Case 1/2015 A 76-year-old Male Patient with Ebstein Anomaly in Natural Course

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Clinical data: Shortness of breath on mild to moderate exertion for 2 months accompanied by heart palpitations. The patient did not report any previous heart disease, had never received specific medication throughout life and did not report cyanosis. He was started on furosemide, hydrochlorothiazide, acetylsalicylic acid (ASA) and losartan, with improvement of symptoms. Laboratory tests showed H= 6,800,000/mm^3; Hct = 63%; Hg = 20.6 g/dL; BUN = 103; creatinine = 2.03; BNP = 286.

Physical examination: normal breathing ; with grade 1/4 cyanosis; arrhythmic pulses, grade 2/4 jugular venous distension. Weight 65 kg, height 170 cm, blood pressure (BP) 100/60 mmHg, heart rate (HR) 82 bpm, oxygen saturation 88%. Grade 1/4 aorta (Ao) palpable on the suprasternal notch.

In the precordium, apical impulse diffusely palpable on the fourth and fifth left intercostal space and slight systolic impulses on the left sternal border. Diminished heart sounds; grade 1/6 coarse systolic murmur on the left sternal border and apex. Liver not palpable.

Laboratory tests

Electrocardiogram (ECG) showed atrial fibrillation, signs of right ventricular (RV) diastolic overload, and left anterior superior hemiblock. Low-voltage QRS complex with qr morphology in V1; RS in V6, with final conduction disturbance in the right bundle branch. QRSA: - 70º, TA: + 30° (Figure 1).

Chest radiograph showed moderately enlarged cardiac silhouette (cardiothoracic ratio: 0.61), resulting from enlarged right chambers. The pulmonary vascular network was slightly decreased, the arch of the pulmonary artery was concave, and the aortic knob was prominent. (Figure 1).

Echocardiogram (Figure 2) showed a thickened dysplastic tricuspid valve with a 19-mm fusion of the septal cusp and defective central coaptation, as well as marked tricuspid regurgitation. The right chambers were much dilated (right ventricle (RV) = 47 mm; left ventricle (LV) = 38 mm; left atrium (LA) = 41 mm; (right atrium (RA) = 36 mm), with the area of the right atrium added to that of the atrialized RV (with 56.5 cm^2), of the functional RV (with 17.6 cm^2), of the LA (with 13.9 cm^2) and LV (with 15.6 cm^2). The Celemajer index was calculated at 1.2. The inferior vena cava was dilated, with 23 mm, with spontaneous contrast enhancement. The tricuspid ring had 57 mm, the mitral ring 35 mm, the pulmonary ring 20 mm, and the aortic ring 24 mm. The LV ejection fraction was 66%, and the pulmonary arteries had 14 mm. There was a foramen ovale with bidirectional shunt and the atrial septum bulged to the left. RV systolic pressure was 30 mmHg.

Clinical diagnosis: Ebstein anomaly with marked tricuspid valve dysplasia and severe tricuspid regurgitation, with mild chronic hypoxemia in natural course.

Clinical reasoning: The clinical elements of cyanotic heart disease with long-standing low pulmonary flow and hypoxemia are expressed by shortness of breath and increased hematocrit. The diminished heart sounds suggest normal arterial position and low pulmonary flow resulting from severe tricuspid regurgitation with mild systolic murmur due to contiguous low-pressure chambers. The right atrium, the atrialized RV and functional RV behave as a true single chamber. RV diastolic overload on the electrocardiogram (ECG) suggests the diagnosis of tricuspid regurgitation. Chest radiograph shows markedly increased right chambers with slightly decreased pulmonary vascular network. Taken together, these elements point to the diagnosis of Ebstein anomaly.

Differential diagnosis: Heart diseases with hypoxemia and dilatation of right chambers are also found in their natural course in long-standing pulmonary stenosis, however with RV systolic overload. Other heart diseases uncommonly manifest as seen in this case.

Management: In face of the long-standing hypoxemic consequences, with unfavorable clinical manifestations, cardiac surgery was considered for tricuspid valve replacement despite the higher risk due to the advanced patient age. For this purpose, other studies of RV viability are necessary to more adequately determine the RV function and, in turn, to support the best treatment option. As a precautionary measure, medical treatment was chosen aiming at treating the cardiac arrhythmia and heart failure, mainly considering the advanced patient age.

Commentaries: Few similar cases of Ebstein anomaly are found in their natural course at advanced ages[1-3]. It is known that only 5% of these patients survive naturally beyond their 5th decade of life. It is a fact that, even with severe tricuspid regurgitation, cases that extrapolate the cardiac compensation phenomena for such a long time are sometimes identified. 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. However, in the presence

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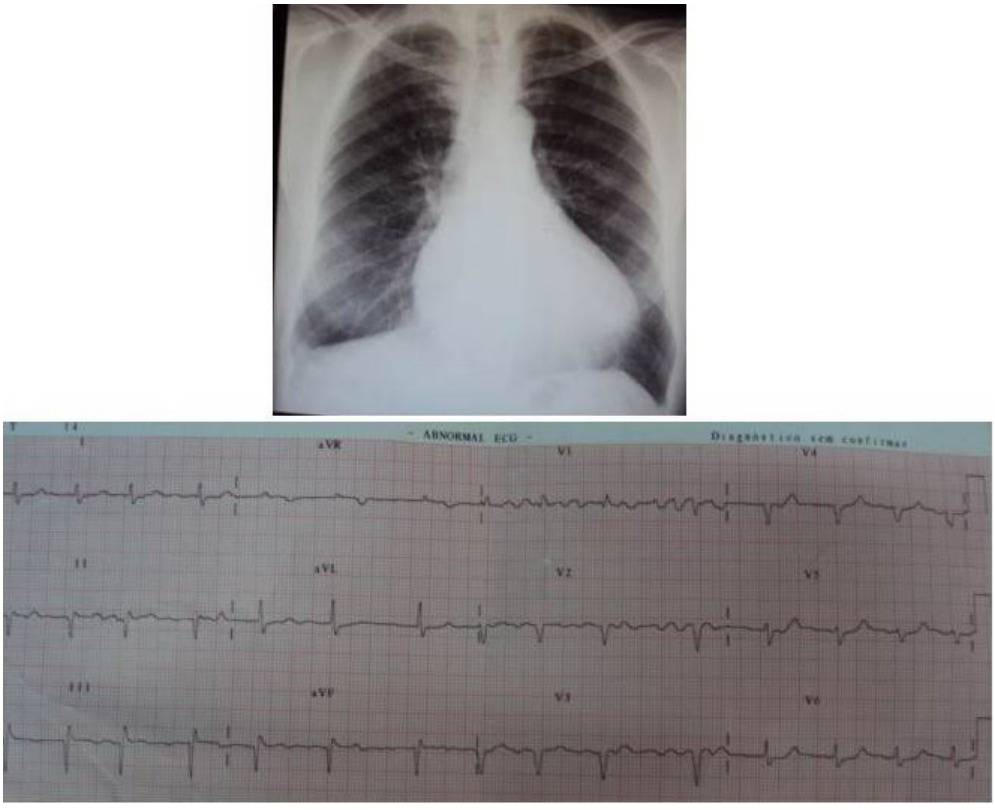

Figure 1 – *Chest radiograph showing enlarged cardiac silhouette due to enlarged right chambers, with pulmonary vascular network close to normal. Electrocardiogram showing atrial fibrillation with right ventricular diastolic overload and left anterior hemiblock.*

of RV dysfunction, the course of the disease is shorter, due to the higher volume overload. Supraventricular cardiac arrhythmia, such as atrial fibrillation, adds more risk to this course, given the greater possibility of thrombi formation and embolism. Even aware of the potentially unfavorable course, it is difficult to indicate surgery because of acquired conditions such as myocardial fibrosis and hypoxemic changes in other organs such as the kidneys and liver, which undoubtedly pose greater risks to elderly patients. Hence, the logical advice for an expectant medical management.

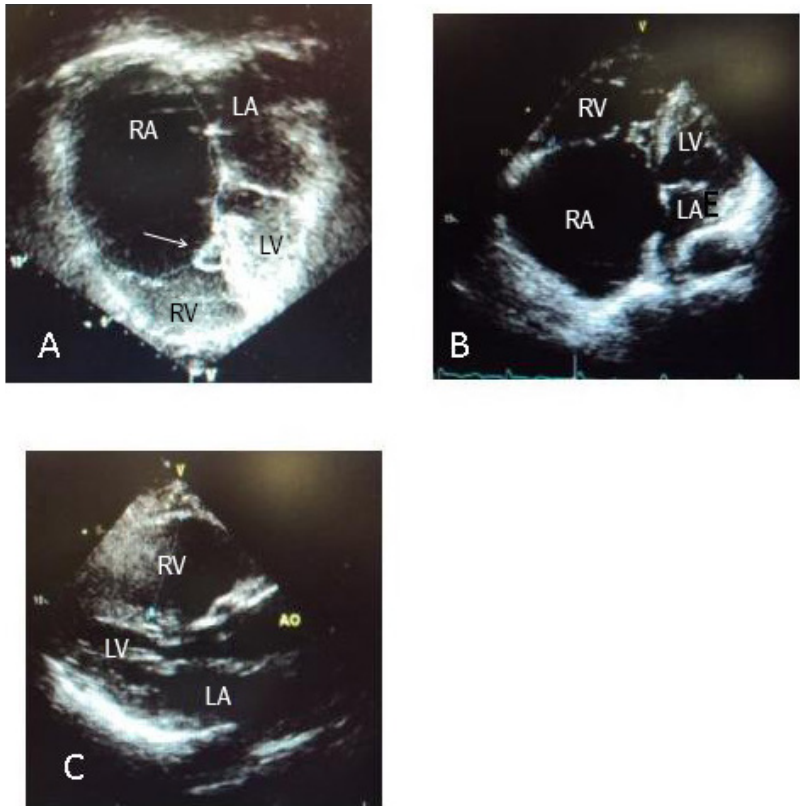

Figure 2 – *Apical four-chamber echocardiogram showing severe enlargement of right cardiac chambers, in A, with clear fusion of the septal cusp in the interventricular septum (arrow). The interatrial septum is bulged to the left and the left chambers are clearly displaced. Subcostal view, showing markedly enlarged right atrium (RA), in B; long-axis view showing, markedly enlarged right ventricle (RV), on the sternal border, with left ventricle (LV) displaced posteriorly, in C. LA: left atrium.*

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