

## Case 6/2013 – 56 years old Woman with Ebstein Anomaly in Heart Failure

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**Clinical Data:** Progressive fatigue on exertion has been noted for two decades, despite operative intervention 10 years ago for the closing of interatrial communication, valvar and tricuspid annulus plastia, and plication of the right atrium. Currently, it is presented with fatigue for small efforts, lower limbs edema and in use of furosemide, hydrochlorothiazide, amiodarone and spironolactone.

**Physical exam:** Slightly dyspneic, cyanotic, diminished pulses. Clear jugular turgence at 30°. Weight: 61 kg. Height: 154 cm. Blood Pressure (BP): 100/65 mmHg. Heart Rate (HR): 80 bpm. Oxygen Saturation = 95%. The aorta was not palpable at suprasternal notch.

In precordium, *ictus cordis* was palpable diffusely and it had discrete systolic impulses in the left sternal border. The heart sounds were markedly hypophonetic and no cardiac murmur was auscultated. The liver was palpable 3 cm from the right costal margin, with discrete ascites. Lower limbs edema was moderate.

### Supplementary tests

**Electrocardiogram** it showed junctional rhythm and signs of right bundle branch block with QRS complex duration (QRS) of 0.16' and low voltage in all leads. Electrical axis of the QRS complex (QRS): +120°, electrical axis of the T wave (AT): +180°.

**Chest Radiography** showed an enlarged cardiac area in very marked degree (cardiothoracic ratio of 0.78), with long and globular atrial and ventricular arches. Pulmonary vascular markings were markedly decreased.

**Echocardiogram** (Figure 1) showed marked increase in right heart chambers, absent septal, anterior and posterior valves mobility and important degree of insufficiency. The right ventricle was so dilated that occupied the apex of the heart. The left cavities were very depressed and compressed by the accentuated increase in the right cavities. Inferior vena cava with 31 mm and suprahepatic veins very dilated and with spontaneous contrast. Left superior vena cava drained in dilated coronary sinus. The tricuspid annulus measured 58 mm in relation to the mitral annulus of 26 mm. The area of the right atrium was 90.1 cm<sup>2</sup> (normal value < 18 cm<sup>2</sup>)

### Keywords

Heart failure; Ebstein anomaly / surgery; Cardiomegaly.

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and with spontaneous contrast. The right ventricle dilated so occupied the apex of the heart, showing pronounced systolic and diastolic dysfunction with Myocardial Performance Index (MPI) = 0.92.

**Chest computed tomography** (Figure 1) stress out the same aspect showed by the echocardiography, calculating that the final diastolic volume of the right ventricle was of 508 mL/m<sup>2</sup> and left ventricle was 48 mL/m<sup>2</sup>. Corresponding function of both ventricles, right and left, was of 15 and of 68%, respectively.

**Blood biochemical analysis:** Hemoglobin: 11 g/dL, urea: 61 mg/dL, creatinine: 2.07 mg/dL; Brain Natriuretic Peptide (BNP): 684 pg/mL.

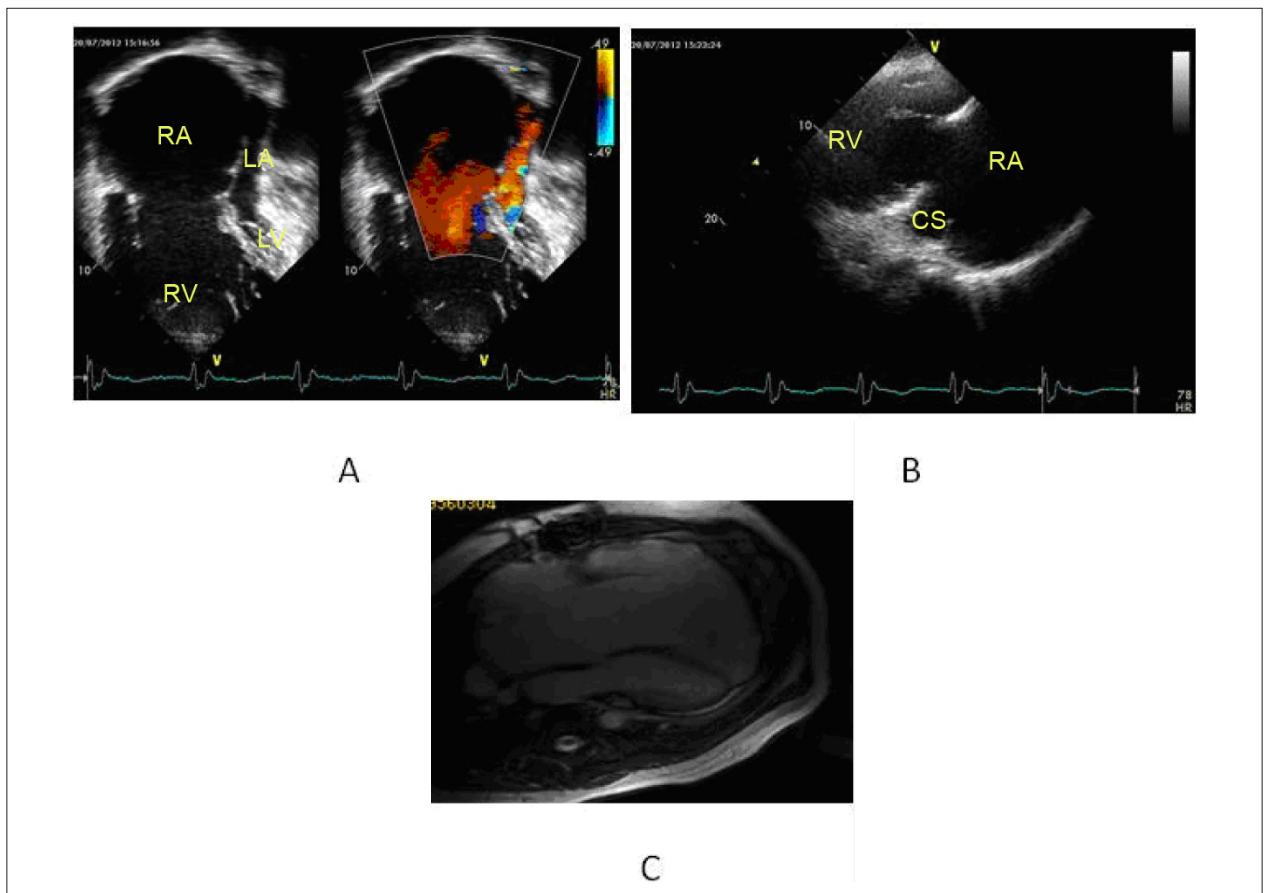
**Clinical Diagnosis:** Ebstein Anomaly in severe heart failure, even after operative correction, 10 years ago.

**Clinical Rationale:** Clinical findings were consistent with the diagnosis of right heart failure due to Ebstein anomaly with severe dysfunction of the right ventricle and tricuspid valve. The absence of heart murmur expressed that the right cavities formed a single cavity with firm adherence of the tricuspid valve into the right ventricular muscle. The great cardiomegaly seen on chest radiograph with decreased pulmonary vascular markings guided to the diagnosis of this anomaly. Other imaging tests, such as echocardiography and cardiac CT, consolidated the severity of the anomaly by severe dilation and right ventricular dysfunction.

**Differential Diagnosis:** The clinical profile of right heart failure allied with the severe increase in the cardiac area seen on chest radiography, could also be present in pericardial processes, which are accompanied by severe pericardial effusion or pericardial cysts and yet in cardiac tumors.

**Conduct:** Due to the strong repercussion of the Ebstein anomaly with heart failure with functional class IV, extreme dilatation of the right cavities with clear compression of the left cavities and with cardiac arrhythmia, that heart transplantation was considered as the only viable therapeutic solution. The surgical repair of Ebstein anomaly with necessary replacement of the tricuspid valve would not be enough to reverse the long-standing ventricular dysfunction and under extreme operative risk.

**Comments:** Despite the Ebstein Anomaly being constituted in the greater longevity among all congenital heart diseases, rarely, however, it reaches the sixth decade of life. The unfavorable evolutionary problems, frequently in adulthood are related to right heart failure, to supraventricular and ventricular arrhythmias and thromboembolic phenomena. Therefore, most of these defects must be operated in the first decade of life, even without strong hemodynamic disturbances, particularly because it is today considered that cardiac surgery is corrective and even curative, as it happens with the cone technique introduced in our means by Da Silva<sup>1</sup>.



**Figure 1** - Echocardiography in the apical 4-chamber view stands out the diagnostic elements of Ebstein anomaly with tricuspid septal valve coupling with strong increase of the right cavities responsible for the extreme rebound of the left cavities (A), strong enlargement of right atrium and coronary sinus with tricuspid posterior valve still in subcostal view (B), and the contrast of the right cavities very enlarged and compressed left cavities in cross section view of the chest CT (C). RA: right atrium; LA: left atrium; CS: coronary sinus; RV: right ventricle; LV: left ventricle.

## References

1. da Silva JP, da Silva Lda F. Ebstein's anomaly of the tricuspid valve: the cone repair. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu.* 2012;15(1):38-45.