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# An Unusual Cause of Gradual Increasing Shortness of Breath in a 75 Years Old Man

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## Abstract

Ebstein anomaly is a congenital abnormality of the tricuspid valve in which there is apical displacement typically of the septal leaflet as well as tethering of the lateral leaflet to the ventricular wall. This results in coaptation of the tricuspid leaflets in a position displaced toward the right ventricular apex and creates an atrialized portion of the right ventricle. The degree of displacement can be highly variable and can range from as little as 12 mm to several centimeters. A variety of cardiac abnormalities are associated with Ebstein's anomaly, including atrial septal defect, conduction system abnormalities, patent foramen ovale, pulmonary stenosis or atresia and ventricular septal defect. Ebstein's anomaly is mild in most adults who have it, so they do not need surgery. But sometimes the tricuspid valve leaks severely enough to result in heart failure or cyanosis. Then surgery may be required. When patients of all ages are taken together, the predicted mortality is approximately 50% by the fourth or fifth decade. We present here an uncommon presentation of this complex congenital heart defect in a 75 years old man.

## 1. Introduction

Ebstein's anomaly is a rare congenital heart defect occurring in 1 per 200000 live births and accounting for 1% of all cases of congenital heart disease. This anomaly was described by Wilhelm Ebstein's in 1866 in a report titled, "Concerning a very rare case of insufficiency of the tricuspid valve by a congenital malformation."<sup>1</sup> The clinical spectrum is variable and is heavily dependent on the severity of the deformity and the presence of associated defects.<sup>2</sup> Patients surviving into adulthood without surgical valve repair or replacement may develop a variety of signs and symptoms, including cyanosis (because of right to left shunting across an ASD or patent foramen ovale), dyspnea and fatigue (due to decreased cardiac output), or palpitations.

Ebstein's anomaly is an abnormality of the tricuspid valve in which the septal leaflets and often the posterior leaflet are displaced into the right ventricle and the anterior leaflet is usually malformed, excessively large, and abnormally attached or adherent to the right ventricular free wall.<sup>3</sup>

## 2. Case History

The patient, Mr. Liakot Ali, 75 years of age, smoker, non-Diabetic having gradual increasing of shortness of breath and exertional chest pain for last 1 month. He was totally asymptomatic before 1 month and living active life. Initially he had breathlessness and chest pain after heavy work which at present is even at rest. On examination, pulse rate

110/minute, BP: 130/90 mmHg, moderate bilateral leg oedema, bilateral basal lungs crepitation. Pan-systolic murmur in tricuspid area (Grade: 2/5) and audible 3<sup>rd</sup> heart sound. Electrocardiogram (ECG) shows right bundle branch block and right atrial enlargement (RBBB & RAE), CXR: hugely enlarged cardiac shadow with lung congestion. Echocardiogram Shows rudimentary/small size Right ventricle (RV), Large size right atrium (RA), Apical displacement of septal cusp of TV (23.0 mm), anterior and

septal wall of Left ventricle (LV) hypokinetic, LV EF-45% to 48%. His complete blood count, renal function, liver function tests were normal. So our patient diagnosed as a case of Ebstein's anomaly with Tricuspid regurgitation (Grade-2/5) with mild pulmonary hypertension (PASP-40 mmHg) with ischemic heart disease with bi-ventricular heart failure. After getting adequate treatment now patient is symptoms free and resume his ordinary activity.



*Fig. 1. Chest X-ray showing Cardiac shadow enlarged.*

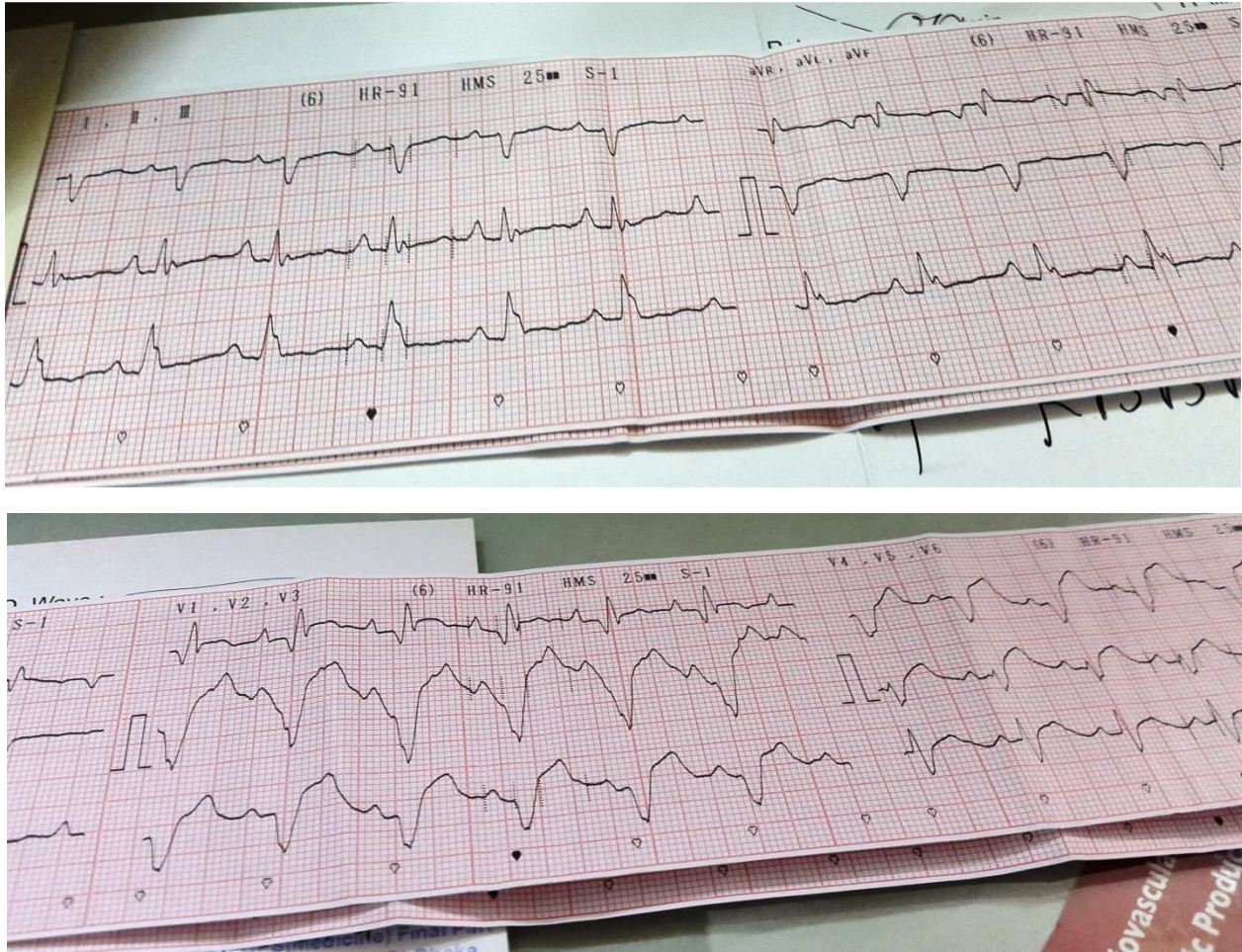


Fig. 2. Electrocardiogram showing complete right bundle branch block and right atrial enlargement.

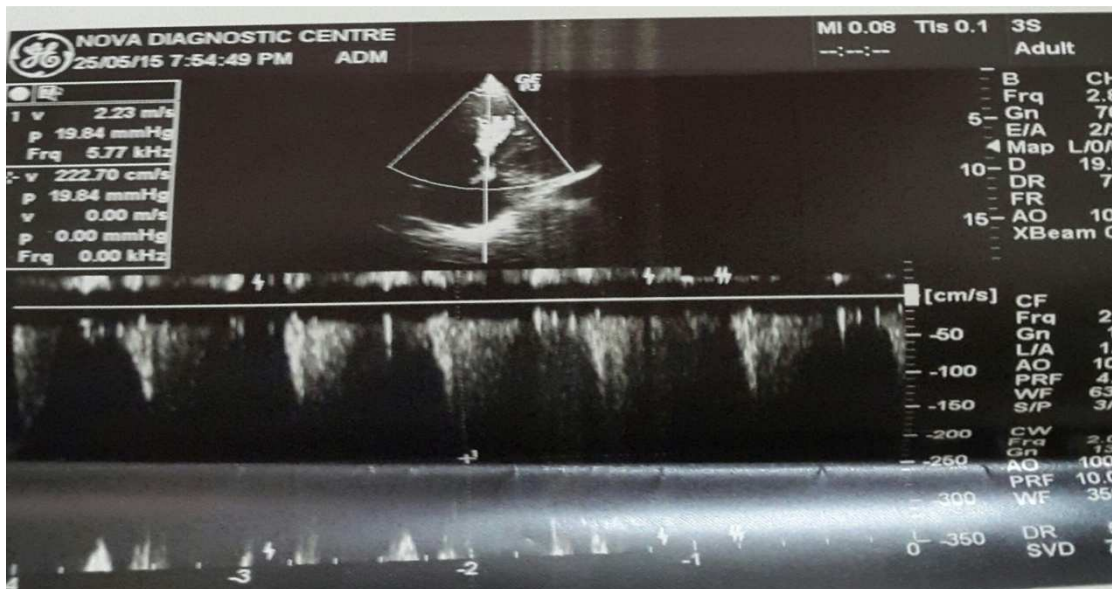


Fig. 3. Echocardiogram with color flow mapping (CFM) and CW showing tricuspid regurgitation.



Fig. 4. Apical displacement (23mm.) of septal cusp with elongated anterior leaflet giant right atrium with small right ventricle.

### 3. Discussion

The natural history of patients with Ebstein's anomaly depends on its severity. When the tricuspid valve deformity is severe, symptoms usually develop in newborn infants. Few similar cases of Ebstein's anomaly are found in their natural course at advanced ages.<sup>4</sup> It is known that only 5% of these patients survive naturally beyond their 5<sup>th</sup> decade of life. It can be stated that this anomaly is well tolerated provided the RV function is preserved, as seems to have been the case in this patient.<sup>5</sup> Supraventricular cardiac arrhythmia, such as atrial fibrillation, adds more risk to this course, given the greater possibility of thrombi formation and embolism.<sup>6</sup>

Approximately one-half of reported patients develop symptoms of cyanosis and right-sided heart failure in early infancy. Mild forms of Ebstein's anomaly may not cause symptoms until later in adulthood. Even some people with very abnormal valves may have minimal, if any, problems. Ebstein's anomaly symptoms may develop slowly over many years and include: shortness of breath on exertion, palpitation, leg swelling, and cyanosis.<sup>7</sup>

The best echocardiographic view for the evaluation of Ebstein's anomaly is the four-chamber view. Of principal importance is the accurate recording of the level of insertion of the septal leaflet of the tricuspid valve relative to the annulus. Apical displacement of this insertion site is optimally assessed in this view and is the key to diagnosis.<sup>8</sup>

Treatment of Ebstein's anomaly is complex and dictated mainly by the severity of the disease itself and the effect of accompanying congenital structural and electrical

abnormalities. Treatment options include medical therapy, radiofrequency ablation, and surgical therapy. Antibiotic prophylaxis for infective endocarditis, treatment for heart failure and anti-ischaemic drugs if associated IHD (Ischemic heart disease) present.<sup>9</sup> The more common surgical approach has been tricuspid valve reconstruction or replacement, usually with bioprosthesis.<sup>10</sup> Other approach, to avoid TV replacement several repair techniques including annuloplasty, closure of commissures, and creation of a "monocusp" valve.

### 4. Conclusion

In any case, the management of patients with Ebstein's anomaly is complex and has to be individualized. Patient with Ebstein's anomaly should receive continued care from a cardiologist with expertise in congenital heart defects. Patient with mild tricuspid valve leakage are unlikely to require surgery but moderate to severe regurgitation may need initial or subsequent surgery. Tricuspid valve repair with plication of the atrialized ventricle was effective in improving the functional status of an older patient with a late presentation.

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