

# Clinical Follow-up of a Pregnant Woman with Cor Triatriatum

Rogério Tasca, Manuela Gonçalves Tasca, Paulo Artur Amorim, Isabel Cristina do Nascimento, Orlando Carlos Glória Veloso, Carlos Scherr

Ecodoppler Exames Cardiológicos, Hospital Pan Americano - Rio de Janeiro, RJ - Brazil

Cor triatriatum (CT) is a rare congenital cardiac anomaly that usually becomes symptomatic in the first years of life. If the condition is not associated with other cardiac defects, and depending on the degree of communication between the upper chamber and left atrium (LA), patients may reach adulthood. We report a case of an asymptomatic, adult, female patient with CT diagnosed by transthoracic echocardiogram (TTE) and followed-up during pregnancy.

## Introduction

First described in 1868<sup>1</sup>, cor triatriatum is a congenital malformation secondary to a failure of the common pulmonary vein resorption during cardiogenesis. This genetic error leads to the persistence of a diaphragm-like membrane dividing the LA into a posterosuperior chamber receiving the pulmonary veins and an anteroinferior chamber connected to the left atrial appendage (LAA) and draining into the left ventricle via the mitral valve. Anatomical variations and associations with other cardiac malformations, such as atrial septal defect (ASD) and anomalous pulmonary venous return<sup>2</sup>, are common. Communication between both left atrial chambers may be made through one or more orifices and may be small, large or absent, depending on the membrane dividing them. In the absence of other associated defects, the size of the opening(s) will determine the degree of obstruction to pulmonary venous return and, thus, the patients' clinical outcome. Elevations in pulmonary venous pressure and pulmonary vascular resistance may cause pulmonary artery hypertension (PAH); the more restrictive the communication is between both cavities, the earlier this may happen. Right cor triatriatum (dexter), resulting from the persistence of the right sinus venosus valve, is a rare heart disease that may lead to right atrium obstruction<sup>3</sup>.

## Case Report

A 34-year-old white woman, four weeks pregnant, was referred by her obstetrician for cardiological evaluation due to past history of congenital heart disease. At 18, she had seen a cardiologist for palpitations, and a transthoracic echocardiography (TTE) diagnosed a congenital heart defect.

## Key words

Cor triatriatum; heart defects, congenital; adult; pregnancy.

She did not know exactly what type of defect she had, nor did she have her previous supplementary examinations. Back then no drugs were prescribed and, despite being instructed to visit the cardiologist periodically, she did not. The patient had no cardiovascular complaint, including during physical exertion. Physical examination revealed regular heart rate and rhythm with normal S1 and S2 and no murmurs. Her heart rate was 65 beats per minute, and her blood pressure was 110 x 60 mm Hg.

Both the electrocardiogram (Figure 1) and chest X-ray (Figure 2) were within normal parameters. TTE (Figure 3) revealed a membrane dividing the LA into an upper chamber, accepting the pulmonary veins, and a lower chamber communicating to the LLA and mitral valve. The interatrial septum (IAS) was intact. Chamber dimensions and biventricular contractility were normal. Color Doppler imaging demonstrated two flows with increased velocity from the upper to the lower chamber, consistent with two orifices with different diameters through which the pulmonary veins are drained (figure 4). On pulsed Doppler, a peak pressure gradient of 4.8 mm Hg (mean of 2.7 mm Hg) was found between the two chambers. Pulmonary artery systolic pressure was estimated in 25 mm Hg by a mild tricuspid regurgitation. Based on these echocardiographic findings, the diagnosis was made of mildly obstructive CT without hemodynamic repercussion. The patient went back to the referring obstetrician with instructions for prophylaxis against infective endocarditis (IE) should any contaminated procedure be performed.

Transthoracic echocardiograms were performed at the 16th and 27th gestational weeks. In the latter, a slight increase in pressure gradient was observed between the upper and lower left atrial chamber, as compared with the first examination (maximum of 7.8 mm Hg, mean of 3.4 mm Hg). The pregnancy course was uneventful. Although vaginal delivery was not contraindicated, this patient underwent a cesarean section at 36 weeks of gestation, upon the obstetrician's decision. At the last cardiological and echocardiographic examinations, performed two years later, the patient's clinical condition was unchanged, she was asymptomatic and the echocardiographic findings were similar to those of the initial examination.

## Discussion

Despite being a rare condition, if it is not accompanied by other heart malformations cor triatriatum can be easily corrected surgically<sup>4</sup>, but early diagnosis of symptomatic cases is critical. Although rare in adults<sup>5</sup>, CT was described in septuagenarian and octogenarian patients<sup>5,6</sup>. In both, similar to the case herein presented, no other congenital

Mailing address: Rogério Tasca •  
Rua Souza Lima, 325/801 - 22081-010 - Rio de Janeiro, RJ - Brazil  
E-mail: tasca@cardiol.br  
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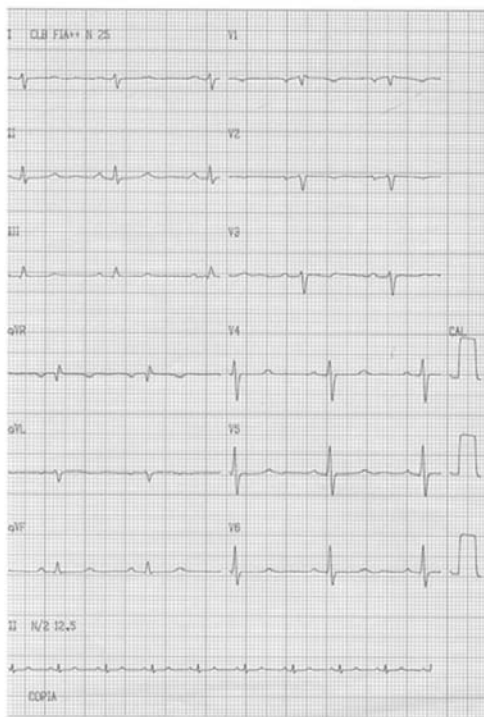


Fig. 1 - Electrocardiogram.

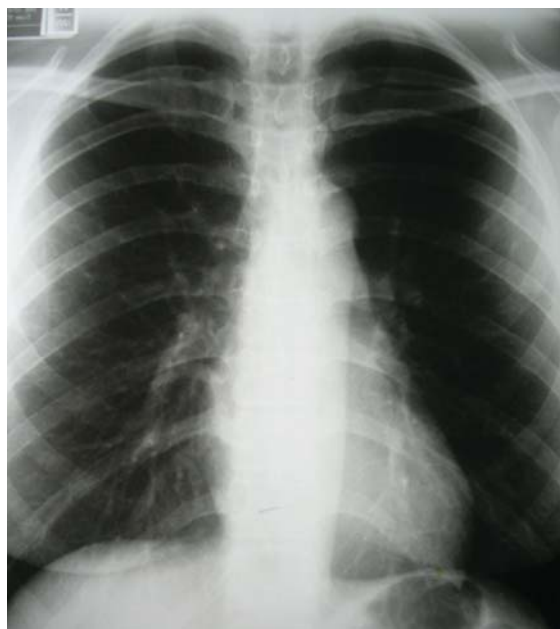


Fig. 2 - Plain chest X-ray (anterior view).

structural defects existed, and the membrane caused little, if any, restriction to the pulmonary veins flow. Non-invasive diagnosis can be made by TTE<sup>7</sup>, and color flow Doppler makes it possible to visualize the opening(s) between the

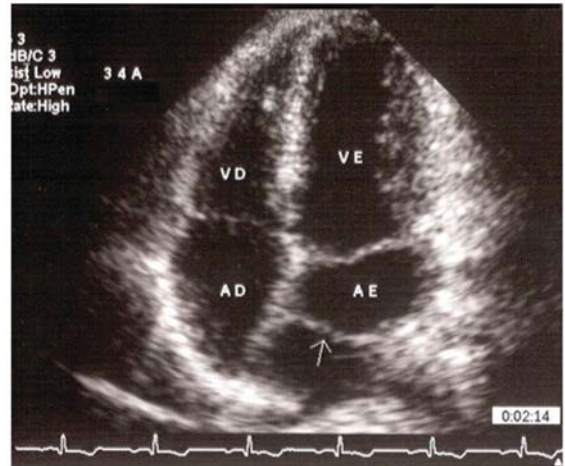


Fig. 3 - Apical four-chamber view revealing a membrane (arrow) dividing the LA into a lower chamber communicating with the mitral valve and an upper chamber receiving the four pulmonary veins. This view shows the two lower pulmonary veins.

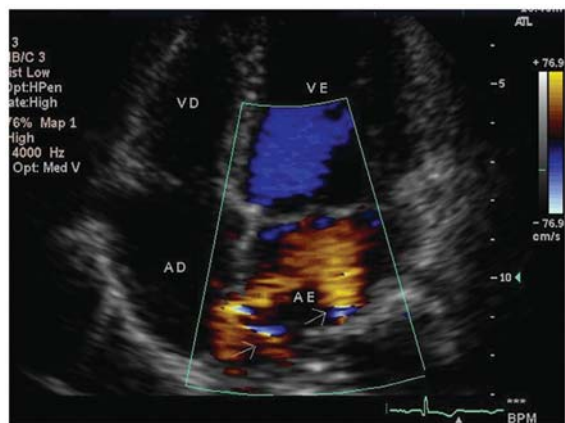


Fig. 4 - Apical four-chamber view obtained with color Doppler flow showing two flows (arrows) through the orifices forming the communication between two chambers.

two chambers, especially in the case of gradient between them, because the turbulence caused by increased blood flow velocity is easily detected. Using pulsed Doppler, this pressure gradient can be estimated. In the case of this patient, the two flows with different velocities allowed identifying the presence of two orifices, the more anterior being smaller, since it showed a higher-velocity flow. There is no report in the literature of pressure gradient variations through the CT membrane being monitored by Doppler echocardiography during pregnancy. In this patient, TTE examinations have demonstrated a rise in pressure gradient, secondary to an increase in blood volume, which usually occurs between weeks 24 and 28 of gestation.

Transesophageal echocardiogram (TEE) is indicated when more accurate anatomical details are needed to determine optimal surgical repair or assess the presence of associated

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lesions. As our patient was asymptomatic and there was neither evidence of PAH nor structural defects, we chose not to perform TEE. Even though few reports exist so far on the subject, real-time three-dimensional echocardiography, recently introduced commercially, will allow greater accuracy regarding the anatomical aspects of CT<sup>8</sup>. Among the differential diagnoses to be considered is congenital supralvular mitral stenosis<sup>9</sup>, which may produce signs and symptoms similar to those of obstructive CT during childhood. Uncharacteristically, TTE findings of this lesion showed that the membrane lies more inferior, very close to the mitral leaflets, rendering the LAA proximal. Although we found no specific reports regarding the

need of prophylaxis for infective endocarditis in adult patients with CT, we recommend that antibiotic therapy be used in case of contaminated procedures, because of the turbulent flow through the membrane within the LA. Female patients with non-obstructive CT have no contraindication for vaginal delivery<sup>10</sup>. In the present case, the cesarean section was the obstetrician's choice, concurred to by the patient, rather than a contraindication.

### Potential Conflict of Interest

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

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