

Cor Triatriatum Sinistrum in Symptomatic Adult: Case Report

Francilayne Moretto dos Santos, Laura Gomes Flores, Felipe Rodrigues Dias, Leandro Shigueru Ikuta Ueda, Eduardo Frederico Philippi

Centro Universitário Faculdade Assis Gurgacz, Toledo; Hospital São Lucas, Cascavel, PR; Unitom - Unidade de Diagnóstico por Imagem, Cascavel, PR; Intervent Hemodinâmica, Cascavel, PR – Brazil

Introduction

Cor triatriatum is a rare congenital malformation, accounting for 0.1 to 0.4% of all congenital heart defects. The pathophysiology results from the failure to reabsorb the common pulmonary vein during the embryonic formation of the heart. Because of this, there is persistence of a fibrotic membrane, dividing the left atrium (LA) into a posterosuperior cavity that receives the pulmonary veins and an anteroinferior cavity attached to the left atrial appendage (LAA), communicating to the left ventricle (LV) through the mitral valve.¹⁻⁴ Atrial septal defect (ASD) and pulmonary venous return anomalies are malformations that may be associated.⁵

Case Report

V.G.S., female, 46 years old, cook. The patient reported dyspnea on major exertion and long-standing nocturnal paroxysmal dyspnea. The patient denied chest discomfort, palpitations and syncope. As comorbidities, the individual reported glaucoma, a cervical disc hernia that took her away from work one year prior, and depression. Smoker for 39 years/pack of cigarettes.

Cardiological physical examination in regular rhythm, two-stroke, no murmur and no fixed split second heart sound; pulmonary examination with no abnormalities. Electrocardiogram (ECG) was normal. Echocardiogram (ECHO) revealed LA with preserved diameter; presence of ostium secundum ASD of approximately 8 mm, without hemodynamic repercussion; cor triatriatum membrane; mild tricuspid regurgitation estimating systolic pressure in the pulmonary artery of 26 mmHg. LV with preserved wall dimensions and thickness; and preserved left ventricular systolic function (Figures 1 and 2). Spirometry was normal. The 5-minute BRUCE exercise test showed no signs of exertional ischemia, reaching the maximal heart rate at the peak of exertion, with 5.13 METS, and interrupted by fatigue. New York Heart Association (NYHA) functional class II.

As the echocardiogram showed no signs of ASD shunt with hemodynamic repercussion and disagreement with the

symptoms and low cardiorespiratory capacity, we chose to perform cardiac catheterization, considering the possibility of a false-negative result in the exercise test.

At catheterization, unobstructed coronary arteries were observed, as well as contrast recirculation to LA and right atrium through ASD, and QP/QS calculation of 1.81. Pulmonary systolic pressure (PSP) was 34 mmHg, pulmonary diastolic pressure (PDP) was 17 mmHg and the mean pulmonary artery pressure was 21 mmHg (Figure 3).

Cardiac magnetic resonance imaging (MRI) in a high field instrument (3 Tesla) with paramagnetic contrast injection showed cardiac chambers of preserved dimensions; LA with two proximal and distal chambers, with no obstructive signs between them, with greater opening measurement of 2.5 cm. ASD with predominance of LA flow to the right, with no significant hemodynamic repercussion (Qp/Qs: 1.3). Preserved biventricular systolic function and absence of myocardial fibrosis (Figure 4). The set of findings was compatible with cor triatriatum sinistrum, associated with the ASD.

Because we did not note any overloads of right chambers and hemodynamic repercussions, neither on ECHO nor on ASD MRI, we chose to advise the patient to stop smoking and follow her up, conducting an echocardiography every 6 months.

Discussion

Cor triatriatum sinistrum is a rare congenital malformation, in which a membrane, usually a fibromuscular septum, divides the LA into two different cavities. The proximal or superior chamber drains blood from the common pulmonary vein while the distal chamber, which is considered the true atrium, is in contact with the atrioventricular valve and contains the septum and the atrial appendage.⁵

The location of the membrane is essential to differentiate between cor triatriatum and the supralvalvar mitral ring: the cor triatriatum membrane is located above the LAA, associating it with the anteroinferior chamber, whereas, in the supralvalvar mitral ring, the appendix is associated with the posterosuperior chamber.⁶

According to the number and size of the fibromuscular septum orifices, according to Löffler, the disease is classified into three groups: group 1 is composed of a membrane without aperture, in which the accessory chamber drains the blood into the right chambers; group 2 presents small fenestrations and in small quantity, resulting in a high-pressure gradient; group 3 involves a septum with several fenestrations, developing no or slight obstruction.⁶

The last group, of the patient of this report, rarely presents symptoms, and is usually diagnosed in adult life. On the other hand, groups 1 and 2 are associated with the

Keywords

Heart Defects Congenital; Cor Triatriatum/physiopathology; Adult; Echocardiography/methods; Cardiac Catheterization; Heart Septal Defects, Atrial; Diagnostic Imaging.

Mailing Address: Francilayne Moretto dos Santos •

Rua Albei Antônio Richard, N° 124, Cancelli. Postal Code 85811-290, Cascavel, PR – Brazil

Email: franmoretto92@gmail.com

Manuscript received May 21, 2018; revised June 10, 2018; accepted July 27, 2018.

DOI: 10.5935/2318-8219.20180044

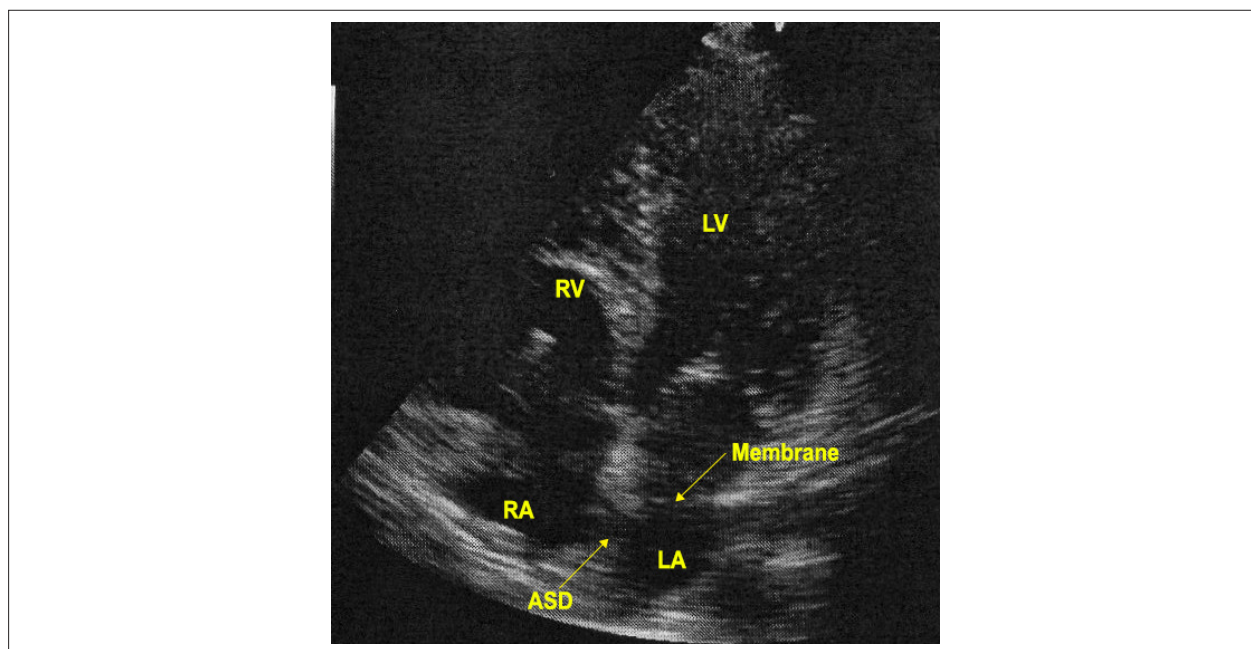


Figure 1 – Transesophageal echocardiography in 4-chamber apical section showing the cor triatriatum membrane and atrial septal defect above it.

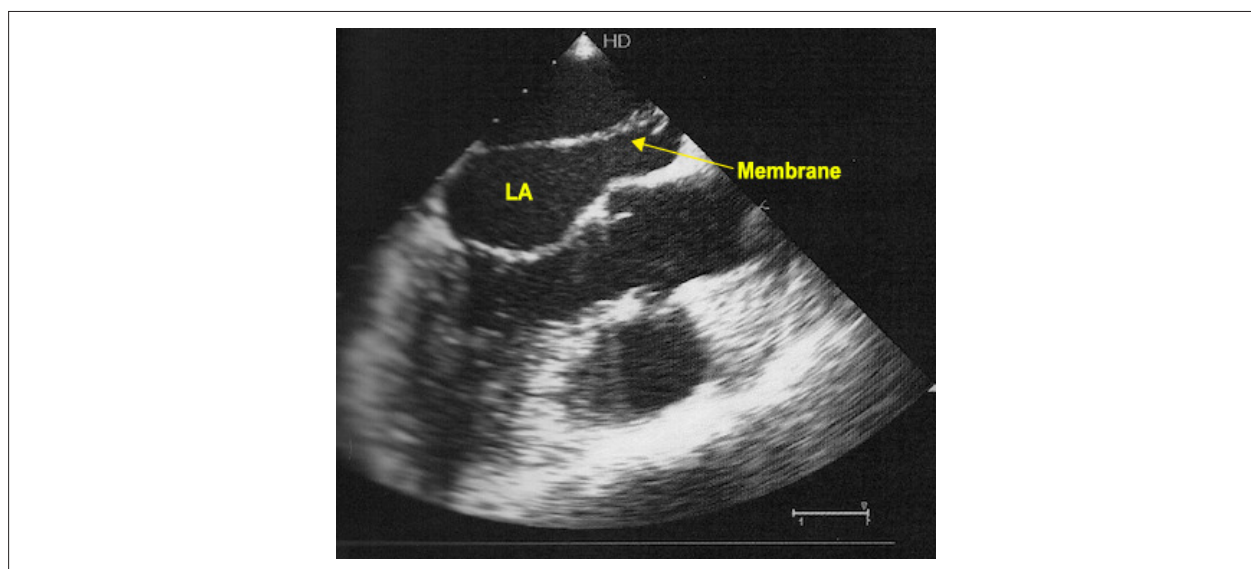


Figure 2 – Transesophageal echocardiography showing cor triatriatum membrane.

presence of significant symptoms in childhood and higher mortality at earlier ages.⁵

In more than 80% of the cases, this disease is associated with other congenital heart diseases, such as ostium secundum ASD, persistent left superior vena cava, aortic regurgitation with dissecting aneurysm and anomalous pulmonary vein connection.^{5,6}

Newborns and infants usually manifest heart diseases with varying degrees of dyspnea, which basically results from the intra-atrial pressure gradient, with consequent increase

in the proximal left atrial chamber pressure and pulmonary congestion. Adults, who in the absolute majority of the cases are included in Loeffler's group 3, usually present symptoms only when the intraarterial membrane orifices develop fibrosis or calcification. In this case, the most common manifestations are dyspnea, orthopnea and hemoptysis.⁵

Physical examination can reveal signs of this anomaly by means of pulmonary auscultation as, classically, in the presence of pulmonary hypertension, a diastolic murmur with high intensity P2 is auscultated. In the ASD case, we can observe fixed split second heart sound.^{5,6}

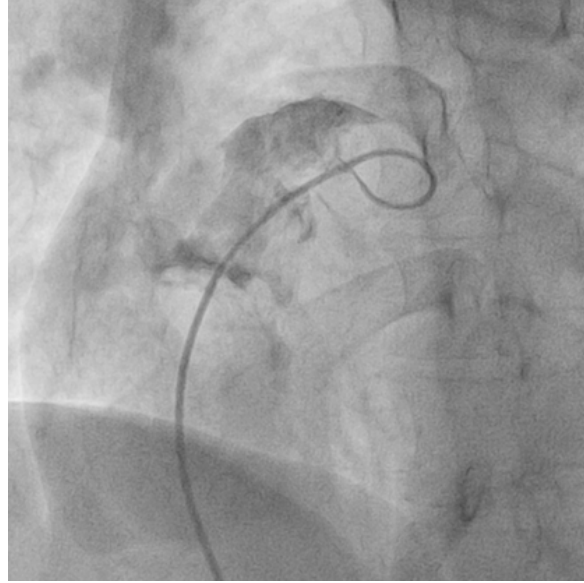


Figure 3 – Contrast injection near the atrial septal defect.

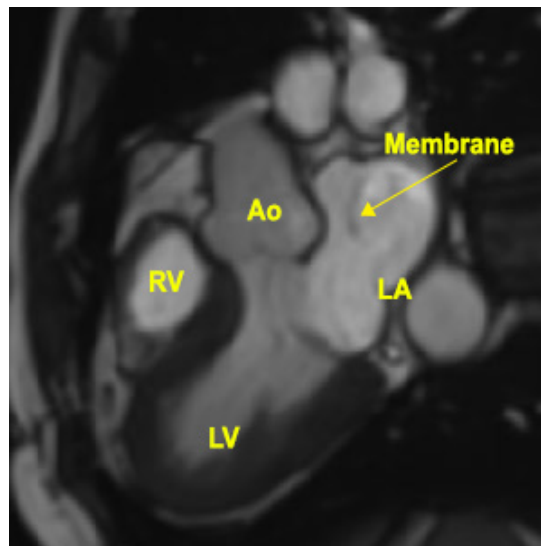


Figure 4 – Cardiac resonance CINE imaging, 3-chamber view, showing left atrial membrane subdividing into anterior and posterior chambers.

The main diagnostic method is echocardiogram, due to its convenience and the ability to delineate the morphology of the intra-atrial septum, in addition to the presence of associated cardiac lesions. However, catheterization, computed tomography and cardiac MRI are also capable of diagnosing this anomaly.⁵⁻⁷

The electrocardiogram is normal in most cases, but in the presence of pulmonary hypertension, signs of right ventricular overload and axis deviation to the right can be found. Although cardiac catheterization is rarely used at present for diagnosing cor triatriatum sinistrum, it shows a normal

left atrioventricular gradient. To analyze the transmembrane gradient and the obstruction degree, echo Doppler is used and, to measure the orifice, a three-dimensional transthoracic echocardiography can be performed. In cases where image quality is limited, transesophageal echocardiography should be performed for diagnostic confirmation. MRI and computed tomography are also useful in the diagnosis of this anomaly, since they reveal the presence of accessory membrane. Currently, MRI in all surgical patients is recommended because of its ability to portray membrane fenestrations more accurately, and for the presence of some associated flow turbulence.^(5,7,8)

Management of patients with cor triatriatum depends on the degree of obstruction between the LA chambers. Definitive treatment is surgery, which is reserved for patients with symptoms secondary to severe intra-atrial obstruction. Asymptomatic patients require strict clinical follow-up to ensure an early drug or surgical approach when severe symptoms arise, since a more severe symptomatology makes the prognosis less favorable after repair.^(5,6,9)

Authors' contributions

Research creation and design: Santos FM, Flores LG; Data acquisition: Santos FM, Ueda LSI, Philippi EF; Data analysis and interpretation: Santos FM, Flores LG, Ueda LSI, Philippi EF; Statistical analysis: Santos FM, Flores LG;

Manuscript writing: Santos FM, Flores LG, Dias FR; Critical revision of the manuscript as for important intellectual content: Santos FM, Flores LG, Ueda LSI, Philippi EF.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

Sources of Funding

This study had no external funding sources.

Academic Association

This study is not associated with any graduate programs.

References

1. Nassar PN, Hamdan RH. Cor Triatriatum Sinistrum: classification and imaging modalities. *Eur J Cardiovasc Med.* 2011;1(3):84-7.
2. Raheja H, Namana V, Moskovits N, Hollander G, Shani J. Cor Triatriatum Sinistrum. *Arq Bras Cardiol.* 2018;110(1):101.
3. Junior RA, Cunha AJ, Yared F, Masselli DB, Machado RL, Silva CE, et al. Cor Triatriatum Restritivo em Paciente Adulto Assintomático. *Arq Bras Card Imagem Cardiovasc.* 2014;27(1):32-4.
4. Lambert J, Oechslin E, Tsang W, Osten MD, Thavendiranathan P. Cor Triatriatum Sinister with secundum atrial septal defect. *Cardiovasc Imaging Case Rep.* 2017;1(4):141-6.
5. Nassar PN, Hamdan RH. Cor Triatriatum Sinistrum: classification and imaging modalities. *Eur J Cardiovasc Med.* 2011;1(3):84-7.
6. Jacobs A, Weinet L, Goonewardena S, Gomberg-Maitland M, Lang R. Three-dimensional transthoracic echocardiography to evaluate cor triatriatum in the Adult. *J Am Soc Echocardiogr.* 2006; 19 (4): 468-4.
7. Markham R, Rahman A. Multimodality imaging of cor triatriatum sinistrum diagnosed in an adult female. *Int j Cardiol.* 2015 March;185:90-2.
8. Isik O, Akyuz M, Ayik MF, Levent E, Atay Y. Cor triatriatum sinistrum: a case series. *Turk Soc Cardiol.* 2015;44(1):20-3.
9. Saxena P, Burkhart H, Schaff H, Daly R, Joyce L, Dearani J. Surgical repair of Cor Triatriatum Sinister: The Mayo Clinic 50-year experience. *Ann Thorac Surg.* 2014;97(5):1659-63.