

Surgical Management for an Adult Female Patient With Ebstein Anomaly of the Tricuspid Valve With a Subvalvular Membrane With Severe PS and Multiple VSDs: A Case Report

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ABSTRACT

Ebstein's anomaly is a rare and complex heart defect that affects the tricuspid valve and is accountable for around 1% of congenital cardiac abnormalities. It is one of the most common congenital causes of tricuspid valve regurgitation. Ebstein's anomaly often is diagnosed prenatally, due to its severe cardiomegaly. Some individuals with this anomaly do not experience complications until adulthood and even then, they have mostly minor complaints like exercise intolerance. Atrial septal defect is most commonly (70-90%) associated with Ebstein's anomaly. However, ventricular septal defect (VSD) can be associated with 2-6% of the cases. This report presents a case of surgical intervention for a 20-year-old female with Ebstein's anomaly with severe pulmonary stenosis (PS) and multiple VSDs.

INTRODUCTION

Ebstein's anomaly is a disorder that primarily involves the tricuspid valve (TV). The anomaly is named after Wilhelm Ebstein, who in 1866 first described the defect's symptoms on the heart of 19-year-old Joseph Prescher [O'Leary 2010; Yuan 2017].

The EA produces several anatomical variations [Stulak 2007] and is characterized by:

- Embryonic failure of delamination of the septal inferior and anterior leaflets of the TV, resulting in adherence of the leaflets to the underlying right ventricular (RV) myocardium
- Apical displacement of the tricuspid leaflets' annular hinge points (septal > inferior > anterior) with an anteroapical shift in the functional TV orifice toward the RV outflow tract
- Dilatation of the atrialized portion of the RV along

with anterior leaflet fenestrations, redundancy or tethering and muscularization

- Dilatation of the anatomic TV annulus, thus typically results in severe regurgitation [Dearani 2015]

Genetic factors in Ebstein anomaly are heterogeneous. Most cases are sporadic; familial Ebstein is rare but may warrant genetic testing and family evaluation in this subset of patients [Dearani 2015]. Ebstein anomaly's most common symptoms are cyanosis, dyspnea, decreased



Figure 1. Chest x-ray showing cardiomegaly.

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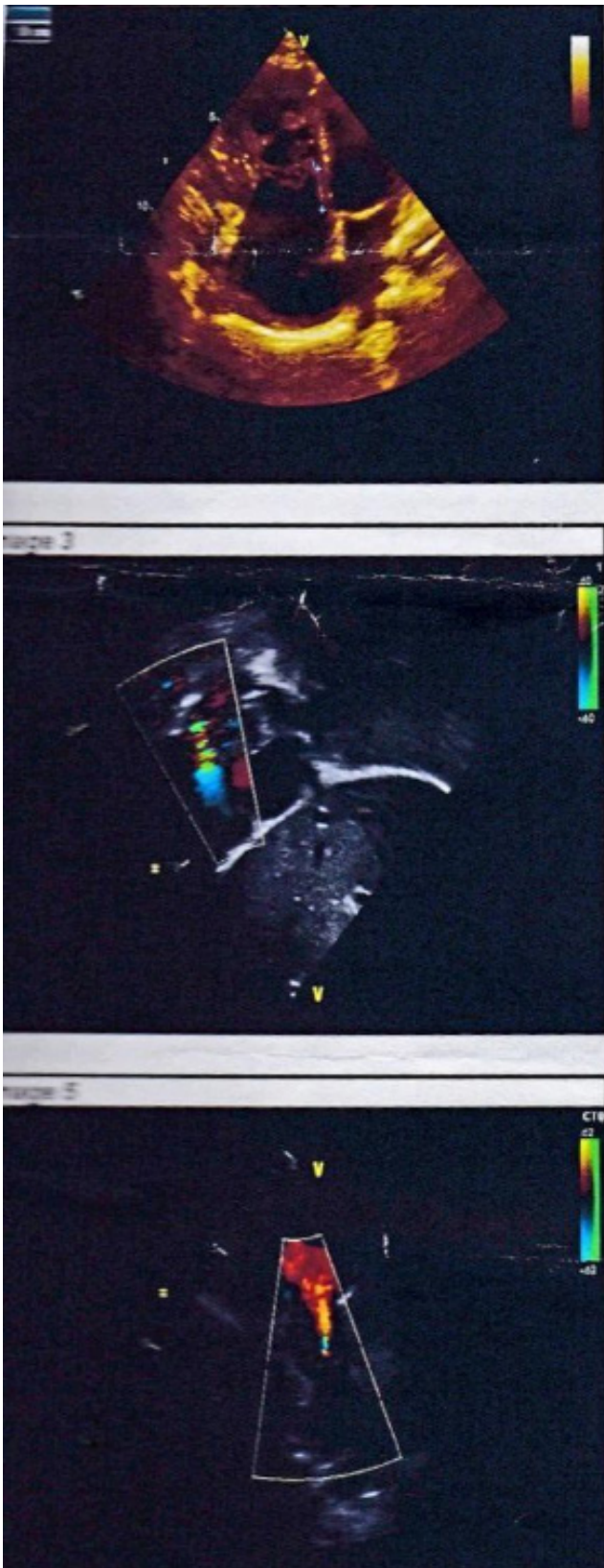


Figure 2. Transthoracic echocardiography showing atrialized RV.

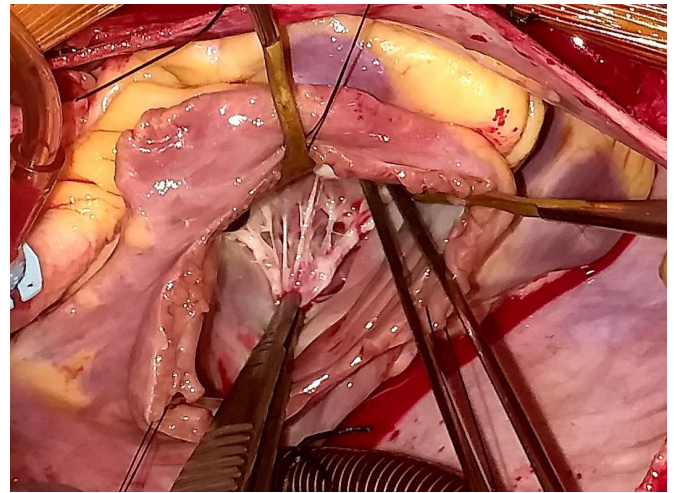


Figure 3. Redundant anterior leaflet of TV.

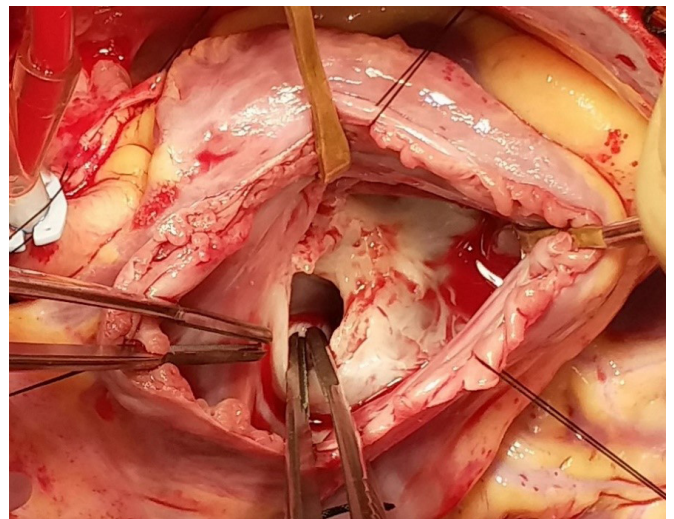


Figure 4. VSD through tricuspid valve orifice.

exercise tolerance, tachycardia, chest pain, and syncope. Arrhythmias are common as well and include accessory conduction pathways (Wolff-Parkinson-White syndrome) in about 15–20% of the defected ones and around 30–40% of those affected patients will develop atrial tachyarrhythmias by age 50 [Khositseth 2004; Stulak 2015]. Other commonly associated cardiac defects include atrial septal defect or patent foramen ovale (70–90%), ventricular septal defect (2–6%), and RV outflow tract obstruction that can occur as a secondary to structural abnormalities (pulmonary valve stenosis or pulmonary atresia), branch pulmonary artery stenosis or patent ductus arteriosus [Brown 2008; Da Silva 2019; Veen 2019]. In cases where pre-excitation is present (Wolff-Parkinson-White syndrome), preoperative electrophysiologic study and ablation usually are recommended at age of 4 to 5 years and typically followed by surgical repair of the anomaly. Hunter and Lillehei first reported surgical treatment of EA in 1958 [Hunter 1958]. Common indicators for

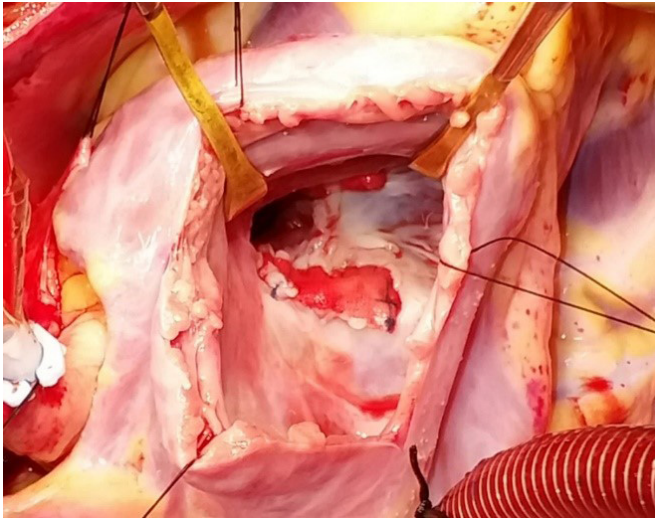


Figure 5. VSD closed with Dacron patch.

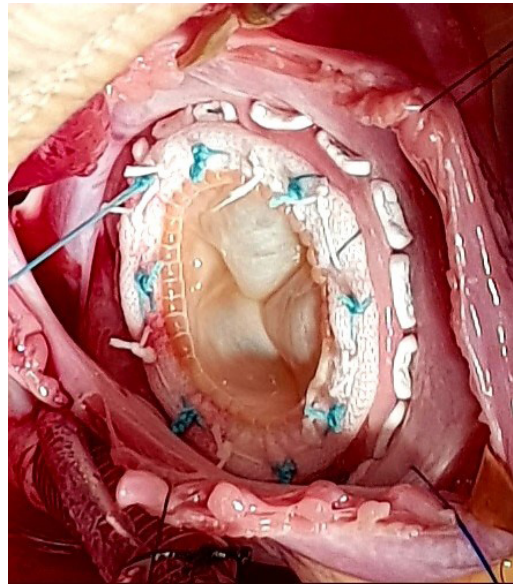


Figure 7. Tricuspid valve replaced with HANCOCK II tissue valve.

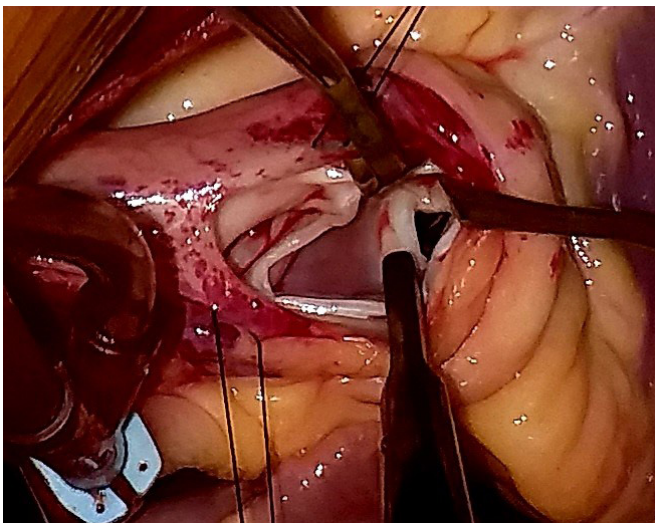


Figure 6. VSD through PAtomy.

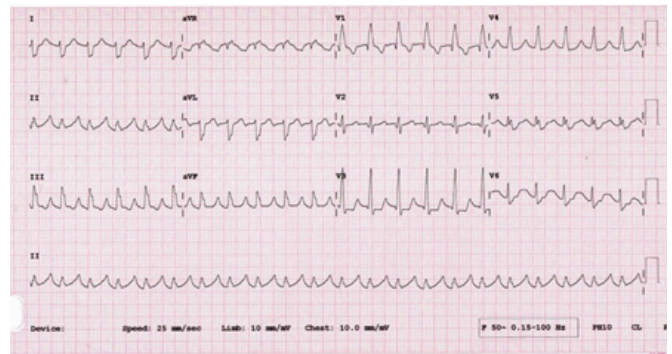


Figure 8. Postoperative ECG.

surgery include symptoms like fatigue, decreased exercise tolerance, cyanosis and shortness of breath or progressive RV enlargement, RV dysfunction, or the onset of atrial tachyarrhythmias [Dearani 2015]. Operative management routinely consists of TV repair or replacement, selective plication of the aRV, right reduction atrioplasty, closure (or subtotal closure) of any atrial septal communications, and correction of other associated anomalies [Brown 2008]. Here, we present a case for the surgical correction of Ebstein anomaly with multiple VSDs, including subvalvular PS.

CASE REPORT

A 20-year-old female was admitted into the medical ward with complaints of increasing dyspnea and

palpitations that had been present for the last six months. Her examination was unremarkable, with the exception of a harsh systolic murmur at the left parasternal and apical area. She had no other significant medical condition, and biochemical investigations were within a reasonable limit. The patient's BMI was 20.41 kg/m². ECG displayed RBBB with normal sinus rhythm, and only her chest X-ray showed mild cardiomegaly (Figure 1). However, transthoracic echo demonstrated intact IAS. Her IVS displayed the following: basal anterior membranous part of IVS formed a pouch, and there was perimembranous VSD (6mm x 5mm), while TV annulus measured 34 mm. The septal leaflet was attached 18mm more distally to the posterior IVS than the MV level and redundant anterior leaflet. Noncoaptation presented in between the anterior and posterior leaflets, producing mild to moderate TR. The RV appeared dilated and mildly impaired (TAPSE 14), whereas LV function was well preserved. The subvalvular membrane caused severe pulmonary stenosis, and the

third chamber measured 29 mm. The patient's color flow indicated a subvalvular membrane with severe subvalvular PS (PPG: 103mmHg) and bidirectional shunt flow across VSD, together with mild to moderate TR (PPG:80mmHg) (Figure 2). Cardiac cath showed generalized oxygen desaturation, and no PLSVC. However a VSD with Right to left shunt and severe PS (infundibular) found. Pressure gradient from RV to PA measured 80 mm Hg, yet favorable PA anatomy. The RV, on the other hand, showed coarsely trabeculated hypertrophy, while the LV graphic displayed VSD. Aortography results confirmed that the coronary arteries were in a normal position, and there was no MAPCA. The patient's Euroscore II for in-hospital mortality was 2.33%. This case was discussed in the heart team meeting, and we decided to perform an elective surgery for correcting the abnormalities presented.

Operative findings: The patient was prepared for standard open-heart surgery. Transesophageal echo confirmed tricuspid valve pathology, pulmonary stenosis and two VSDs: one in perimembranous and other in the muscular area. Cardiopulmonary bypass was established by standard aorto bicaval cannulation and myocardial protection with antegrade cold blood cardioplegia (del Nido) solution through aortic root. The temperature was reduced to 30°C. Opening of the right atrium native tricuspid valve revealed severe distortion (Figure 3). IAS was found intact, and on further inspection of VSD identified perimembranous and muscular position. Muscular VSD was closed by Dacron patch with interrupted prolene sutures (Figure 4). PA was opened as well and revealed a muscle band, plus a sub-pulmonic ring that was causing the RVOT obstruction. Both adequately were resected. Within the same incision, perimembranous VSD also was closed with Dacron patch (Figures 5 and 6). Atrialized RV was plicated. Initially cone repair was attempted and tested with saline, which did not show any satisfactory results. Therefore, the tricuspid valve was replaced with 27mm HANCOCK II PORCINE Heart Valve (Figure 7). RATomy wound was closed. The heart was weaned from CPB with small doses of Dobutamine and Noradrenaline. Postoperative TEE confirmed a normally functioning prosthetic valve with no residual VSD and normal RVOT flow. Total CPB time was 163 mins, and cross-clamp time was 137 mins. The patient was extubated the following day and her subsequent postoperative days were uneventful. Postoperative ECG did not show any significant changes (Figure 8). Transthoracic echo prior her discharge demonstrated a normally functioning prosthetic tricuspid valve, no paravalvular leakage, trivial TR (PASP: 20 mm of Hg), no VSD flow, no RVOTO, good LV (LVEF 58%) and fair RV systolic function, and no pericardial effusion. Our center previously experienced Ebstein's anomaly corrections only on children. This was the first time this procedure was performed on an adult. Furthermore, the case displayed uncommon associations, such as multiple VSD and pulmonary stenosis. Despite it, successful valve replacement took place.

DISCUSSION

Many surgical techniques have been reported so far. This primarily is due to the infinite anatomic variability with this anomaly [Dearani 2015]. However, high incidence of tricuspid dysfunction and tricuspid valve replacement (TVR) has been observed [Da Silva 2007; Danielson 1992; Carpentier 1988]. In 2004, the cone reconstruction of TV was proposed by Da Silva et al, shifting the paradigm of EA surgical management [Da Silva 2004]. TV replacement was reserved for valves not amenable to repair, older adults (>55–60 years of age), cases with massive RV or tricuspid annular dilatation [Brown 2009]. Bio-prosthetic (porcine) valve replacement generally is preferred due to its relatively excellent durability of the porcine bio-prosthesis in the tricuspid position, and therefore long-term warfarin anticoagulation therapy can be avoided [Brown 2009]. When the bio-prosthetic valve deteriorates, percutaneous “valve-in-valve” insertion is an alternative option [Hasan 2011; Roberts 2011]. Mechanical valves on this procedure are associated with a higher frequency of valve malfunction and thrombotic complications compared with mechanical valves in other cardiac positions, mainly when RV function is reduced [Brown 2009; Said 2014]. During a valve replacement in Ebstein's anomaly, the suture line deviates to the atrial side of the atrioventricular node and membranous septum. This is done to avoid the conduction tissue and right coronary artery injury. The prosthesis, as a result, is in an “intra-atrial” position. The coronary sinus is left to drain into either the RA or right ventricle, depending on its proximity to the conduction tissue. In contrast to tricuspid repair, a tricuspid replacement can be performed without a cross-clamp once intracardiac communications have been closed. This is particularly important when RV function is reduced or left ventricular function is below normal [Dearani 2015]. Preoperative functional capacity of the patient (NYHA class) is specifically reported as the most important parameter of survival [Attie 2000].

Follow up: The patient was reviewed after two years. She was found entirely asymptomatic (NYHA I), fully mobile, active, and completely independent. The heart team intends to follow up with her once a year.

CONCLUSION

Ebstein anomaly is a complicated form of congenital heart disease with variable clinical presentations. In this condition, the visualization of the apically displaced septal tricuspid leaflet is an excellent diagnostic feature. Appropriate preoperative assessment of the patient, both clinical and by cardiac echo, is the prerequisite. With a sound knowledge of the cardiac anatomy, accurate scheming of surgical outcomes, routine follow ups, multidisciplinary team approach and better management, an experienced center can ultimately improve the prognosis of such patients. However, there are few factors to consider for favorable outcome, such as ventricular function, presence of polycythemia, a patient's sex, and right ventricular outflow tract obstruction [Brown 2008].

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