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CASE REPORTS

Surgical Management of Ebstein's Anomaly with Atrial Septal Defect in a Young Lady: A Case Report

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Abstract:

Ebstein's anomaly is a rare congenital heart defect accounting for <1% of all cases. It is commonly associated with other cardiac malformations particularly, 50% of the patients are associated with atrial septal defect. We report a 22-year-old lady diagnosed to have Ebstein's anomaly with small atrial septal defect. She was surgically managed in Bangabandhu Sheikh Mujib Medical University by replacing the defective tricuspid valve withSt Jude Medical Epic porcine bio-prosthetic heart valve along with plication of atrialized portion of right ventricle and direct closure of the atrial septal defect.This resulted in excellent symptomatic improvement.

Introduction:

Ebstein's anomaly (EA) of Tricuspid Valve (TV) is a rare congenital heart defect (CHD) described in 1866 by a German physician Wilhem Ebstein (1836-1912). The incidence is 1-5 in 2,00,000 live births, accounting for <1% of all cases of CHD. At autopsy of the original case, Ebstein described, an enlarged and fenestrated anterior tricuspid leaflet; hypoplastic posterior and septal leaflets, thickened and adherent to the right ventricle (RV), thin and dilated atrialized portion of RV, enlarged right atrium (RA) and a patent foramen ovale (PFO).[1] However, the anatomical hallmark of this malformation is, a downward displacement of the hinge point of the septal and posterior tricuspid leaflets below the annulus due to delamination of these tricuspid valve leaflets from the underlying myocardium during embryogenesis.[2] More than 30% of patients with EA have association with other cardiac defects e.g. Pulmonary atresia or stenosis, VSD, PFO or ASD, TOF, PDA and congenitally corrected TGA. ASD is present in 50% of patients with EA.[3] Patients may present at any age and the disease has a highly variable clinical course.[4] Three cardinal pathologic features predominate in patients with EA. These are RV abnormality, TV abnormality and accessory conduction pathways (WPW syndrome).[5] Older children and adults usually present with fatigability, reduced exercise tolerance, effort dyspnea and cyanosis. Medical management with diuretics and anti-arrhythmic drugs may be used for symptomatic management of heart failure and arrhythmia, however most patient requires surgery. Immobility and morphology of the TV prevents repair in about 20-30% of the patients with EA and therefore warrant valve replacement.[6]

Case Report:

A 22-year-old young mother presented with history of palpitation, exertional dyspnea and undue fatigue for 3 years. These symptoms went worse after the delivery of her only baby 1.5 years earlier. She is normotensive, non-diabetic and non-smoker. Her physical examination revealed a regular pulse rate of 106 beats/minute, blood pressure 110/70 mm of Hg and normal auscultatory finding of the chest and heart except a systolic murmur of grade 4/6 along the left lower parasternal area. ECG shows sinus tachycardia with heart rate 106 beats/minute. Chest X-ray revealed, cardiomegaly with cardiothoracic ratio of 0.65 and normal pulmonary vascular markings. Color Doppler echocardiogram showed, septal leaflet is short (12mm) and displaced about 20mm from AV junction and loop like shape and tethered to IVS; basal insertion of posterior leaflet (28mm) is extremely displaced about 52mm from AV junction and apical end is tethered to the opposite IVS; anterior leaflet is long (81mm), sail like, basal insertion is within normal limit but the apical part is tethered to the RV free wall, moderate tricuspid regurgitation, grossly dilated right atrium due to atrialization of RV (size: 79mm x 36mm), smaller right ventricle (effective volume = 71ml) and a small ASD (8mm) with left to right shunt. The patient was therefore diagnosed as EA of TV (Carpentier Type-C) with ASD and was planned to undergo surgery.

With all aseptic precautions, under GA with ET intubation, a standard median sternotomy was done. Thymus was

dissected out and Pericardiotomy was done. Cardiopulmonary Bypass (CPB) was established with bicaval and aortic cannulation. Aortic cross clump was applied. Cardiac arrest was achieved by antegrade cardioplegia under mild hypothermia (32^{o} C). Right atriotomy was done and TV was approached through RA. TV annulus was found grossly dilated, leaflets were thinned out, septal and posterior leaflets were rudimentary and anterior leaflet was sail like. The defective valve was unfavorable for repair as the morphology was absolutely distorted (Fig-1). TV was replaced with St Jude Medical Epic porcine bio-prosthetic heart valve of 31mm size with complete preservation of subvalvular structures (Fig -2). A small ASD was noted and repaired by direct closure. RA was closed and aortic cross clamp was released. Patient weaned from CPB uneventfully. After adequate haemostasis, chest was closed in layers keeping 2 drains (24Fr) in retrosternal and right pleural space attached with water seal drainage bag and 1 epicardial pacing wire in the RV wall. 4 units of whole blood was transfused peroperatively. Total cross clump time was 33 minutes and total bypass time was 70 minutes. Per-operative period was uneventful. Patient was extubated 6 hours after the surgery. Postoperatively patient recovered well with minimal inotrope support. Drain tubes were removed on 4th POD and Patient was discharged on 8th POD in a satisfactory condition with advice to take oral warfarin 5mg daily for 3months. On subsequent follow-up, patient stated, she is enjoying almost a symptom-free life.

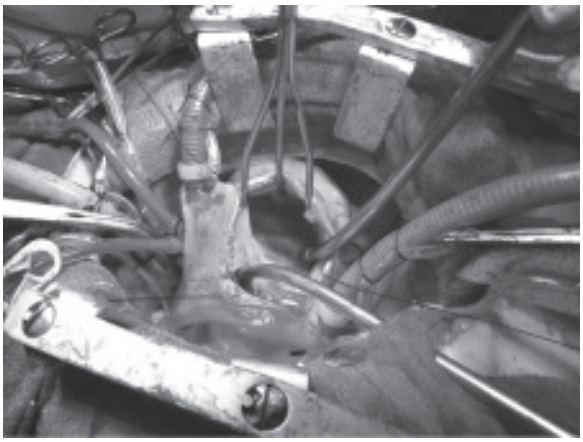

Fig.-1: *View of tricuspid valve through right atrium. Note that, posterior and septal leaflets were rudimentary. A cardiotomy sucker was passed through the atrial septal defect into left atrium.*

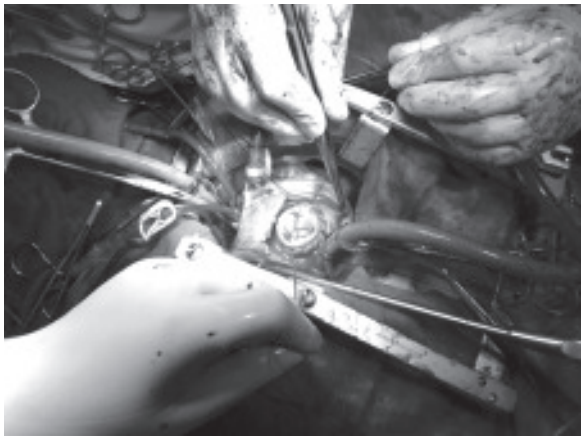

Fig.-2: *Tricuspid valve is replaced by St Jude Medical Epic porcine bio-prosthetic heart valve of 31mm size.*

Discussion:

The structural abnormality in EA is the TV leaflets are displaced apically from the TV annulus and therefore, the actual orifice is located inside the RV cavity at the junction of inlet and trabecular portions of RV leaving a large ventricular cavity merged with the right atrial cavity (atrialized right ventricle). Only the outlet and trabecular portion remain as effective ventricle (functional right ventricle). Interatrial communications are present in 80-95% of patients with EA[7]. One third present with an ASD and majority of the reminder as PFO.[8] Our patient also presented with similar findings.

EA is a rare disease but due to recent advancement of diagnostic facilities, cases are diagnosed and reported at a higher incidence. Cardinal symptoms of EA are cyanosis, arrhythmia and right heart failure.[1] Others manifestations are Paradoxical embolism and Endocarditis. Many patients first experience symptoms as adults, but onset can occur at birth or in infancy or in childhood.[3] However, patient may be asymptomatic and could incidentally be diagnosed. In this case, patient presented with palpitation, exertional dyspnea and undue fatigue in her adulthood. Multiplicity of heart sound and murmurs from the right heart with a wide and persistently split second heart sounds are typical. Digital clubbing depends on the degree of cyanosis. [9] Our case presented with normal heart sounds and systolic murmur of tricuspid regurgitation but cyanosis and clubbing were absent. Chest X-ray vary from normal to typical globe shaped heart with narrow pedicle, cardiomegaly, normal or low pulmonary vascularity[7] coincides what we have also found in this case. 2D echocardiography is considered as gold standard[7] and can delineate the TV defect and concomitant other cardiac malformations. An interatrial or interventricular defect usually found to have a right to left shunt and therefore,

present as central cyanosis. Similarly, on echocardiography our patient is also found to have EA and a small ASD but with left to right shunt, therefore, patient did not present with cyanosis. Most patients with EA present with abnormal ECG. Abnormalities include sinus tachyarrhythmia, tall and broad P-wave with complete or incomplete RBBB, first degree AV block, WPW syndrome (25% cases), atrial flutter or fibrillation (25% cases). [1,3] However, our case presented with sinus tachycardia on ECG.

With the ongoing medical and surgical advancement, several treatment options are described by several authors. Surgical modalities depend upon the severity and type of defect in EA. No EA has similar presentation and there are varying ranges of defect in TV and also other concomitant cardiac malformations. Therefore, each case should be individualized. Indications for surgery in EA is not clearly defined and the ideal mode of surgical management remains controversial.[10,11] A study on 8 patients with EA who underwent TV replacement showed good early clinical improvement and excellent long-term result.[12]Another study showed excellent durability in 158 patients who received a primary tricuspid bio-prosthesis due to unfavorable tricuspid morphology for repair. [13] A comparative study between mechanical and bio-prosthetic heart valve on 333 patients who required a valve replacement, showed superior late survival with those who received porcine bio-prosthesis. [14]

In our setting, we performed replacement of the defective TV with St Jude Medical Epic porcine bio-prosthetic heart valve with plication of atrialized portion of right ventricle and direct closure of the ASD.

Conclusion:

Ebstein's anomaly is rare but not unusual. With recent advancement, cardiac surgeons are now-a-days dealing such cases by replacing the defective valve with bio-prosthetic valve which give good postoperative outcome, reducing the morbidity as well as the mortality by ensuring a good symptom free life after surgery.

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