

**NEMBORN WITH
CONGENITALLY
CORRECTED
TRANSPOSITION OF
THE GREAT ARTERIES:
CASE REPORT**

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Abstract: Congenitally corrected transposition of the great arteries is a rare congenital malformation in which there is an atrioventricular connection and discordant arterial ventricle. Its prevalence is around 0.5% among congenital heart diseases and is usually accompanied by other defects, as well as associated with the development of complications such as total atrioventricular block and right ventricular dysfunction around 30 years of age, which limit survival. In this case, we describe a patient with congenitally corrected transposition of the great arteries, mild insufficiency of the tricuspid valve, diagnosed with congenital syphilis and treated neonatal jaundice. Despite this, she was discharged with 13 days of life, in good condition.

Keywords: Cardiology, Congenital Anomalies, Congenital Heart Diseases.

INTRODUCTION

Congenitally corrected transposition of the great arteries is a rare congenital malformation with an approximate prevalence of 0.5%, among congenital heart diseases, being more common in males, and may be associated with other congenital defects, such as ventricular septal defect, pulmonary stenosis and other tricuspid valve dysplasias on the left side.^{2,6,8}

In this case, there is a discordant atrioventricular connection accompanied by a discordant ventricular arterial connection, that is, the right atrium is connected to the morphological left ventricle that connects to the pulmonary artery, while on the left side, the left atrium connects to the morphological right ventricle, which is connected to the aorta artery³.

In general, these patients remain asymptomatic until right ventricular (RV) failure due to pressure overload, because in this case the RV functions as the systemic ventricle, the failure manifests as severe heart

failure that may result from other associated injuries.⁷

CASE REPORT

NB of P.C.S.S., female, born by cesarean section due to cardiac malformation (transposition of the great arteries) diagnosed in second semester ultrasound and confirmed with echocardiogram at birth. He was diagnosed with congenital syphilis due to inadequate maternal treatment, so he was treated for 10 days with crystalline penicillin.

The mother reports adequate prenatal care, but with a diagnosis of gestational syphilis, denies a history of family malformations and addiction, reports a previous abortion, without other comorbidities.

At birth, the newborn was asymptomatic and did not require neonatal resuscitation, gestational age of 37 weeks and 2 days by New Ballard, Apgar 81 and 95, normal weight of 2610 grams, blood type A Rh positive, mother being A Rh negative. Physical examination appropriate for age and cardiovascular examination demonstrating normodynamic precordium, palpable ictus in the 3rd intercostal space in the left midclavicular line, absence of valvular shock, regular heart rhythm, two-stroke heart sounds, second heart sound hyperphonestic, without murmurs, clicks or clacks.

An echocardiogram performed on the first day of life showed: situs solitus, normal venoatrial connection; atria with normal dimensions, interatrial septum with patent foramen ovale, measuring 3 mm; discordant atrioventricular connection, two valves; tricuspid valve with mild regurgitation, without typical appearance of Ebstein's anomaly; normal mitral valve; normal-sized ventricles, good ventricular contractility; intact interventricular septum; discordant ventriculoarterial connection, two valves; competent ventricular arterial valves; normal

thoracic aorta; pulmonary artery and branches without alterations; normal pulmonary flow; ductus arteriosus open, small, about to be closed. Conclusion: congenitally corrected transposition of the great vessels of the base; mild tricuspid valve insufficiency and patent ductus arteriosus, consistent with fetal echocardiography performed at 21 weeks and 1 day of gestational age

During the period of hospitalization, the newborn had no complications, except for late neonatal jaundice. Thus, the NB was discharged with 13 days of life, after treatment for congenital syphilis, exclusively breastfed, asymptomatic, without the use of medication, being referred for cardiological and childcare outpatient follow-up.

DISCUSSION

Due to the rarity of congenitally corrected transposition of the great arteries, few cases are diagnosed prenatally. Its diagnosis in this period is possible through morphological ultrasound, associated with fetal echocardiography, being a challenge when, in addition to the transposition, other associated defects are not found.⁵

Its prevalence is 0.02 per thousand live births, which represents approximately 0.5% to 1.4% of congenital heart diseases. Thus, this report is extremely important for the scientific community, due to the scarcity of scientific articles mainly in Portuguese.^{5,9}

With advancing age, some factors contribute to the gradual development of heart failure and symptoms of decompensation, whose causes are associated with right ventricular dysfunction and tricuspid valve insufficiency, arrhythmias and other associated injuries.⁵

In this sense, the subsequent appearance of left atrioventricular valve regurgitation, systemic ventricular dysfunction and complete atrioventricular block means that few survive beyond 50 years of age, even after

being asymptomatic for several years. The manifestation of symptoms, such as fatigue and dyspnea, is mainly due to right ventricular failure resulting from pressure overload.^{4,7}

In asymptomatic adults, the main diagnostic method is transthoracic echocardiography, cardiac catheterization or magnetic resonance imaging is rarely used. However, the diagnosis can also be made by electrocardiogram associated with chest X-ray. On the electrocardiogram, a deviation in the QRS axis can be found, with atypical septal activation, demonstrated by the absence of Q waves in the left precordial leads. While the chest X-ray shows dextrocardia or an abnormal cardiac silhouette resulting from the L-position of the aorta⁹.

When necessary, magnetic resonance imaging is useful to assess left-sided flow obstructions that may occur without defects in the ventricular septum, and may occur in the pulmonary valve or before it. The indication of cardiac catheterization can be used to assess pulmonary vascular resistance in older patients without left-sided flow obstruction.¹

Early diagnosis is fundamental, in view of all the possible consequences in adult life resulting from late identification.⁵

The treatment of this condition depends on the clinical presentation and associated defects, ranging from clinical treatment with drugs (angiotensin-converting enzyme inhibitors, diuretics, digitalis), to palliative (physiological) or definitive (anatomical) surgical treatment, with the arterial exchange⁷.

Surgical intervention is reserved for symptomatic patients and for those asymptomatic with declining (systemic) right ventricular function and tricuspid regurgitation. In asymptomatic patients who do not fit this definition, the surgical approach is controversial considering that they have a favorable prognosis and surgical intervention has many risks and long-term consequences.¹

Heart transplantation may be the only option in patients who have a contraindication for heart repair surgery, older age, and those with poor previous surgical results.¹

Finally, the patient reported is under outpatient follow-up with Cardiology, performing serial echocardiograms in order to monitor systemic ventricular function.

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