

## Corrected Transposition of the Great Arteries: Late Clinical Presentation, in the Fifth Decade of Life

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The corrected transposition of the great arteries, rare congenital cardiopathy, is related to the largest incidence of cardiological complications. We report a case in which the clinical presentation of the disease occurred in the fifth decade of life, with tricuspid insufficiency, occasion that the patient was submitted to valvar replacement.

### Introduction

Congenital cardiopathies present prevalence from 3,7 to 8/1.000 alive born (4,8/1.000 in Latin America), 5% to 7% of the which present transposition of the great vases, anatomically defined by atrioventricular concordance and ventriclearterial discordance. The transposition of the great vases is the main cause of cyanotic cardiopathy in the neonatal period, and has predominance in the masculine gender (60% to 70%). The corrected transposition of the great arteries (CTGA) is characterized by concomitant atrioventricular and ventriclearterial discordance, with approximate prevalence of 0,5% to 1,4% of all of the congenital cardiopathies<sup>1,2</sup>. It is commonly associated to several congenital defects, total atrioventricular block and right ventricular dysfunction, what limits the survival of these patient, who do not, in great majority, live over 50 years of age<sup>3</sup>.

This condition can remain asymptomatic for several years in individuals without associated cardiopathies, can only present electrocardiogram (ECG) or x-ray with alteration, due to the unusual position of the ventricles and vases. Over the time, the systemic ventricle can become insufficient, fail, due to pressure overload, causing fatigue and dyspnea. The treatment of this condition depends on the clinical presentation and the associated defects, can go from the clinical treatment, with drugs (inhibitors of angiotensine-converting enzymes, diuretic, digitalization), to the palliative surgical treatment (physiological) or definitive (anatomical), with the arterial replacement.

### Key words

Transposition of great vessels; tricuspid valve insufficiency; heart defects, congenital.

### Case Report

Patient female, 47 years of age, with diagnosis of CTGA at 18 years of age (after evaluation motivated by non-specific thoracic discomfort to the left during physical exercise), accompanied periodically by cardiologist and gave up sporting activities. Received diagnosis of systemic arterial hypertension at 40 years of age.

About four years ago, tricuspid (systemic valve) insufficiency was diagnosed, through echocardiography, in periodic exam, developing in the last two years with dyspnea and progressive fatigue (functional class III by New York Heart Association). The patient, then, was referred to Casa de Saúde São José for surgery (elective) of valvar replacement. At physical exam, in the preoperative, presented blood pressure of 140/70 mmHg, pulse of 76 bpm, irregular heart rhythm (chronic atrial fibrillation [AF]), ictus diverted to the sixth intercostal space (previous axillary line), holosystolic murmur (4+/6+) more evident in mitral focus with irradiation to dorso, B3 in the tip and fine stertorous breathing in lung bases. The laboratory exams (blood count, biochemistry), in the preoperative, were shown unaffected, and ECG demonstrated AF rhythm. The preoperative thorax x-ray (fig. 1a) evidenced cardiomegaly, aortic arch to the right and prominent hilum.

Preoperative echocardiogram (ECHO) revealed systemic tricuspid atrioventricular valve (AV), with serious regurgitation to Doppler in colors, right atrioventricular AV valve bicuspid, diameter of the left atrium of 4,5 cm, and ejection fraction (Teichholz) of 56,5%. Transesophageal perioperative ECHO (fig. 2) demonstrated (pre- extracorporeal circulation) ventricular inversion associated to the transposition of the great arteries, with anatomical preservation of the atria and venous drainages, right ventricle (RV) dilated, with moderate dysfunction and diffuse hypokinesia of their walls, systemic AV valve with significant reflux, with thickened leaflets and lack of coaptation due to prolapse of the antero-septal leaflets.

During surgery, tricuspid systemic AV valve was found, with aspect of myxomatous degeneration and with rupture of papillary muscle, this substituted by metallic valve St. Jude® nº 31 (fig. 1b). The patient developed without intercurrents in the postoperative, was released from hospital in six days, using cumarín, captopril, spironolactone and furosemide. Option was made to maintain clinical treatment of the AF and continuous ambulatorial surveillance.

### Discussion

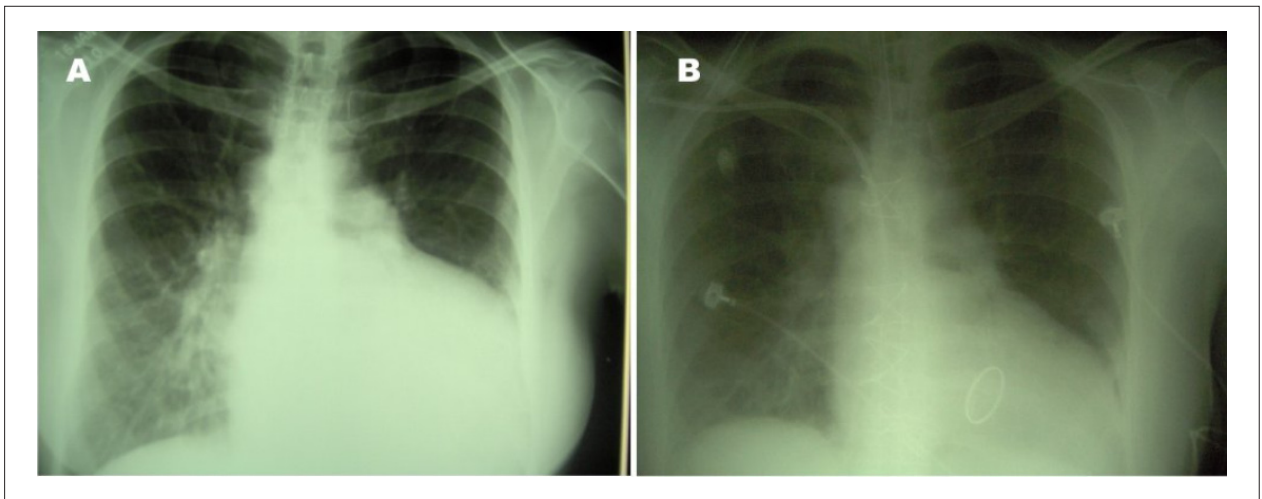
CTGA is a rare congenital cardiopathy. When there are no associated defects, as in the subject case, it is still rarer,

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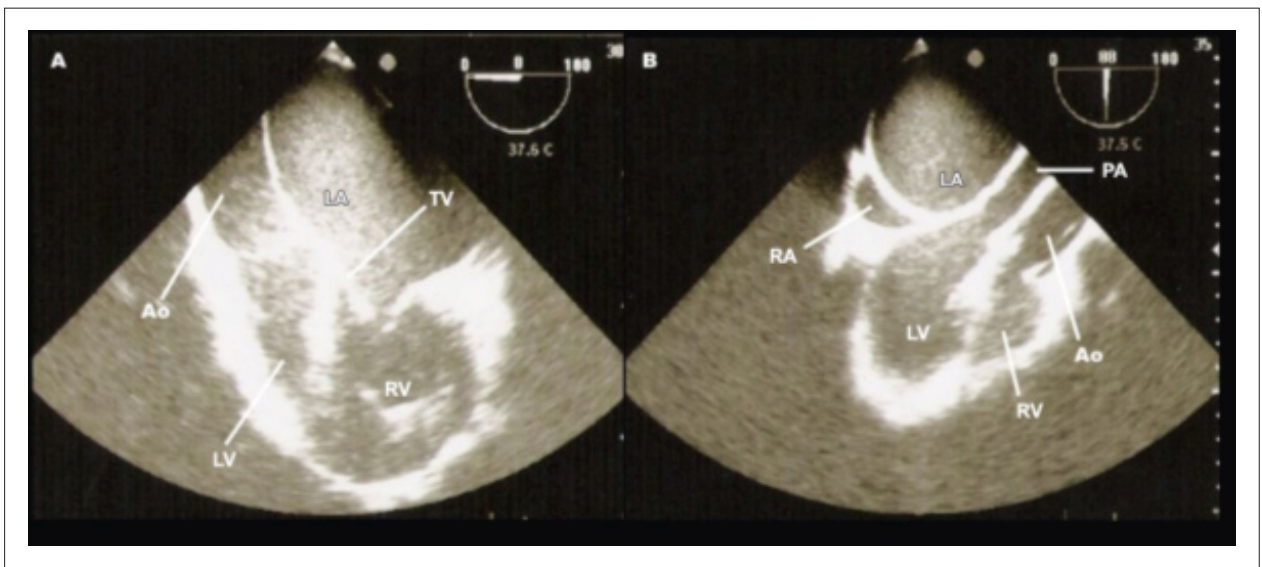
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**Figure 1** - In A, preoperative thorax x-ray, evidencing cardiomegaly and aortic arch to the right; In B, presence of the metallic valve in left atrioventricular location(mitral topography), in immediate postoperative.



**Figure 2** - Perioperative echocardiogram of the patient; In A, ventricular inversion and tricuspid valve closer to the heart apex; In B, anatomy of the heart and great arteries, in another incidence and with more details; LA - left atrium; RA - right atrium; LV - left ventricle; RV - right ventricle; Ao - aorta; TV - tricuspid valve; PA - pulmonary artery.

in 1% of the cases<sup>1,2</sup>. In these situation when there is no associated defect, the survival is longer. For these factors (absence of associated defects, lingering evolution and late presentation, with more than four decades of clinical silence), the relevance and the peculiarity of the described case can be noticed. The natural history of this condition is not very well defined and it depends on the associated defects and of their repercussion<sup>1,2</sup>, as well as of the capacity of RV to preserve its function. Several authors have been studying the prevalence, the natural history and the prognostic of CTGA, in the presence and in the absence of associated defects. Beauchesne et al.<sup>4</sup>, when studying the presentation and the patients' prognostic (from 20 to 79 years) with not

operated CTGA, they obtained prevalence of systemic valvar regurgitation of 59%, and 68% of the patients underwent some surgical intervention, including systemic AV valvar replacement in all of them, without precocious mortality.

The group of Graham et al<sup>5</sup> evidenced that, at 45 years of age, 67% of the patients with CTGA presented congestive heart failure,(CHF) an extremely common complication in fourth and fifth decades, and 25% of the patients without associated lesions had that condition. These authors also concluded that the tricuspid insufficiency is strongly associated to the right ventricular dysfunction and congestive heart failure (CHF), but it is still speculative whether this is the cause or just a secondary complication of the other forementioned conditions.

## Case Report

The group of van Son et al.<sup>6</sup>, in study with 40 patients that aimed to evaluate the late results of the AV valvar replacement in patients with CTGA, in the period from 1964 to 1993, concluded that the results of this type of surgery have been improving significantly during the decades, and they suggest that the operation be considered at the first sign of progressive ventricular dysfunction at the serial clinical evaluation and by the echocardiography, to preserve the systemic ventricular function. Such conduct was followed in the subject case, with precocious surgical intervention. The accomplishment of precocious diagnosis is important and nowadays is possible still in the gestational phase, with fetal echography or by post-natal echocardiography in cases that present murmur or signs of ventricular dysfunction, so that the most effective

clinical and/or surgical treatment can be instituted in due time, altering in this way, the natural history of the disease.

### Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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There were no external funding sources for this study.

### Study Association

This study is not associated with any graduation program.

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