

Cor Triatriatum Sinistrum in an Asymptomatic Adult: A Case Report

Triatriatum Sinistrum em Adulto Assintomático: Relato de Caso

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Abstract

Cor triatriatum is a rare congenital heart anomaly often diagnosed in early childhood. This case study features an adult with an incidental finding of cor triatriatum sinistrum. Based on the clinical presentation, the patient was treated conservatively. Cor triatriatum sinistrum echocardiographic image findings of this patient are presented along with a narrative review of the literature about this disease.

Introduction

Cor triatriatum sinistrum (CTS) is a rare congenital heart disease that consists of a fibromuscular membrane obliquely dividing the left atrium into two chambers, one proximal, which receives the pulmonary veins, and the other distal, which contains the left atrial appendage and is continuous with the mitral valve. CTS was first observed in 1868¹ by Andral and Church; in 1905, Borst called it "cor triatriatum." It occurs in 0.1% of clinically diagnosed cases of congenital heart disease (CHD) and in 0.4% of autopsy cases of CHD. The prevalence of CTS in the general population is likely less than 0.004%. The embryological origin is controversial; some theories have been created to explain it, the most commonly accepted being the poor incorporation of the common pulmonary vein into the left atrium creating two chambers separated by a septum. Several congenital anomalies may be associated, primarily the ostium secundum atrial septal defect (ASD) and anomalous pulmonary venous drainage.²

CTS can be classified by the pulmonary venous drainage and the presence of ASD using Lam classification (Table 1).³

The clinical presentation of CTS depends on the total area of the orifice(s) and the presence and severity of any associated defects. Most patients are symptomatic since early childhood with findings similar to severe mitral stenosis (dyspnea and pulmonary congestion).³ In the natural history of the disease, 75% of untreated affected patients die during childhood⁴ due to congestive heart failure and pulmonary edema. Patients with relatively large orifices may remain asymptomatic for life and their condition be incidentally discovered,⁵ or they may develop symptoms secondary to stenosis by fibrosis and calcification or mitral regurgitation (MR) after the second and third decades of life.²

Keywords

Cor Triatriatum sinistrum, Heart Defects, Congenital, echocardiogram.

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Table 1 – Lam classification.

Type	Description
A	The proximal chamber receives the pulmonary veins, while the distal chamber contains the left atrial appendage and the mitral valve. There is no defect in the interatrial septum.
A1	Interatrial communication between the right atrium and the proximal chamber.
A2	Interatrial communication between the right atrium and the distal chamber.
B	The pulmonary veins drain into the coronary sinus.
C	No anatomical connection between the pulmonary veins and the proximal chamber.

Source: Lam CR, Green E, Drake E. Diagnosis and surgical correction of 2 types of triatrial heart. *Surgery* 1962; 51:127–37. Saxena P, Burkhardt HM, Schaff HV, Daly R, Joyce LD, Dearani JA. Surgical repair of cor triatriatum sinister: the Mayo Clinic 50-year experience. *Ann Thorac Surg*. 2014;97(5):1659-63.

In adults, dyspnea, hemoptysis, and orthopnea are more common² due to pulmonary venous drainage obstruction, pulmonary venous/arterial hypertension, and heart failure. Palpitations may also occur due to the presence of cardiac arrhythmias (extrasystole or atrial fibrillation)⁶ and systemic thromboembolism due to intra-atrial thrombus formation.⁷

Physical examination findings will vary depending on the associated anomalies. Electrocardiogram (ECG) and chest X-ray findings also depend on the anatomical changes consequent to the severity of these concomitant pathologies.

Echocardiography (ECHO), whether two-dimensional, three-dimensional, or transesophageal, is an important tool in the diagnosis of this congenital condition.⁸

CTS management depends on the hemodynamic effects of the atrial membrane. Patients with restrictive and symptomatic orifices should undergo surgical membrane resection at any age.

This report describes the case of a patient with CTS evaluated with two- and three-dimensional transthoracic Doppler ECHO, transesophageal ECHO, and cardiac magnetic resonance imaging (CMRI) and the management adopted for the case.

Case report and discussion

An asymptomatic 52-year-old man attended a scheduled medical check-up. He was a car driver born in the state of MG, Brazil. He was not using any medications. Pathological history: He reported being hospitalized several times as a child with shortness of breath. His mother said it was asthma. He had a normal childhood and played like other boys his age. He denied having diabetes mellitus, rheumatic fever, alcoholism, or smoking. Family history: His brother and mother died of acute myocardial infarction (AMI), but he did not remember exactly at what age. Physical examination: Normal color. Medium height. Blood pressure: 140/110 mmHg, Heart rate: 72 bpm. An irregular heart rhythm with 2 sound and a B1 split on inspiration was noted. The patient

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presented systolic regurgitation murmur ++/(6) in mitral focus. The lungs were clinically clean. The pulse was irregular but universally palpable and of good amplitude. ECG: Sinus rhythm with several ventricular extrasystole and S/V, left atrial enlargement, right atrial enlargement, and an incomplete right bundle branch block. Although the chest X-ray findings were normal, we observed a posterior left atrial increase in the posteroanterior view (Figure 1). Two-dimensional ECHO (2D ECHO) with Doppler showed thoracic aortic ectasia (3.7 cm/2.3 cm/m² in the sinuses of Valsalva and 3.25 cm/2.0 cm/m² in the ascending proximal segment). Bi-atrial augmentation and mild to moderate MR were also noted. The interatrial septum was intact. The ejection fraction calculated using Simpson's method was 78%. Pulmonary arterial systolic pressure (PASP) was estimated at 33 mmHg. Imaging findings were suggestive of CTS (Figure 2). Abdominal ultrasound (11/24/20) revealed hepatic steatosis and a renal cyst.

A three-dimensional ECHO (Figure 3) showed a clear left intra-atrial membrane (cor triatriatum). MR was moderate to severe due to mitral annular displacement and mitral valve prolapse (MVP). The leaflet was inserted in the posterior part of the left atrium but not in the muscular mitral annulus crest. This type of prolapse is often associated with cardiac arrhythmia.

Transesophageal ECHO showed (Figure 4) enlarged left cavities and slightly decreased global LV systolic function (45.95% ejection fraction using the Teichholz method) with diffuse wall hypokinesia. LV wall hypertrophy was evident. Cor triatriatum was noted in the LA with a maximum gradient of 11 mmHg (mean, 4 mmHg). He also presented MVP with signs of myxomatous degeneration and high posterior leaflet insertion into the LA. Doppler showed moderate MR. Tricuspid valve prolapse with signs of myxomatous degeneration were evident. Mild tricuspid regurgitation was noted with an estimated PASP of 21 mmHg.

CMRI confirmed the CT findings, ruling out other possible associated myocardial structural anomalies (Figure 5).

The patient also underwent a stress test on a treadmill and remained asymptomatic, reaching 9 metabolic equivalents at the end of the test, when he reached 100% of the maximum predicted frequency.

Discussion

CTS is a rare congenital heart malformation with an estimated incidence of 0.1–0.4% among CHDs. In cor triatriatum, the atrium is divided into two parts by a tissue fold, membrane, or a

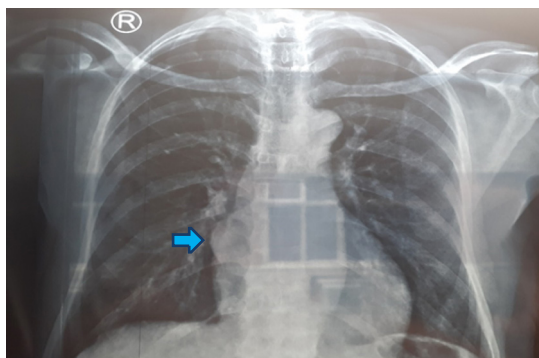
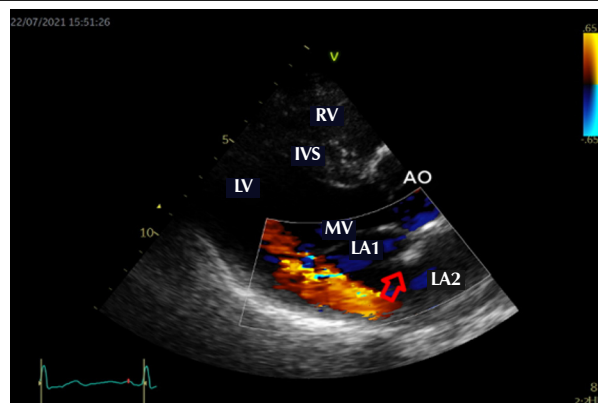
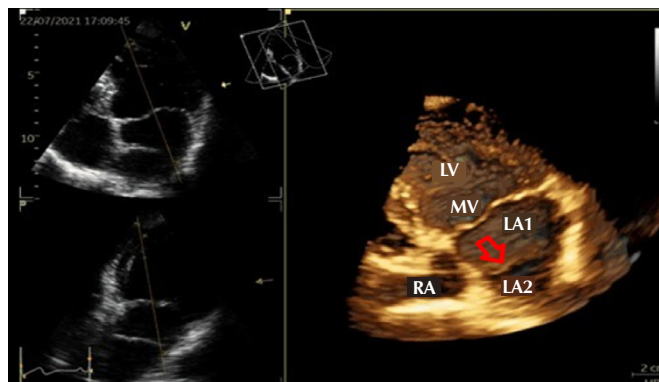


Figure 1 – Chest X-ray in posteroanterior position. The arrow indicates posterior left atrial enlargement.



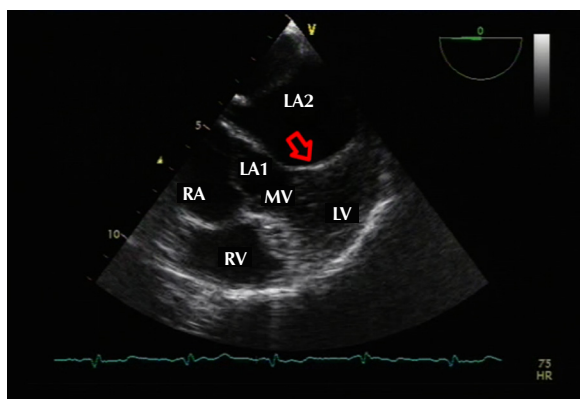
AO, aorta; IVS, interventricular septum; LV, left ventricle chamber; LA1, left atrium chamber; LA2, left atrium chamber 2; MV, mitral valve; RV, right ventricle.

Figure 2 – Transthoracic color Doppler echocardiogram in parasternal section and longitudinal axis. The red arrow indicates the membrane dividing the LA into two chambers (LA1 and LA2).



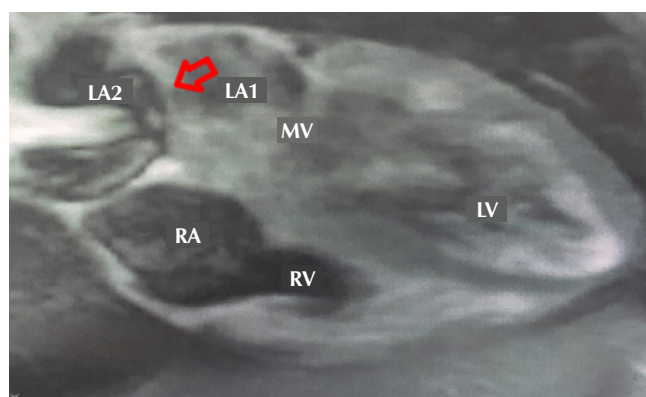
LA1: left atrium chamber 1; LA2: left atrium chamber 2; LV: left ventricle chamber; MV: mitral valve; RA: right atrium.

Figure 3 – Three-dimensional echocardiogram. The red arrow indicates the membrane dividing the LA into two chambers (LA1 and LA2).



LA1: left atrium chamber 1; LA2: left atrium chamber 2; LV: left ventricle chamber; MV: mitral valve; RA: right atrium; RV: right ventricle.

Figure 4 – Transesophageal echocardiogram. The red arrow indicates the membrane dividing the LA into two chambers (LA1 and LA2).



LA, left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle.

Figure 5 – Cardiac magnetic resonance image. The red arrow indicates the membrane dividing the LA into two chambers (LA1 and LA2).

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fibromuscular band that can occur in the CTS and the right atrium. In the pediatric population, this anomaly may be associated with major congenital cardiac defects, while in adults, cor triatriatum is often an isolated and even rarer finding. The literature reports that adult patients with CTS are rarely asymptomatic.^{2,5} ECHO is the most common imaging technique for diagnosing cor triatriatum, although transesophageal ECHO is also needed to precisely define membrane anatomy, its relationship to other structures, and the pulmonary venous drainage pattern.⁸

As the patient was asymptomatic with a New York Heart Association functional class I, we chose to control his systemic blood pressure with an angiotensin II AT1 receptor blocker (ARB) and to follow him up.

Conclusion

During a routine check-up visit, an asymptomatic man was diagnosed with CTS. This congenital condition is rare in

adulthood. We performed several imaging tests and a functional test and performed a narrative review of the literature. We opted for conservative treatment and observational follow-up. Imaging findings classified our patient as Lam type A (Table 1). The patient signed an informed consent form allowing the publication of his data.

Authors' contributions

Alves M: study design; Amaral CSS, Morcerf FAP: data collection; Alves M: data analysis and interpretation; Alves M: manuscript writing; Alves M: critical review of the manuscript for important intellectual content; Morcerf FAP: images.

Conflict of interest

The authors have declared that they have no conflict of interest.

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