Tricuspid Atresia and Double Outlet Left Ventricle: A Rare Association in Adulthood

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Introduction

Tricuspid atresia is the third most common form of cyanotic congenital heart disease with a prevalence of about 10 cases per 1,000 live births.[1] It is defined by the absence of a direct connection between the right atrium and the right ventricle.[2] Double outlet left ventricle (DOLV) is a very rare anomaly occurring in approximately 0.003 to 0.09 per 1,000 live births.[3] In this malformation, the aorta and the pulmonary artery emerge wholly or predominantly from the morphological left ventricle.

These anomalies occurring in isolation rarely achieve adulthood without any surgical intervention. The coexistence of the two diseases, natural evolving in adulthood, is an extremely rare event.

Case Report

This female 25-year-old patient from a rural area in the North of Brazil developed cyanosis at the age of 2 months during breastfeeding and crying. At age 9, she was referred for evaluation at a tertiary center for cardiac catheterization, the result of which was not conclusive due to the complexity of the anomalies. The possibility of surgical treatment was not considered.

The patient developed normal height and weight below the percentile for her age, starting puberty at age 14. Cyanosis has always been severe on exertion. At 17, she presented seizures secondary to brain abscess, clinically treated successfully. This condition in adulthood is characterized by dyspnea on moderate exertion and severe cyanosis on exertion. On physical examination, BMI was 15.4, arterial saturation ranging from 88% to 92%, slight digital clubbing, propulsive wishbones, pectus carinatum, cardiac auscultation with first hyperphonetic heart sound, more intense in the mitral area, slight hyperphonesis A2 and holosystolic murmur +++/6+ at the 2nd and 3rd LICS. ECG showed sinus rhythm, signs of left ventricular hypertrophy with repolarization abnormalities (negative T waves from V2 to V6, DI, AVL and bottom wall), biatrial overload, ÂQRS: +60 and HR: 75 bpm.

Transthoracic echocardiography revealed atrial situs solitus, levocardia and cardiac apex to the right. Due to the severe thoracic deformity, the study was complemented with transesophageal echocardiography, which showed large atrial septal defect, tricuspid atresia, rudimentary right ventricle with unrelated ventricular septal defect and functionally single left ventricle (Figure 1). Aorta was anterior and emerged from the left ventricle (Figure 2). Longitudinal section of the middle esophagus revealed the presence of double outlet left ventricle with vessels separated by muscular band (Figure 3). The pulmonary artery was posterior with bicuspid pulmonary valve and moderate stenosis — maximum transvalvular gradient of 50 mmHg (Figure 4). Nuclear magnetic resonance imaging defined anterior outflow of the aorta, aortic arch to the left with normal emergence of vessels and well-developed pulmonary artery branches (Figure 5).

Discussion

Adult patients with congenital heart disease (CHD) in natural evolution are increasingly rare today due to advances in the diagnosis and surgical treatment of these diseases in children. However, in impoverished populations, we can still find adults with CHD, mostly acyanotic heart diseases with discrete shunt or Eisenmenger Complex.

Tricuspid atresia is a cyanotic heart defect in which the development of pulmonary arterial hypertension occurs early between the fourth and the eighth week of life. Survival of cases not treated surgically will depend on a balanced pulmonary circulation obtained through protective pulmonary stenosis, avoiding elevation of pulmonary vascular resistance, with report of survival until the sixth decade.[4] The main complications described are heart failure, endocarditis, paradoxical embolism, cerebral abscess, chronic hypoxemia, which were the main causes of death.

DOLV, unlike the double outlet right ventricle, is a rare anomaly, with a complex embryological concept and few cases described in the literature. In 1974, Anderson et al.5 described the sixth case in the literature in a 6-year-old child, conducted by angiography, and proposed an embryological hypothesis for the condition, called conal differential resorption.[5] From that work, DOLV was then recognized as a congenital heart defect. Later on, Van Pragh et al.[6] published a review of 109 cases of DOLV, describing 26 variations of the heart disease.

Keywords

Heart Defects, Congenital/surgery; Tricuspid Atresia/surgery; Ventricular Dysfunction Left/surgery; Echocardiography.

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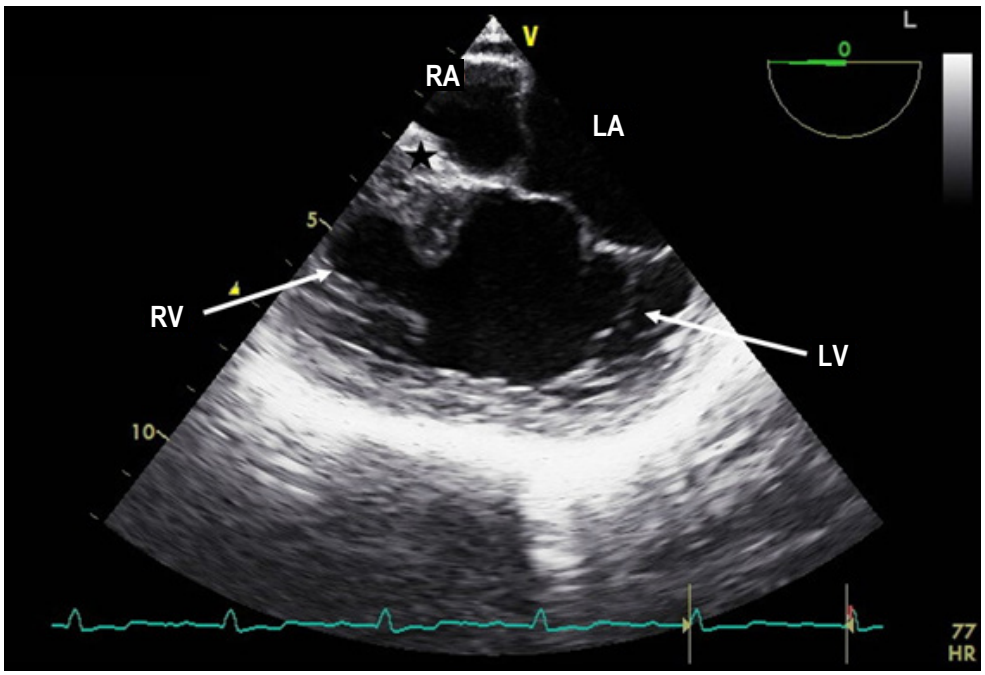

Figure 1 – *TEE - Four-chamber section at 0° revealing tricuspid valve atresia, rudimentary right ventricle and ventricular septal defect not related to large vessels. RA: right artery; LA: left artery; RV: right ventricle; LV: left ventricle.*

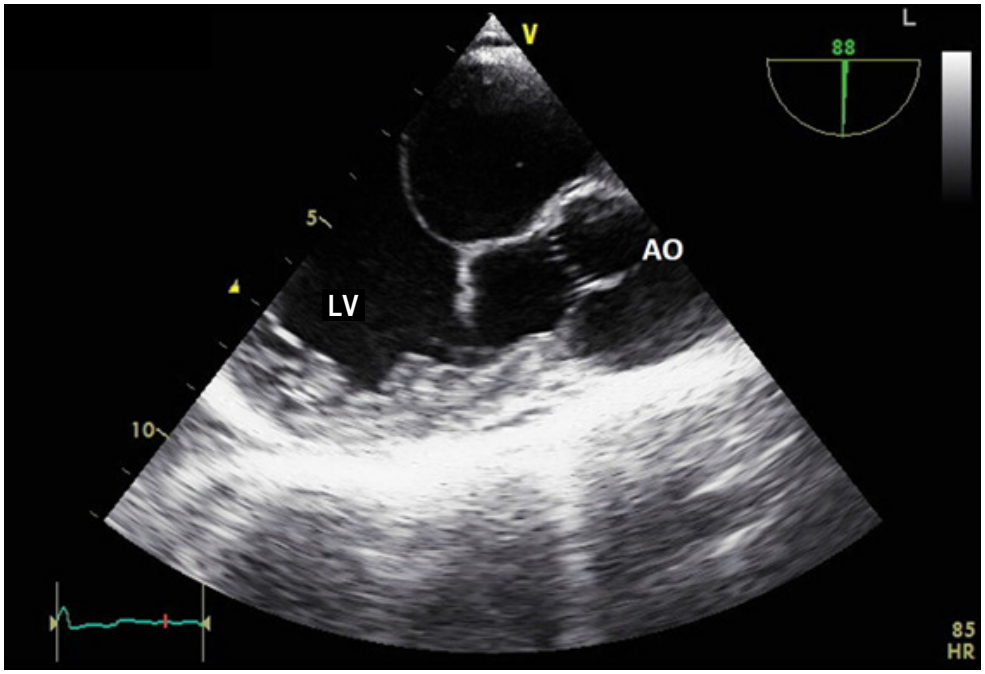

Figure 2 – *TEE - Longitudinal section of the middle esophagus at 90°. Note the origin of the left ventricle aorta. LV: left ventricle; AO: aorta.*

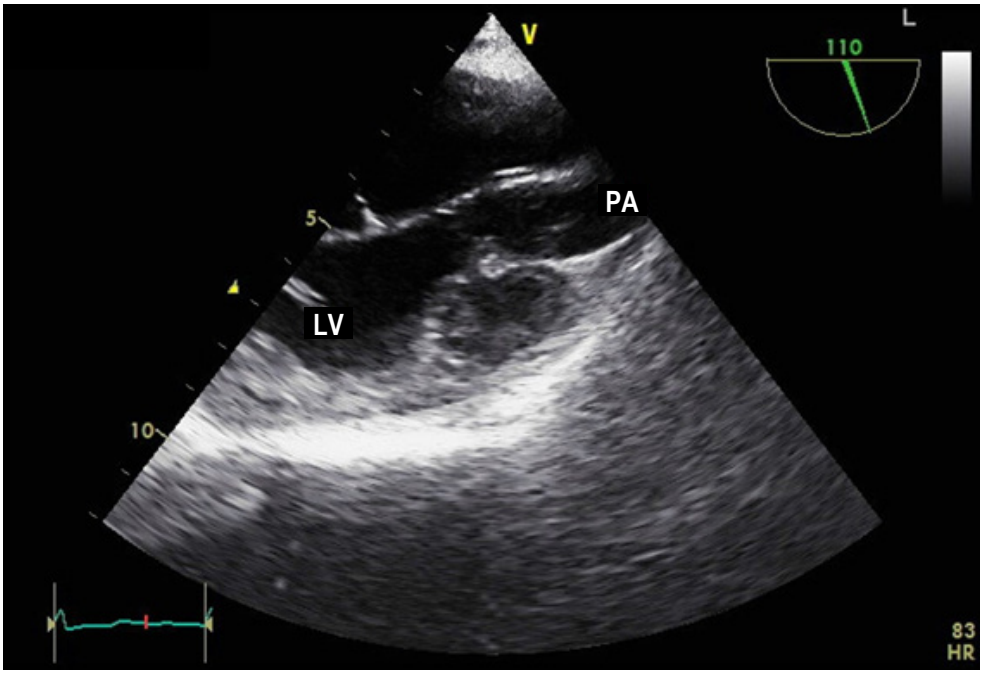

Figure 3 – *TEE - Longitudinal section of the middle esophagus at 110°. Origin of the left ventricular pulmonary artery. LV: left ventricle; PA: pulmonary artery.*

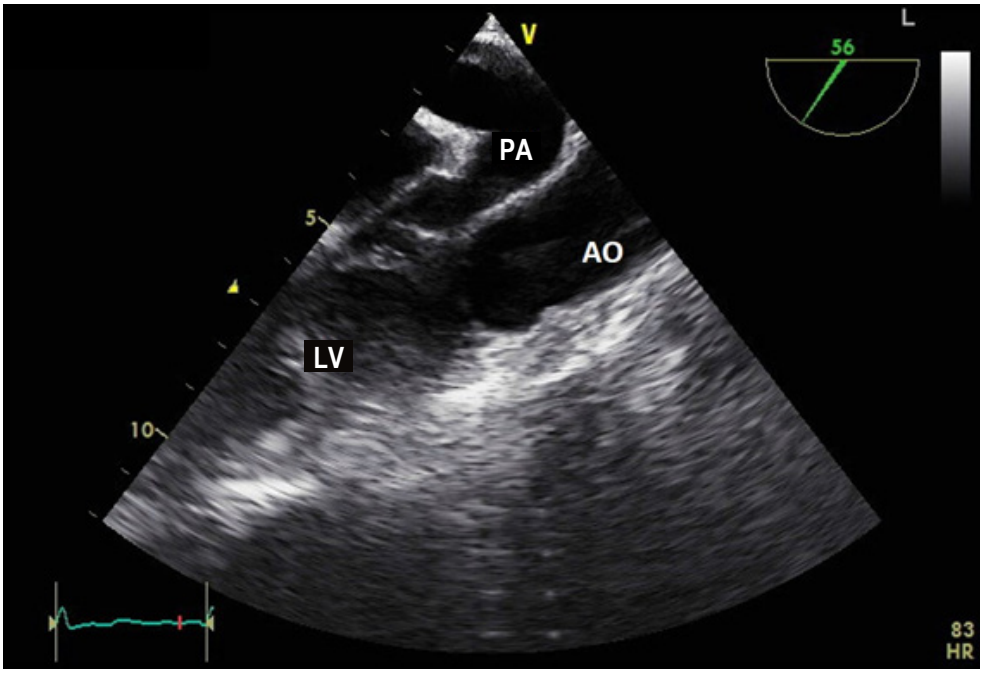

Figure 4 – *TEE - Longitudinal section of the middle esophagus at 56°. Note the origin of the aorta and left ventricular pulmonary artery separated by muscle band. LV: left ventricle; PA: pulmonary artery; AO: aorta.*

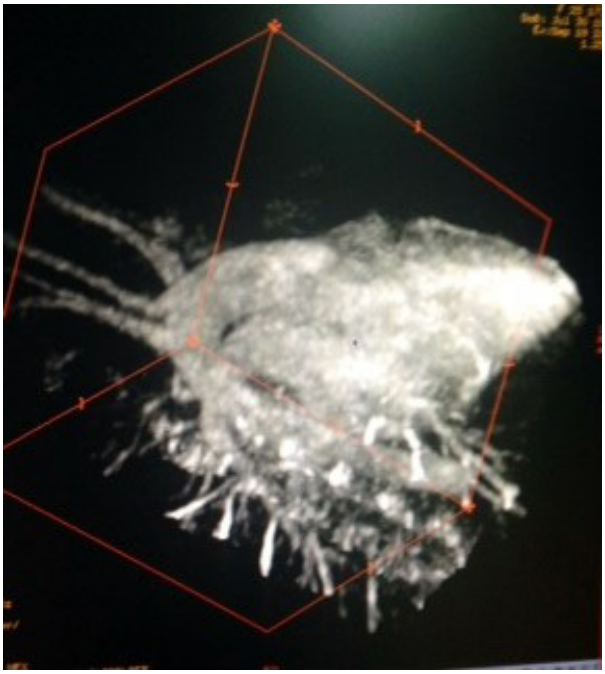

Figure 5 – *NMRI reveals posterior origin of the pulmonary artery and the anterior aorta, both emerging from the left ventricle. Aortic arch with normal vessels.*

Considering the clinical behavior of the malformation, its diagnosis is one of the most challenging ones among many congenital heart diseases.[7-9]

In this case, late diagnosis of this complex CHD was attributed to both the social condition of the patient and to the absence of a previous evaluation with transesophageal echocardiography, considered the most accurate noninvasive test in cases of congenital heart disease in adulthood. Transesophageal echocardiography defined the emergence of the pulmonary artery from the left ventricular cavity, a finding previously unknown and not evident in the catheterization conducted in childhood. Despite the symptoms related to chronic hypoxia and the history of brain abscess, the clinical outcome was favorable, considering the potential of short-term complications of malformations in natural evolution. The presence of pulmonary stenosis naturally regulated pulmonary blood flow, similar to an effective surgical bandage, preventing the development of pulmonary hypertension and allowing acceptable levels of arterial oxygen saturation.

The treatment recommended for a heart disease similar to the one described in children would be a surgical approach, such as the Fontan procedure. However, considering the time of evolution and the potential complication of this approach in adults, the proposed treatment was cardiac transplantation.

Authors' contributions

Research creation and design: Rocha IEG, Pazin IC; Data acquisition: Rocha IEG; Data analysis and interpretation: Rocha IEG, Pazin IC; Manuscript drafting: Rocha IEG, Pazin IC; Critical revision of the manuscript for important intellectual content: Lopes LM; Analysis of all clips and echocardiographic data and selection of photos: Lopes LM.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

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Academic Association

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