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CASE REPORT

## Atresia of tricuspid valve and hypoplasia of the right ventricle

Graziela Cruz e Silva<sup>1</sup>, Jussara Silva Lima<sup>2</sup>, Leda Maria Prado Palis<sup>1</sup>, Larissa de Oliveira Braga<sup>1</sup>

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### Abstract

Tricuspid atresia is a cyanotic congenital heart disease, due to agenesis (absence of right atrioventricular connection) or imperfection of the tricuspid valve, resulting in no direct communication between the right atrium and ventricle. Corresponds to 2.7% of all congenital heart diseases. In this case report we describe the handling of an infant with tricuspid atresia and hypoplastic right ventricle, with extensive interatrial (IAC) and interventricular communication (IVC), as well as a brief review of the literature on pathology. The patient, male, was admitted to our Service with one month and twenty-eight days of life. He presented sudden respiratory failure after choking and apnea, being intubated in the city of origin and transferred to our Service. Upon admission, systolic murmur, increased cardiac area on chest radiography, and signs of pulmonary hyperflow were observed. Echocardiogram showed 9 mm CIA, tricuspid valve atresia with right ventricular hypoplasia, significant pulmonary hyperflow, 8.5 mm VSD, absence of ductus arteriosus, dilated left atria and ventricles. Initiated clinical treatment of heart disease and programming of the surgical procedure. Tricuspid atresia consists of complete absence of right atrioventricular connection. CIA and IVC or persistent ductus arteriosus, are associated with obligatory lesions to maintain pulmonary flow. The condition predominates in males and is associated with right ventricular hypoplasia in 100% of cases. Surgical treatment is mandatory for the disease. The patient reported received clinical treatment, including diuretics, vasodilators and digitalis; In addition, surgery was scheduled to correct the malformation.

<sup>1</sup> Resident Physician in Pediatrics.

<sup>2</sup> Pediatrician: Professor of the Residency in Pediatrics of the University Clinics Hospital of the Federal University of Triângulo Mineiro.

### Correspondence to:

Graziela Cruz e Silva.

Hospital De Clínicas, Universidade Federal do Triângulo Mineiro - UFTM. Rua João Alfredo, nº 560, Apto 602, Bloco A, Nossa Senhora da Abadia, Uberaba - MG. Brazil. Zip Code: 38025300.

## INTRODUCTION

Tricuspid atresia is a form of congenital heart disease with cyanosis, which arises from either the agenesis (absence of right atrioventricular connection) or the imperforation of the tricuspid valve. Consequently, there is no direct communication between the right atrium and the right ventricle. The survival of children with tricuspid atresia depends on the presence of an interatrial septum defect that allows the passage of blood from the right atrium to the left atrium. It is a rare malformation, corresponding to 2.7% of all cases of congenital heart disease. The etiology is unknown.<sup>1</sup> Our study reports the experience with an infant referred to our service with a late echocardiographic diagnosis of tricuspid atresia with right ventricular hypoplasia.

## OBJECTIVES

The objective of this case report is to describe the management of an infant with tricuspid atresia and right ventricular hypoplasia in addition to broad interatrial (IAC) and interventricular communication (IVC) as well as to briefly review the literature with respect to this condition.

## METHODS

This report was based on our direct following of the case, including assessment of the medical history and daily physical examinations, as well as the collection and analysis of data from the medical record and literature review.

## CASE REPORT

A male patient aged 1 month and 28 days was admitted to our service, presenting with sudden respiratory insufficiency and apnea after choking. He was intubated in his hometown and transferred to our service. Upon admission, a panfocal systolic murmur could be heard on auscultation, a thoracic radiograph showed an enlarged cardiac silhouette, and there were signs of pulmonary hyperflow. Echocardiography was performed, evidencing a 9mm IAC, tricuspid valve atresia with right ventricular hypoplasia, significant pulmonary hyperflow, an 8.5mm IVC with a resulting 30mmHg gradient, absence of the arterial channel, and dilated left atrium and ventricle. The patient was transferred to intensive care unit intensive care

unit (ICU), and the clinical treatment of the heart condition was initiated along with the planning of the surgical procedure.

## DISCUSSION

Tricuspid atresia involves a complete absence of the right atrioventricular connection. IAC and IVC or persistence of the arterial channel are associated lesions that are mandatory to preserve pulmonary flow.<sup>2</sup> It has been demonstrated that the condition is more frequent in males and is associated with right ventricular hypoplasia in 100% of cases.<sup>3</sup> Our patient also displayed some characteristic features of the malformation, such as broad IVC with no pulmonary stenosis. This caused pulmonary hyperflow that produced heart failure symptoms. The approach covers several aspects, but surgical treatment is mandatory for this condition.<sup>2</sup> Mortality increases with increased age at surgery because prolonged volume overload in the functioning ventricle and persistence of cyanosis contribute to a gradual deterioration of the myocardium.<sup>4</sup> The patient described in this report was treated clinically, namely with the administration of diuretics, vasodilators, and digitalis; moreover, a surgical intervention was scheduled, aiming to correct the malformation.

## CONCLUSION

Our experience shows that the clinical and surgical management of tricuspid atresia may achieve a satisfactory outcome in our setting, provided the pediatrician recognizes the cyanotic congenital heart disease at an early stage and the patient is timely transferred to a reference pediatric cardiology center.

## REFERENCES

1. Mattos SS, Rodrigues JV, Severi R, Nunes M, Cunha CEG, Melo VB, et al. Manuseio da atresia tricúspide em neonatos. Relato de três casos e revisão da literatura. *J Pediatr.* 1994;70(1):33-8.
2. Campos Júnior D, Burns DAR, Lopez FA. *Tratado de Pediatria.* 3ª ed. Barueri: Manole; 2014.
3. Blasubramanian S, Tacy T. Tricuspid valve (TV) atresia. UpToDate [acesso 2017 Nov 27]. Disponível em: <https://www.uptodate.com/contents/tricuspid-valve-tv-atresia>
4. Mastalir ET, Kalil RAK, Horowitz ESK, Wender O, Sant'Anna JR, Prates PR, et al. Desfechos Clínicos Tardios da Cirurgia de Fontan em Pacientes com Atresia Tricúspide. *Arq Bras Cardiol.* 2002;79(1):51-5.