Ebstein's Anomaly Corrected by Tissue Valve: Case Report

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Key words:
Ebstein's anomaly, WPW (Wolff-Parkinson-White) Syndrome, Celermajer's index, Tissue valve.

Abstract:

Ebstein's anomaly is a rare form of congenital heart disease with incidence of 1% of all congenital heart diseases. There are two modes of surgical correction of Ebstein's anomaly: either biventricular repair with or without tricuspid valve replacement, or palliative univentricular repair consisting of bidirectional Glenn shunt or Fontan procedure. We treated a case of severe form of Ebstein's anomaly with ASD secundum with WPW syndrome. Radiofrequency ablation was done to treat WPW syndrome preoperatively. Celermajer's index is a prognostic indicator for tricuspid valve repair or replacement. On 04.03.08 tricuspid valve was replaced with 31 mm Carpentier-Edwards bovine pericardial valve under cardiopulmonary bypass. Postoperative period was uneventful. Follow up echo done on 01.06.08 which revealed normally functioning tissue valve found in tricuspid position TR Grade I. So, in conclusion, preoperative evaluation and workout of Celermajer's index is essential before surgical intervention for decision of tricuspid valve repair or replacement. Last but not the least, any event of arrhythmia should be properly evaluated.

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Introduction:

Ebstein's anomaly is a rare form of congenital heart disease with incidence of 1% of all congenital heart diseases. Age of presentation depends upon the degree of morphological and functional variation of the disease itself which may be of mild to severe form. Ebstein's anomaly consists of downward displacement of septal leaflet of tricuspid valve from tricuspid valve ring towards the apex.[1-3] There are two modes of surgical correction of Ebstein's anomaly: either biventricular repair with or without tricuspid valve replacement, or palliative univentricular repair consisting of bidirectional Glenn shunt or Fontan procedure. We treated a case of severe form of Ebstein's anomaly with ASD secundum with WPW syndrome.

Case history:

Khurshida Akter, a 17-year-old girl from Chittagong, got admitted on 29.01.08 in the Cardiac Surgery Department of National Institute of Cardiovascular Diseases (NICVD) hospital, with complaints of palpitation, exertional dyspnoea with growth retardation. On examination, she was ill-looking with below average body built, mildly pale, RR 18/m, HR- irregular, having thrill and a systolic flow murmur in pulmonary area and a pansystolic murmur in tricuspid area. She was clinically diagnosed as a case of ASD secundum with tricuspid regurgitation. On investigation, CXR- mild cardiomegaly, ECG- WPW (Wolff-Parkinson-White) syndrome, Echo Doppler (Fig 1)- Ebstein's anomaly, ASD secundum with L>R shunt, severe TR. On echo evaluation, cardiologist suggested that

$$\text{Celermajer's index} = \frac{\text{RA Area + Area of atialized RV}}{\text{LA area + LV area + functional RV area}} = 0.75$$

This indicates that there is 10% chance of mortality if a biventricular repair is attempted. So, it was decided to correct this anomaly with a tissue valve implantation in tricuspid annulus and closure of ASD.[4] Radiofrequency ablation for WPW syndrome was done on 18.02.08.

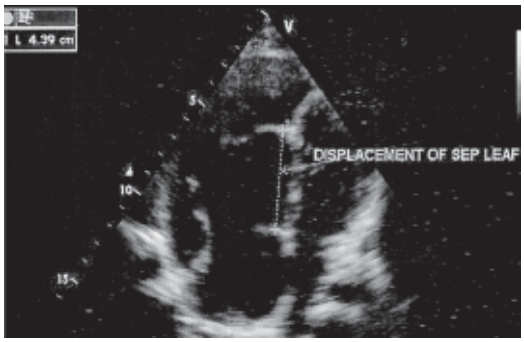

Fig.-1 *Ebstein's anomaly of tricuspid Valve (apical 4-chamber view).*

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Operative procedure:

Operation was performed on 04.03.08 under G/A with all invasive and noninvasive lines. Median sternotomy done. Cardiopulmonary bypass was established with bicaval cannulation and aortic cannulation. After snaring SVC and IVC cannulae, aorta was crossclamped. Antegrade cold blood cardioplegia was administered, heart arrested in diastole. RAtomy was done. Interior of RA, RV, and through ASD, LA, LV were examined. Four pulmonary veins drained to LA. Mitral valve was competent and of adequate orifice. LV was normal. Atrialized part of RV was examined.There was mild to moderate RVOT obstruction. RVOT was 9 mm.

Anterior leaflet of tricuspid valve was excised; 31 mm Carpentier-Edwards bovine pericardial valve was implanted in tricuspid position with pledgetted valve suture with technique of moderate plication of atrialized RV. ASD secundum was closed by a right atrial patch and RVOT enlarged with pericardial patch after resecting hypertrophied bands. Right atrium was closed. Cross-clamp time was 91', extracorporeal circulation time 137'. Patient weaned from cardiopulmonary bypass; chest was closed in layers after ensuring haemostasis keeping two chest drains tube in situ.

Postoperative Period:

Patient was transferred to ICU. ICU recovery was uneventful. Before discharge, Echo evaluation was done. Effective RV was 36X39 mm. RVOT 17 mm; good functioning tricuspid valve with mild central regurgitation. RVEF 49%, LVEF 62%. No flow across ASD closure patch was found.

Patient was discharged on 13.03.08 in good condition with anticoagulant treatment i.e. tab aspirin 75 mg and tab warfarin 2.5 mg daily with advice to come on follow up visit after 1 month. On follow up visit, patient was found clinically and echocardiographically improved.

On follow up echo done on 01.06.08, normally functioning tissue valve was found in tricuspid position and TR Grade I (Fig 2).

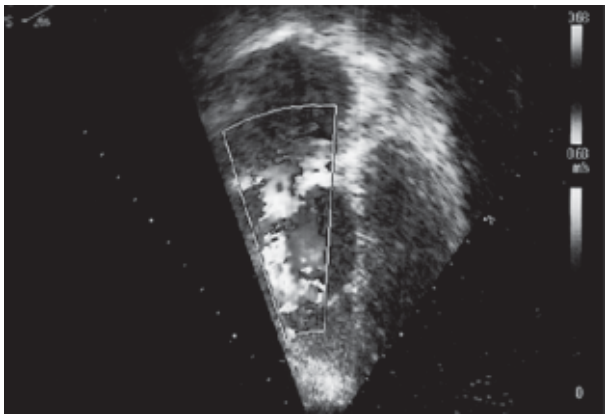

Fig.-2: *Follow up Echo done on 01.06.08*

Discussion:

Ebstein's anomaly is defined as a congenital heart disease in which the septal leaflet of tricuspid valve or posterior leaflet or both are displaced downward into the right ventricle, the anterior leaflet is long and sail-like with atrialization of the right ventricle causing tricuspid regurgitation. Atrial septal defect secundum type is a common association, about 60%. In about 14% cases Wolff-Parkinson-White (WPW) syndrome occurs. Ebstein described this anomaly in 1866[5]. Sometimes it is associated with other complex congenital heart diseases eg. Pulmonary atresia or stenosis, or RVOT obstruction and congenitally corrected transposition of great arteries. The Ebstein's disease was first described by Arndein in 1927.[6]

This condition may remain asymptomatic in childhood. Sometimes, the disease may come to light due to associated anomaly or during a routine medical check up.

Due to tricuspid regurgitation and presence of atrial septal defect patient develops cyanosis and heart failure and later on clubbing.[7] Some patient may develop breathlessness, cyanosis, cardiomegaly and heart failure in first week of life.Though may remain asymptomatic, their exercise tolerance is less than normal children. Oxygen saturation at rest is a major predictor of exercise intolerance.[8] When patient becomes symptomatic, breathlessness on exertion, low oxygen saturation, cyanosis, clubbing and signs of heart failure may be found. Palpitation and vague chest pain may be the leading symptoms in patients who have WPW syndrome. In cyanotic patient there is polycythemia. The patient sometimes develops malar flash.

On examination, the patient may present with breathlessness, dependent edema, engorged neck veins and hepatomegaly. Auscultation of heart reveals wide splitting of first sound with accentuation of the delayed component caused by closing of the large anterior tricuspid leaflet.[9-10] Pansystolic murmur may be evident at the lower end of left sternal margin due to tricuspid regurgitation.

Chest X-Ray shows cardiomegaly with right ventricular preponderance. ECG may present incomplete right bundle branch block with features of right ventricular hypertrophy or WPW syndrome, if it is present.

Echocardiography is diagnostic; 2D, M mode and Doppler study can give the detail information regarding the anomaly.[11-12] Work out of Celermajer's index is essential to decide for tricuspid valve repair or replacement.

Cardiac catheterization may be required in adolescent and adult patients to determine the presence of associated anomalies and evaluation of pulmonary vasculature.

Surgical intervention is aimed at repair of tricuspid valve and closure of atrial septal defect.[13]

In this particular case, electrophysiological study and ablation of Kent's bundle required due to presence of WPW syndrome. The accessory conduction pathway could be divided during surgery.[14] Due to Celermajer's index 0.75 tricuspid valve replacement than repair was considered safe in this case, which has a risk of >10% death if a valve repair is attempted. Valve repair often requires an annuloplasty ring. the real danger in valve repair and implantation of valve in tricuspid position is danger to conduction system of the heart.[15] In this particular case, use of a bovine pericardial tissue valve Edward life science perimount bovine pericardial valve is expected to serve for 20-30 years in this low pressure zone.[16] Atrialized portion of right ventricle is plicated during valve replacement.

The closure of ASD is usually done by a pericardial patch. In this case, the hugely dilated right atrial wall is used to close the large ASD secundum. The pericardium is used to enlarge the RVOT after resecting hypertrophied bands.

Conclusion:

Ebstein's anomaly is a congenital heart disease. Preoperative evaluation and workout of Celermajer's index is essential before surgical intervention for decision of tricuspid valve repair or replacement. Last but not the least, any event of arrhythmia should be properly evaluated.

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