Case Report

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One and a Half Ventricle Repair of a Rare Ebstenoid Tricuspid Valve Orifice with a Hypoplastic Right Ventricle

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Abstract

Primary tricuspid valve disease related to its congenital malformation is rare. Ebstein's anamoly is a rare congenital condition affecting the septal and posterior leaflets of the tricuspid valve. Ebstein's anamoly has a wide spectrum of pathological features. We describe the case of 33 year old female with Ebsteinoid tricuspid valve orifice, severely hypoplastic right ventricle and thrombus in a giant right atrium, with intact interatrial and interventricular septa. The patient was managed surgically with Kays tricuspid annuloplasty and right Bidirectional Glenn anastomosis constituting one and a half ventricular repair with satisfactory outcome.

Keywords: Ebstenoid Tricuspid valve; Hypoplastic Right ventricle; Right atrial clot; One and a half ventricle repair; Kays annuloplasty.

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INTRODUCTION

Ebstein's anamoly is a rare complex congenital anamoly accounting for nearly 1 percent of all congenital heart defects.[1] The Ebstein's anamoly classically has downward displacement of the hinge point of septal and posterior leaflet of tricuspid valve in association with dilatation of annulus, atrialisation of right ventricle and also the anamolies of the anterior leaflet. Dysplasia of the leaflets is also seen in Ebstein's anamoly. There are several morphological variants of Ebstein's anomaly of tricuspid valve. It also needs to be differentiated from rare varieties like congenital unguarded tricuspid valve orifice which usually has patent right ventricular outflow tract and dilated right ventricle. The clinical presentation is also heterogenous based on the anatomical variants.[3] The surgical strategies of management of this anamoly include valve repair, valve replacement, one and a half ventricle repair, single ventricle repair and transplantation. Patient presenting early as neonates with severe right ventricular and tricuspid valve dysfunction need aggressive surgical approach. The late presenting patients with milder form the disease can be managed with valve repair. However in someone and a half ventricular repair has been preferred when the right ventricle is severely hypoplastic and fails to support the pulmonary circulation.[4] We report a case of Ebsteinoid tricuspid valve with a hypoplastic right ventricle and right atrial thrombus who underwent Kays tricuspid annuloplasty and right Bidirectional Glenn anastomosis constituting one and a half ventricular repair with satisfactory outcome.

CASE REPORT

A 33-year-old female patient, a known case of Rheumatoid arthritis, hypothyroidism presented with complaints of dyspnoea on exertion NYHA class III and frequent palpitations since 1 month. She had no complaints of facial swelling, pedal edema, jaundice or abdominal distension. On examination, she had pulse rate of 120 perminute and blood pressure of 100/70 mm of Hg. There was no cyanosis and clubbing. Jugular venous pressure was raised. Cardiac examination revealed a grade 2/6 ejection systolic murmur in the tricuspid area. Her haemogram, liver and renal function was normal. Her coagulation profile were within normal limits. Chest x-ray was suggestive of enlarged cardiothoracic ratio of 0.7 with right atrial enlargement, left ventricular type of apex, and clear costophrenic angles and normal lung fields. Electrocardiogram showed atrial fibrillation.

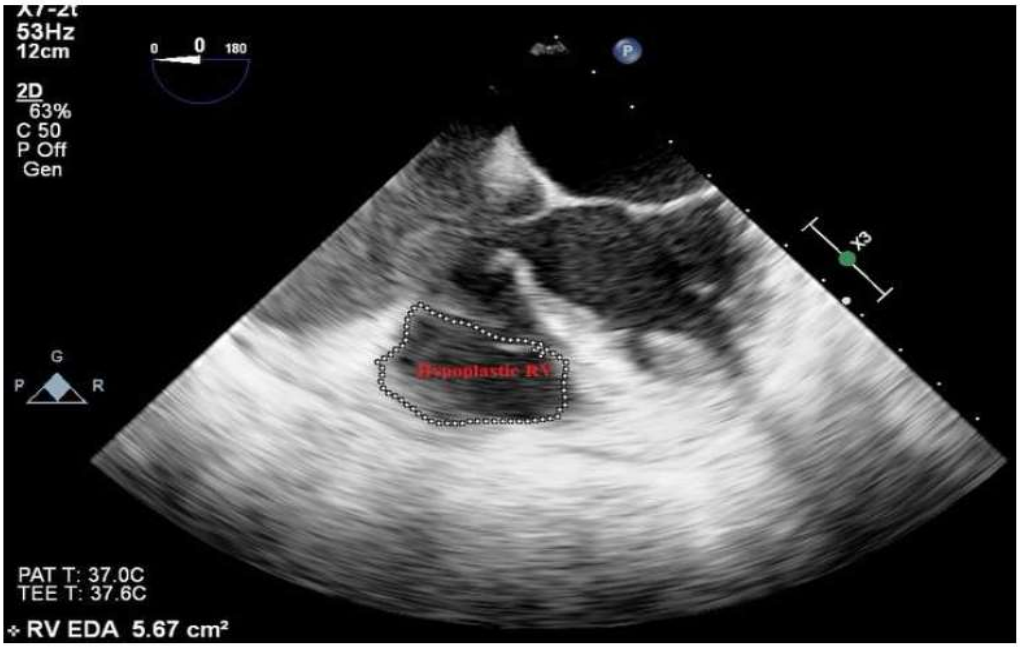

Fig. 1: Transesophageal Echocardiography (Midesophageal 4 Chamber view) showing Hypoplastic right ventricle.

Two dimensional echocardiography showed thrombus of size 3 × 1.9 cm seen in a hugely dilated right atrium. Ebstein's anamoly of the tricuspid valve with moderate low pressure tricuspid

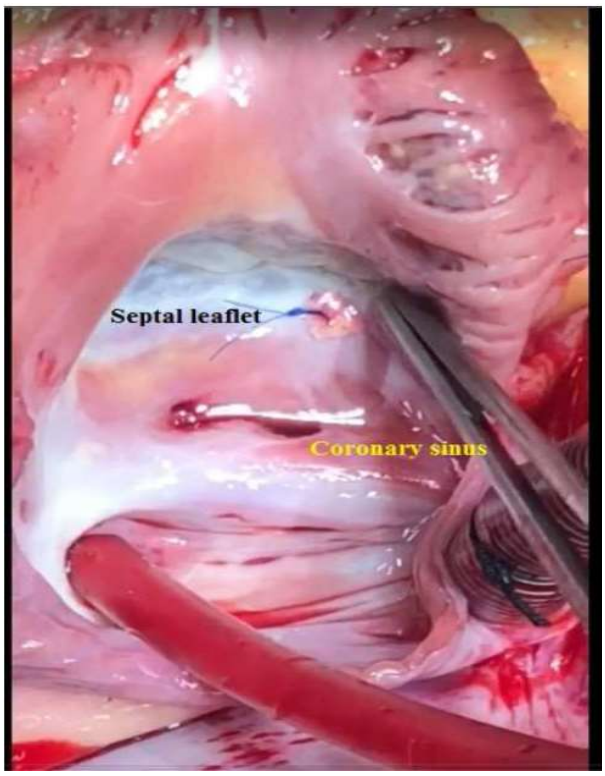

Fig. 2: Intraoperative image of Ebsteinoid tricuspid valve with no significant displacement of the leaflets repaired by Kaysannuloplasty

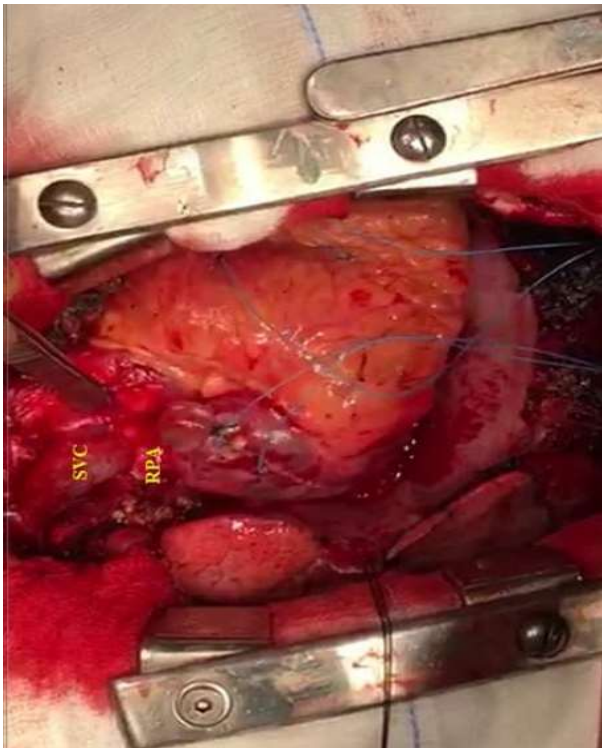

Fig. 3: Intraoperative image showing Right sided Bidirectional Glenn shunt as a part of one and half ventricle repair

regurgitation. True right ventricular cavity was only one third of the normal. Mild mitral regurgitation was noted with good left ventricular function. The interatrial and interventricular septa were intact. Computerized tomography was corroborative of echocardiographic findings and no evidence of pulmonary embolism. Cardiac magnetic resonance imaging showed non enhancing well defined lesion in the right atrium and features of Ebstein's anamoly with hypoplastic right ventricle.

The surgical procedure was performed by median sternotomy using moderately hypothermic cardiopulmonary bypass, and cold blood cardioplegic arrest. Intraoperatively, hugely dilated right atrium and hypoplastic right ventricle with almost one third of the normal cavity. The mean pulmonary artery pressure was 18 mmHg. Right atrium was opened. There was a huge thrombus in the right atrium and its appendage. The right atrial appendage was dilated. The tricuspid annulus was dilated with thin, fragile, non-coapting leaflets with moderate to severe tricuspid regurgitation on saline test. The septal tricuspid leaflet was not displaced significantly as in classical Ebstein's anamoly. However there was slight tethering of the septal leaflet to the septal wall seen. The main, right and left pulmonary arteries were of good size. The interatrial and interventricular septum were found intact. The coronary sinus, coronary arteries, and pulmonary venous drainage were normal. Innominate vein was present. There was no patent ductus arteriosus or left superior venecava. Though the right ventricular size was small, the classical ''atrialised portion" of the right ventricle was not noted. After removing the thrombus, thorough wash was given with normal saline. The tricuspid annulus size was reduced by Kays annuloplasty using simple figure of eight suture plication at the hinge of the septal and the posterior leaflet. The atrial septectomy of size 1 cm was performed. Right atrial closure was done and its appendage ligated. Right sided Bi-directional Glenn or Cavopulmonary anastomosis was performed by dividing the superior venecava at its junction with the right atrium followed by the end to side anastomosis to right pulmonary artery. The azygous vein was ligated. The patient was weaned off bypass with sinus rhythm and needed minimal inotropic support of dobutamine, milrinone and adrenaline. There was significant reduction in central venous pressure from 24 mm of Hg to 10 mm of Hg. Her postoperative course was uneventful. The duration of mechanical ventilation was 17 hours. The ionotropic support continued for four days. The critical care unit stay was for 5 days. She was discharged on day 8 in sinus

rhythm with prolonged QT interval. Postoperative echocardiography showed patent right bidirectional Glenn shunt to right pulmonary artery with normal phasic venous flow. Moderate size 14mm atrial septal defect shunting bidirectionally was seen. There was mild tricuspid regurgiatation and mild left ventricular dysfunction noted. However post-operative oxygen saturation was 91 percent on room air. Hence she needed administration of diuretics, angiotensin converting enzyme inhibitor Enalapril and oral anticoagulants postoperatively. Now, over 3 months after surgery, she is doing well.

DISCUSSION

The presentation of Ebstein's anamoly in adulthood depends on the structural anatomy. The greater the displacement of the leaflet insertion, more severe are the symptoms. The clinical manifestations are also dependent on the structural and functional changes in the right sided cardiac chambers, tricuspid valve morphology and other associated congenital malformation like atrial septal defect or patent foramen ovale. Arrhythmias are also commonly seen in such patients.[5] In our case patient had atrial fibrillation with dilated right atrium and hypoplastic right ventricle. The risk of paradoxical embolism is high in patients of Ebstein's anamoly with atrial septal defect or patent foramen ovale. Our patient was fortunate to have intact atrial septum which prevented the risk of paradoxical embolism of the large clot found in the right atrium. Our patient had no significant displacement of the tricuspid leaflets which can be accounted to the late onset of symptoms. Our case is a rare variant of Ebstein's anamoly. There is similarity in its clinical presentation with Ebstein's anamoly but the valve morphology is different. Hence we prefer to term this as the "Ebsteinoid tricuspid valve". The other differential diagnosis may be congenital unguarded tricuspid orifice, dysplasia of tricuspid valve with pulmonary atresia as well as intact interatrial septum and Uhlsanamoly which were excluded in our case intraoperatively.

The classical surgical techniques of Ebstein tricuspid valve repair have been demonstrated by Hunter, Lillehei and Hardy and Danielson. The goal of these surgeries is to restore the normal surface of leaflet coaptation. However, Ebstein's anamoly is a right ventricular disease as well. Thus in patients with hypoplastic right ventricle unable to support the pulmonary circulation, one and a half ventricular repair may be considered. The atrialised portion of the right ventricle in classical Ebstein's anamoly contributes significantly to the progression of the right ventricular impairment. Thus some techniques emphasize on the resection or plication of this dyskineticatrialised segment.[6] In our case as there was no such distinct atrialised portion we planned for Kays annuloplasty[7] to treat the dilated tricuspid annulus with moderate to severe tricuspid regurgitation. There was significant reduction in the tricuspid regurgitation in the post-operative period.

The option of the one and a half ventricle repair has emerged as a middle alternative path for the subset of patients with the anatomic or functional features of the right ventricle is borderline and neither suitable for a biventricular repair nor an univentricular pathway. This alternative helps in circumventing the early mortality of high-risk biventricular repair as well as the late complications after a Fontan operation.[8] The risk of known complications of Fontan circulation that includes inferior venecaval hypertension, chronic liver congestion, effusive complication, ascites, protein losing enteropathy, atrial arrthythmias, atrioventricular valve regurgitation, systemic ventricular failure encouraged us to attempt the one and a half circulation in our patient.

In our case, the patient had normal left ventricular function and mean pulmonary artery pressure of 16 mmHg with severely hypoplastic right ventricle. Thus we considered the patient for right sided bidirectional cavopulmonary connection or bidirectional Glenn shunt. In anticipation of the failing right ventricle we considered it rational to perform atrial septectomy. However the creation of the atrial fenestration led to significant arterial desaturation in the patient because of the bidirectional shunting seen post-operatively at the atrial level. Thus we would suggest atrial septectomy preferably only in neonatal patients of Ebstein's anamoly and failing right ventricle with elevated pulmonary resistance and to avoid the same in adults to prevent desaturation. Our patient may need iatrogenic atrial septal defectclosure by interventional method or Fontan procedure in future to furthur improve the functional capacity. We are now managing the patient on diuresis, anticoagulation and angiotensin converting enzyme inhibitors.

CONCLUSION

Ebstein's anamoly has distinct morphological and clinical variants. It is important to repair the tricuspid valve based on its structural and functional

analysis. One and a half ventricular repair is useful in patients of Ebstein's anamoly with hypoplastic right ventricle.

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