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Case Report



# Cor triatriatum dexter in adulthood

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#### ABSTRACT

*Cor triatriatum dexter* is a very rare malformation in which the right atrium is divided into two chambers by a membrane. The case of a 24-year-old woman with a history of heart failure due to postpartum cardiomyopathy is reported. She was admitted to the emergency department for an acute exacerbation of her underlying disease. An echocardiogram was performed and *cor triatriatum dexter* was diagnosed. The patient was referred to the cardiovascular surgery department for surgical resection. *Key words: Cor triatriatum,* congenital heart disease / diagnosis, heart failure

## Cor triatriatum dexter en la edad adulta

#### RESUMEN

El *cor triatriatum dexter* es una malformación muy rara, en el cual la aurícula derecha está dividida en dos cámaras por un tabique. Se presenta el caso de una joven de 24 años de edad con antecedentes de insuficiencia cardíaca por miocardiopatía puerperal, que acude al servicio de urgencias por descompensación de su enfermedad de base. Se solicita ecocardiograma donde se diagnostica el *cor triatriatum dexter*. Se envía la paciente al servicio de cirugía cardiovascular para resección quirúrgica.

Palabras clave: Cor triatriatum, cardiopatía congénita / diagnóstico, insuficiencia cardíaca

### INTRODUCTION

*Cor triatriatum dexter* (CTD) is a very rare malformation, with an incidence of approximately 0.025% of all congenital heart diseases, and in which the right atrium is divided into two chambers by a membrane. This abnormality is usually attributed to the persistence of the right valve of the sinus venosus and is associated with other malformations of the right heart<sup>1-6</sup>. The case of a patient admitted with symptoms of right heart failure is reported; she under-

went a transthoracic echocardiography that showed the presence of CTD.

## **CASE REPORT**

Female patient, 24 years of age, with a history of postpartum complicated by congestive heart failure. She was discharged from the maternity ward with a diagnosis of postpartum cardiomyopathy. Due to continuing symptoms, despite treatment, she decided to go to the emergency room and complained about having swollen legs and abdomen.

Physical examination revealed the following signs: pale mucous membranes, tachypnea, blood pressure of 100/70 mmHg, heart rate of 110 beats per minute and respiratory rate of 18 breaths per minute, jugular venous distention, rhythmic heart sounds, holosystolic murmurs grade III/VI in mitral and tricuspid foci, no pulmonary rales, painful hepatomegaly about 5 cm below the costal margin, moderate ascites and lower limb edema up to both knees. A chest radiograph was performed and showed biventricular cardiomegaly with increased pulmonary blood flow and enlarged pulmonary artery (Figure 1). These clinical symptoms



Figure 1. PA chest radiograph.

and the x-ray findings led to requesting a transthoracic echocardiography.

This test showed a small left ventricle with mild global hypokinesia and slightly decreased systolic function (ejection fraction 0.43), moderate mitral regurgitation and mild pulmonary regurgitation. It was seen a dilated right atrium, with the presence of a membrane dividing it into two chambers: a proximal one in which the *venae cavae* flow and a distal chamber, which corresponded with the tricuspid valve (Figure 2). The inferior vena cava was dilated with a reduction of its inspiratory collapse. With these findings, it was concluded that the patient had a CTD that affected the flow and it was decided to refer her to the cardiovascular surgery department where she is waiting for surgery.

## COMMENT

In the fourth week of embryogenesis, the common atrium is separated from the venous sinus by a structure called venous sinus valve. During cardiogenesis, the left veil of this valve is incorporated into the atrial septum and becomes part of the *septum secundum*, and the right veil atrophies and disappears, leaving two remnants: the Eustachian valve, adjacent to the inferior vena cava, and Thebesian valve at the orifice of the coronary sinus. The persistence of this right veil causes the malformation known as CTD. This anomaly is usually associated with other malformations of the right heart, such as hypoplasia or tricuspid atresia, or pulmonary atresia<sup>2</sup>.

The CTD can be diagnosed at any age and may be an incidental finding during routine echocardiographic examination. It usually produces no symptoms, and if they appear, they are directly related to the venous blood flow obstruction caused by the membrane, which in most cases is fenestrated, hence the absence of symptoms. Therefore, some authors do not accept the term of CTD if there is no venous flow obstruction, preferring the term Chiari network<sup>3,4</sup>.

The echocardiographic presence of this malformation in this patient explains the existence of signs and symptoms of right heart failure due to venous flow obstruction, and justifies the use of the term *cor triatriatum dexter*. In symptomatic patients, the treatment of choice is surgical resection, although percutaneous rupture has also been described as an alternative method<sup>5,6</sup>.



**Figure 2.** Transthoracic echocardiography. **A.** Apical four-chamber view. **B.** Subcostal view. Shows the membrane that divides the right atrium into two cavities (arrowhead). AD: right atrium, AE: left atrium, VD: right ventricle, VE: left ventricle

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