

# Ebstein's anomaly with constrictive pericarditis

Apurba Thakur<sup>1</sup>, Redoy Ranjan<sup>2</sup>, Mohammad Samir Azam Sunny<sup>3</sup>, Md. Aftabuddin<sup>4</sup>, Asit Baran Adhikary<sup>5</sup>.

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### Abstract:

**Ebstein's anomaly is a rare congenital cardiac malformation that affects the tricuspid valve commonly, but its association with pericardial disease is even rarer. We report a case of 25 year old man presented with dyspnoea on exertion and fatigability. A diagnosis of Ebstein's anomaly with atrial septal defect (ASD) with constrictive pericarditis was confirmed using transthoracic echocardiography. Peroperatively**

**pericardiotomy was done after meticulous dissection of pericardium. Under cardiopulmonary bypass tricuspid valve was replaced with 29 mm Edward life science porcine tissue heart valve with direct closure of ASD. Patient showed excellent symptomatic improvement and was discharged on 7<sup>th</sup> POD with advice and after 3 months of follow up patient was doing well post operative day.**

*Keywords: Ebstein's anomaly, tricuspid valve, atrial septal defect, constrictive pericarditis, tricuspid valve replacement.*

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### Introduction:

In 1866, Wilhem Ebstein published a scholarly description of a tricuspid valve anomaly that bears his name. On July 7, 1864, he performed an autopsy of a 19 year old, laborer, Joseph Prescher at Breslau and his findings were accurately described and illustrated with excellent drawings by his colleague Dr Wyss<sup>1</sup>. Ebstein's anomaly is a rare congenital heart disorder occurring in approximately 1 % per 200,000 live births and accounting for <1% of all cases of congenital heart disease<sup>2</sup>. The incidence is equal in both sexes.<sup>3</sup> Familial Ebstein's anomaly has been reported rarely.<sup>4</sup> The risk of Ebstein's anomaly exists especially if the lithium drug is taken as medication during 2-6 weeks post conception.<sup>5</sup> Ebstein's anomaly may present at any stage and has a highly variable clinical course. Fetal and neonatal presentation is associated with poor outcomes whereas older children

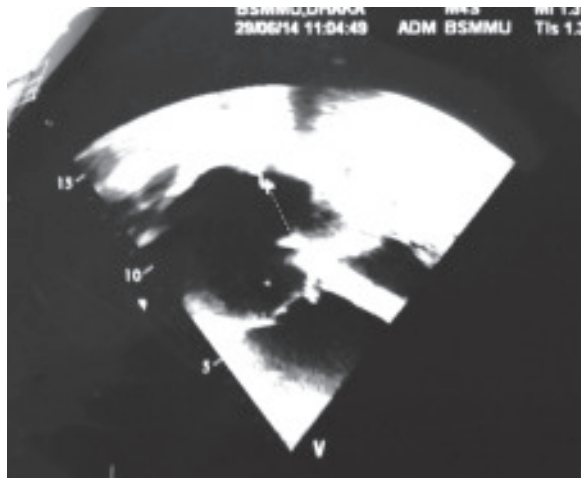
and adults long term outcome is superior.<sup>3</sup> Right ventricular abnormalities, tricuspid valve abnormalities and accessory conduction pathways (WPW syndrome) are the main primary pathophysiologic features predominating in Ebstein's anomaly.<sup>6</sup> An interarterial communication is present in 80-94 % of patients with Ebstein's anomaly. Additional anomalies include pulmonary atresia or stenosis, bicuspid or atretic aortic valves, ventricular septal defect, congenital mitral stenosis etc.<sup>7,8,9</sup> In adulthood, patients usually present with decreasing exercise tolerance, progressive cyanosis, arrhythmias or right heart failure. In the presence of interatrial communication, the risk of paradoxical embolisation, brain abscess and sudden death is increased.<sup>10</sup> Tricuspid valve repair with ASD closure is the preferred operation. However, in about 20 % to 30 % of patients, immobility or morphology of the tricuspid valve prevents repair and valve replacement is required.<sup>11,12</sup> Here, we are presenting a case of Ebstein's anomaly with constrictive pericarditis.

### Case Report:

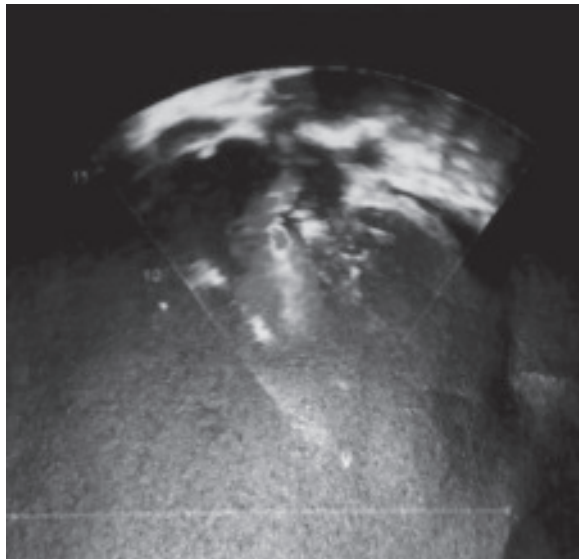
A 25 year old man was admitted in our department with chief complaints of dyspnea on exertion and fatigability for last four months. Physical examinations revealed pulse was 80 beats per minute, regular and normal in volume. Blood pressure was 110/70 mm Hg. His 1<sup>st</sup> heart sound was normal but 2<sup>nd</sup> heart sound was splitted

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1. MS Residency (Cardiovascular and Thoracic Surgery), Phase B, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka.
  2. MS Residency (Cardiovascular and Thoracic Surgery), Phase B, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka.
  3. Medical Officer, Department of Cardiac Surgery, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka.
  4. Professor, Department of Cardiac Surgery, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka.
  5. Professor, Chairman and Chief Special Unit, Department of Cardiac Surgery, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka.

at upper left parasternal border at 2<sup>nd</sup> intercostal space. There was a systolic murmur best heard at left lower parasternal border with inspiratory accentuation. Electrocardiogram revealed sinus tachycardia, right bundle branch block, right ventricular hypertrophy with inverted T waves in leads V<sub>1</sub> to V<sub>4</sub>. The chest X-ray showed marked cardiomegaly. Transthoracic colour doppler echocardiogram revealed a large ostium secundum, ASD (28 mm) with left to right shunt with PASP 53 mm Hg. Dilated RA, RV, PA with mild to moderate PAH. Ebsteinoid deformity of tricuspid valve (septal leaflet is plastered to underlying endocardium and anterior leaflet is sail like),

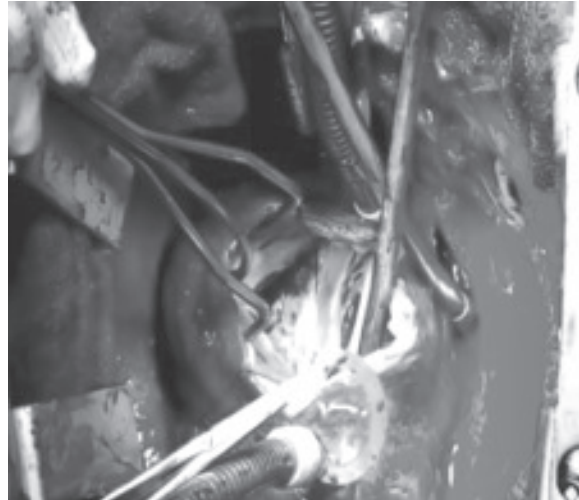


**Fig.-1:** Ebstein's anomaly of Tricuspid Valve (apical 4-chamber view).

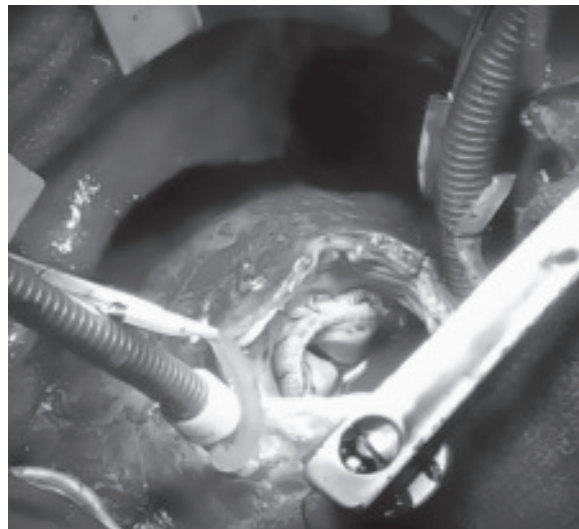


**Fig.-2:** Color Doppler across the ASD and the Tricuspid Valve.

tricuspid valve displacement from tricuspid annulus was 16mm, mild RV systolic dysfunction with TAPSE 12mm and good LV systolic function with LVEF = 76 %. Moderate pericardial effusion (19mm), paradoxical movement of



**Fig.-3:** Anterior leaflet of Tricuspid Valve.



**Fig.-4:** Tricuspid Valve Replacement with 29 mm Porcine Bioprosthesis Valve.

IVS. IVC is 15 mm with presence of restricted respiratory variations. The patient was operated on 21<sup>st</sup> July, 2014. Under general anaesthesia, with all aseptic precaution, standard median sternotomy was done. Pericardiectomy was done with difficulty because there was gross adhesion of pericardium. Cardiopulmonary bypass was established with aortic cannulation (24 F) and bicaval cannulation with SVC (30 F) & IVC (32 F). Heart was arrested with antegrade cardioplegia under mild

hypothermia (32° C). Right atriotomy was done. A large ASD, 20 X 6 mm, secundum, was detected. Tricuspid annulus was dilated. Septal leaflet was rudimentary. Anterior leaflet was sailed approximately 1cm displaced. Tricuspid valve was beyond repairable. Tricuspid valve was replaced with 29 mm Edward life science tissue heart valve with total preservation of subvalvular structures. Direct closure of ASD was done. Right atriotomy was closed in layers. Patient was weaned from bypass without any difficulty. Total cross clamp time was 35 minutes and total bypass time was 60 minutes. After achieving proper haemostasis, chest was closed leaving two mediastinal drainage (retrosternal 24 F) connected to under water sealed drainage. The patient was shifted to ICU with minimum inotropic supports and was extubated 8 hours after arrival at ICU. The patient was discharged on 7<sup>th</sup> POD with advice of taking tablets warfarin sodium 2.5 mg everyday for 3 months followed by antiplatelet therapy. Patient remains asymptomatic in the subsequent follow up and is leading to an almost normal life.

#### Discussion :

Ebstein's anomaly is a malformation of the tricuspid valve and the right ventricle characterized by adherence of the septal and posterior leaflets to the underlying myocardium; downward displacement of the functional annulus (septal>posterior> anterior); dilatation of the "atrialized" portion of the right ventricle; with various degrees of the hypertrophy and thinning of the wall; redundancy, fenestrations and tethering of the anterior leaflet and dilatation of the right atrioventricular junction(true tricuspid annulus) .<sup>13</sup> Regarding embryology, the leaflets and tensile apparatus of the tricuspid valve are believed to be formed mostly by the process of delamination of the inner layers of the inlet zone of the right ventricle. The downward displacement of the leaflets in Ebstein's anomaly suggests that delamination from the inlet portion failed to occur.<sup>14</sup> In our case, peroperative findings were almost similar with others.<sup>13,14</sup> It is not uncommon for Ebstein's anomaly to be undiagnosed until adulthood. The mean age of diagnosis in a study of the natural history of 72 unoperated patients was 23.9±10.4 years.<sup>15</sup> As our reported case was 25 years of age which is consistent with some other authors.<sup>15</sup> The cardinal symptoms in Ebstein's anomaly are cyanosis, right-sided heart failure, arrhythmias, and sudden cardiac death.<sup>10</sup> Children more than 10 years of age and adults often present with fatigue, right-sided heart failure and arrhythmias<sup>10</sup>. Our patient had complaints of dyspnea on exertion and fatigability

for four months. A systolic murmur may be heard at lower left sternal edge due to tricuspid regurgitation.<sup>6</sup> Regarding our patient, 2nd heart sound was widely splitted and fixed at upper left parasternal border and a systolic murmur was heard at lower left parasternal border. Chest radiograph showed marked cardiomegaly with a rounded or boxlike cardiac contour with normal or oligemic lung fields.<sup>6</sup> In our patient, chest radiograph showed only cardiomegaly. The ECG is abnormal in most patients with Ebstein's anomaly. It may show tall and broad P waves with complete or incomplete right bundle-branch block patterns and bizarre morphologies of the terminal QRS pattern result from infra-Hisian conduction disturbance and abnormal activation of the atrialized right ventricle. From 6% to 36% of patients with Ebstein's anomaly have more than one accessory pathway and most accessory pathways are located around the orifice of the tri-cuspid valve. First-degree atrio-ven-tricular block occurs in 42%.<sup>16,17,18</sup> Wolff-Parkinson-White syndrome is found in 30-50%.<sup>17</sup> In our case ECG was showing sinus tachycardia with right bundle branch block with right ventricular hypertrophy and inverted T waves in leads V<sub>1</sub> to V<sub>4</sub>. Two dimensional echocardiography is the diagnostic test of choice for Ebstein's anomaly. Echocardiography allows accurate evaluation of the tricuspid valve leaflets and the size and function of the cardiac chambers. The principal feature of Ebstein's anomaly is apical displacement of the septal leaflet of the tricuspid valve from the insertion of the anterior leaflet of the mitral valve by at least 8 mm/m<sup>2</sup> body surface area.<sup>6</sup> Our patient had 16mm displacement of septal leaflet on echocardiography. Our patient have Great Ormond Street Ratio (GOSR) score 2. Echocardiographing finding of constrictive pericarditis are left ventricular free wall flattening, paradoxical motion of interventricular septum( septal bounce), premature opening of the pulmonary valve, inferior venacava and hepatic vein dilatation with restricted respiratory variation.<sup>23</sup> In our case, paradoxical motion of the IVS and IVC dilatation with restricted respiratory variation were present. Tricuspid valve repair with direct ASD closure is the preferred operation. However in 20-30% of patient, valve replacement is required.<sup>6</sup> Porcine bioprosthetic valve remains a good alternative. Most prefer bioprosthesis to mechanical valves due to relatively good durability and lack of need for anticoagulation.<sup>19</sup> A bioprosthetic valve may offer superior late survival when compared with a mechanical valve when tricuspid valve replacement is required in patients with Ebstein anomaly.<sup>20</sup> Heart transplant is rarely necessary for Ebstein's anomaly. Its indication is usually the presence of severe biventricular dysfunction (LVEF

<25 %).<sup>2</sup> In our, patient tricuspid valve apparatus were beyond repairable and tricuspid valve was replaced with 29 mm Edward life science porcine tissue heart valve. Ebstein's anomaly with pericardial diseases are rarely reported in the literature.<sup>24</sup> In our case, pericardium was densely adherent to the anterior surface of pericardium. No operative sample of the pericardium was taken for histopathological examination. The cause of pericarditis was chronic inflammation as our patient was a case of congenital heart disorder (Ebstein's anomaly with ASD). A 35 years old lady with diagnosis of Ebstein's anomaly with pericardial disease was reported in 2005.<sup>21</sup> Similarly, Ebstein's anomaly with significant pericardial effusion was reported in 1998<sup>22</sup>. Our patient had Ebstein's anomaly with constrictive pericarditis.

#### Conclusions:

Ebstein's anomaly with constrictive pericarditis has been reported in medical literature<sup>24</sup>. Untreated Ebstein's anomaly with constrictive pericarditis can be effectively managed with surgical treatment.

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