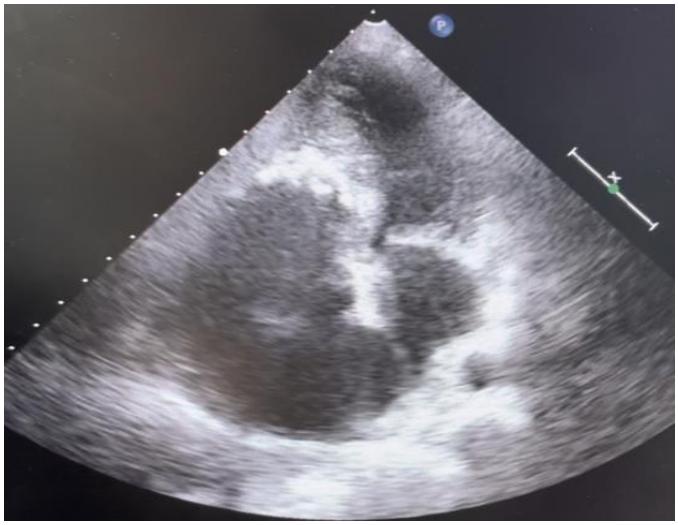

Figure 2: Pre operative Echocardiogram showing displaced valve and obliterated RV cavity.

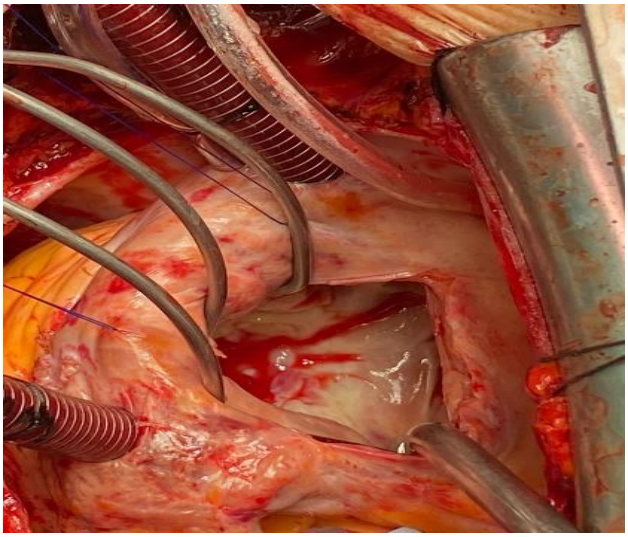

Figure 3: Intra op view of RV cavity filled with calcification

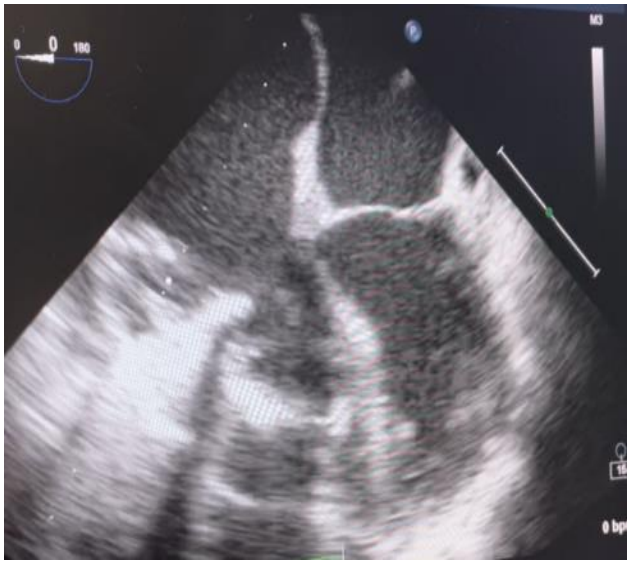

Figure 4: Post op echo demonstrating cleared RV and repositioned Tricuspid valve.

Discussion

Ebstein's anomaly is characterized by atrialization of the right ventricle with a distinguishing feature of apical displacement of the septal leaflet ≥ 8 mm/m2 body surface area. (2)

The Carpentier classification follows: Type A, minimal displacement of septal leaflet attachment with small atrialized RV; Type B, moderate displacement of septal leaflet attachment with large atrialized RV; Type C, important displacement of septal and posteroinferior leaflet attachment with non-atrialized or dyskinetic atrialized RV, restrained anterior leaflet motion, and short chords; and Type D, tricuspid sack.

Sudden cardiac death is one of the serious consequences from EA. (1, 3, 4) attributed to ventricular tachycardia, pulmonary stenosis, and presence of heart failure.

Conclusion

Our patient is unique, due to the buildup calcium in the RV cavity. No guidelines exist for management of adult patients with EA. Even though surgical repair in adult is debated, in view of our patient's severe symptoms and hemodynamic instability, we decided that debulking the RV of the calcium would be lifesaving. Our case offers scope for role of surgery for EA in adult.

Reference

1. Freeman, A. and Byard, R., 2017. "Ebstein Anomaly and Sudden Childhood Death". Journal of Forensic Sciences, 63(3), pp.969-971.

2. Moradi et al. "Complex Ebstein's Anomaly in an 86-Year-Old Iranian Man: A Case Report". J Tehran Heart Cent. 2017 Jan;12(1):39-41.

3. McGee, M. and Collins, N., 2015. "A case report on sudden cardiac death, Ebstein's anomaly and Left ventricular noncompaction. Heart, Lung and Circulation", 24, p.S399.

4. Attenhofer Jost, C., Tan, N., Hassan, A., Vargas, E., Hodge, D., Dearani, J., Connolly, H., Asirvatham, S. and McLeod, C., 2018. "Sudden death in patients with Ebstein anomaly". European Heart Journal, 39(21), pp.1970-1977a.