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Case Report

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Cor triatriatum sinister, a classic but somewhat forgotten anomaly: Apropos of a case

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Images of complementary tests are shown with the patient's consent.

Abbreviations
CHD: congenital heart defects
LA: left atrium
MS: mitral stenosis
PPVV: pulmonary veins

ABSTRACT

A heart with three atria, totaling five chambers in all, was a fact that surprised medicine more than a century ago. This rare congenital heart defect has a very low incidence, which makes it very little known to the young generations of doctors dedicated to cardiovascular diseases. The *cor triatriatum sinister* is usually diagnosed in early childhood, and it is considered a congenital cause of mitral stenosis. Surgical approach is the choice to release the obstacle in order to adequate blood flow through the left atrium. We present the case of a young adult with typical symptoms of mitral stenosis, without apparent key elements on the physical examination and that the echocardiogram showed this anomaly, hardly seen and published in our field in adult patients.

Keywords: Congenital heart defects, *Cor triatriatum*, Mitral estenosis, Left atrium

Cor triatriatum sinister, una anomalía clásica pero un tanto olvidada: A propósito de un caso

RESUMEN

Un corazón con tres aurículas, que sumaba cinco cámaras en total, fue un hecho que sorprendió a la medicina hace ya más de un siglo. Este raro defecto cardíaco congénito tiene muy baja incidencia, lo cual hace que sea muy poco conocido por las jóvenes generaciones de médicos dedicados a las enfermedades cardiovasculares. El cor triatriatum sinister generalmente se diagnostica en la temprana infancia y es considerado una causa congénita de estenosis mitral. La conducta quirúrgica es de elección para poder liberar el obstáculo al adecuado flujo sanguíneo a través de la aurícula izquierda. Se presenta el caso de un adulto joven con síntomas típicos de estenosis mitral, sin aparentes elementos claves al examen físico y con un ecocardiograma que evidencia esta anomalía, apenas vista y publicada en nuestro medio en pacientes adultos.

Palabras clave: Defectos cardíacos congénitos, Cor triatriatum, Estenosis mitral, Aurícula izquierda

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INTRODUCTION

Congenital heart defects (CHD) have an estimated incidence and prevalence of 5-10 cases per 1000 live births. On the African continent, ventricu-

lar septal defect is the most frequent CHD diagnosed in infancy with an incidence of nearly 2-6 cases per 1000 live births[1,2]. Little is known in Angola about the actual prevalence of CHD as there are very few studies providing reliable statistical data[3].

Cor triatriatum is a rare CHD that typically originates in the left atrium (LA) with a varying incidence that is generally estimated to be less than 1 per 10000 live births. This anomaly has been found more frequently among men at a ratio of 1.5:1 with respect to women and is surgically correctable in most cases[4]. *Cor triatriatum* was first described in 1868[5]. It may be an isolated anomaly or part of a number of rather complex disorders in which the left atrium is bisected by a membrane or fibromuscular roll into two distinct chambers (proximal and distal). This anomalous tissue is considered an embryological remnant that persists in the atrial chamber as a result of disorders in cell migration and apoptosis, due to a group of factors yet to be elucidated[6].

In 1949, *Cor triatriatum sinister* was classified (for didactic purposes) by Loeffler[7] into three subgroups according to the integrity of the fibromuscular membrane:

- Type I: No opening in the accessory membrane.
- Type II: Small restrictive openings (fenestrations).
- Type III: Large opening in the membrane with almost no restriction to blood flow from the pulmonary veins (PPVV) to the left ventricle, which explains why these patients can reach adulthood with hardly any symptoms of its presence[7].

Another more recent[8] but less commonly used classification divides it into three types, depending on the relationship between the connection or not of the PPVV with the rest of the LA:

- Type I: All PPVV communicate with the LA.
- Type II: No direct communication between PPVV and the LA.
- Type III: Some PPVV communicate with the LA.

Greater or lesser blood flow obstruction within the atrium generates a pressure overload with retrograde transmission to the pulmonary capillary bed, with the subsequent pressure repercussion in the right chambers showing a physiology typical of mitral stenosis (MS), which explains the clinical symptoms of these patients. That is why several researchers have recognized this CHD as a cause of congenital MS in recent decades[8-10]. The anomaly is frequently diagnosed in childhood and the affected individual rarely reaches adulthood with no symptoms. *Cor triatriatum* is characterized by the classic symptoms associated with MS where dyspnea on exertion, fatigue and palpitations are the most frequent. Isolated cases of syncope, pulmonary and systemic embolism, pulmonary hemosiderosis and sudden death have also been reported[8,9].

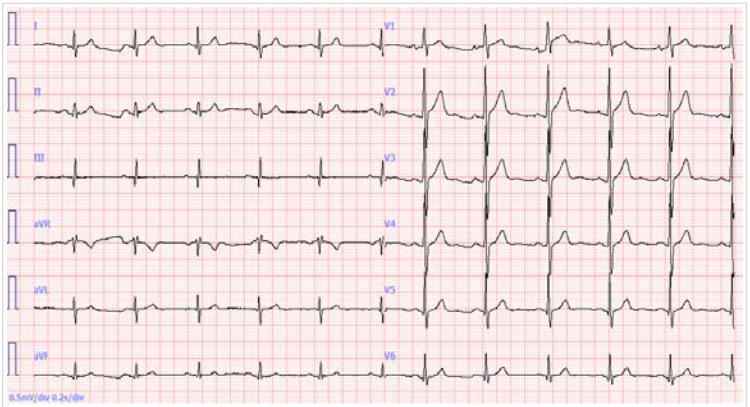

Figure 1. 12-lead electrocardiogram.

Herein, we present a case of *cor triatriatum* in the LA of a young man who presented to the Cardiology Department due to dyspnea on exertion and chest pain of several moths of onset of symptoms.

Herein, we present the case of a young man with *cor triatriatum* in the left atrium who presented to the Cardiology Department due to dyspnea on exertion and chest pain of several months onset.

CASE REPORT

A 40-year-old Chinese man with a history of high blood pressure and a long-time smoker, who was referred to the Cardiology Department complaining of easy fatigue, shortness of breath and a three-month history of chest pain. Physical examination showed no relevant findings, but electrocardiogram, chest x-ray and transthoracic echocardiogram were indicated after personal pathologic history was con-

sidered.

Complementary tests

- 12-lead electrocardiogram (**Figure 1**): Sinus rhythm with narrow QRS is observed and the presence of prominent R waves in V1 is noteworthy since such disturbance may be associated with signs of right chamber overload.
- Plain chest X-ray, posteroanterior view (**Figure 2**): Slight bilateral hilar enlargement, cardiothoracic index within normal range.
- Transthoracic echocardiogram (**Figure 3**): The membrane or fibromuscular roll dividing the LA into two chambers is clearly seen; resulting in the typical image of a triatrial heart (RA, LA^1 and LA^2); therefore, the diagnosis of *cor triatriatum sinister* was established.

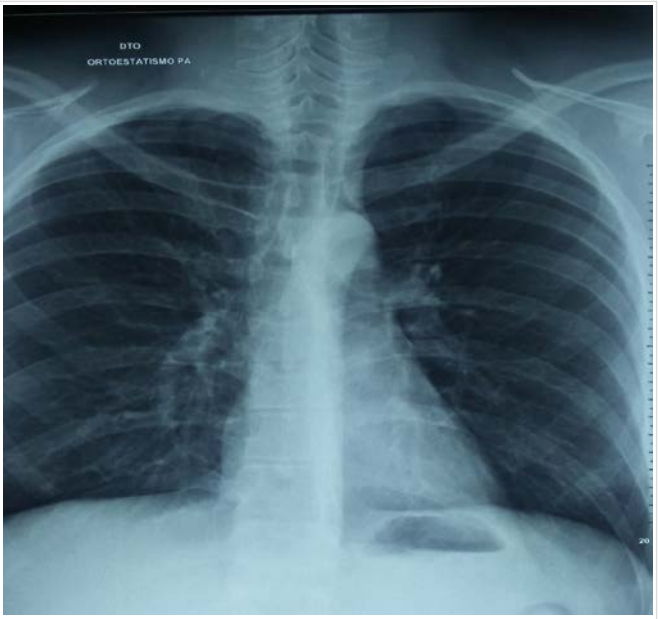

Figure 2. Plain chest X-ray, posteroanterior view.

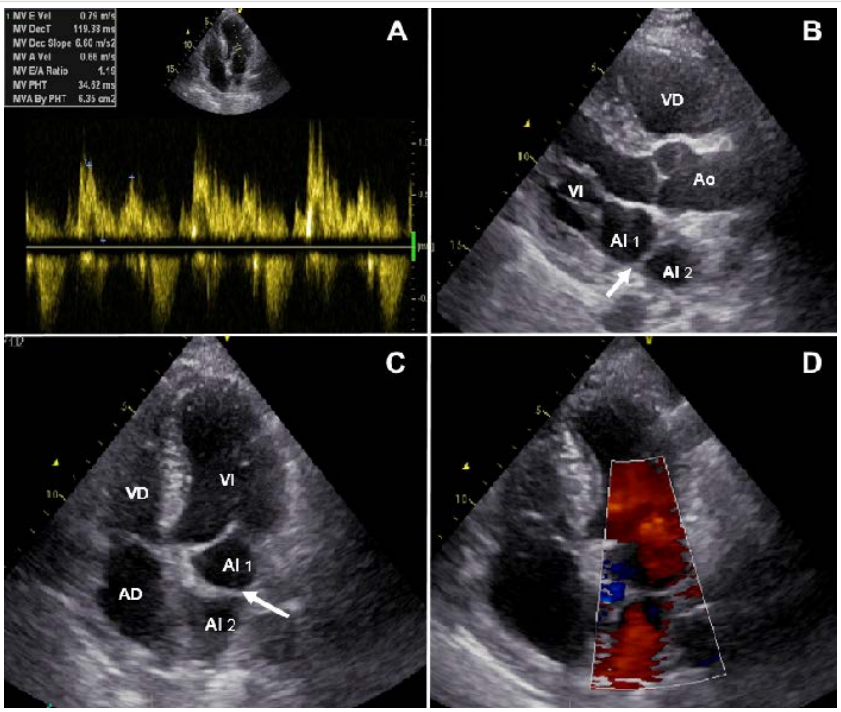

Figure 3. Transthoracic echocardiogram. **A.** Mitral flow with pulsed Doppler. **B.** Two-dimensional mode (parasternal long-axis view) with two chambers (LA1 and LA2) divided by the membrane or fibromuscular roll (arrow). **C.** Apical 4-chamber view showing a typical triatrial heart (*cor triatriatum* - RA, LA1, LA2). Arrow points to membrane in LA. **D.** Color Doppler image of the left chambers.
Acronyms in Spanish. Ao, aorta; AD, right atrium; AI, left atrium; VD, right ventricle; VI, left ventricle.

Further studies were indicated (transesophageal echocardiogram and cardiac computed tomography) to rule out other possible CHD or any associated comorbidity that could explain the symptoms, mainly chest pain, since atherosclerotic disease of the coronary arteries is common in individuals with age and risk factors similar to those of this patient. However, these studies were not carried out as the patient finished his working contract in Angola and, supposedly, went back to China; reason why the work team unfortunately does not know, for sure, the conclusion of the case. We can only speculate that fatigue and dyspnea were the first clinical manifestations of a *cor triatriatum sinister* causing functional MS or that it was simply an incidental finding and not the culprit for all the symptoms that prompted initial medical evaluation and echocardiographic examination.

COMMENT

In 1868, Church[5] reported –for the first time– an unusual anatomical disorder in which the heart of a human corpse had five chambers instead of four as was commonly known. This was due to the presence of a tissue rim, similar to a septum, which divided the LA into two chambers: an upper chamber receiving venous return from the lung through the PPVV and a lower or true chamber which included the left atrial appendage and mitral valve.

The finding was termed *cor triatriatum sinister* and since then to date the literature has reported man-y and dissimilar cases in all kinds of contexts: during childhood, adulthood, associated with complex CHD and in isolation[4-10]. Subsequently, it was observed that this disorder could also compromise the right atrium and thus the anatomical possibility of a right variant (*dexter*), which is much more infrequent, was identified. When the dividing structure was located in the right atrium, it hindered systemic venous return, causing a right heart failure syndrome of varying degree. Today, this membrane is thought to be the result of complex anomalies related to the formation of the Eustachian/Thebesian valves and Chiari network, or associated with defects in the formation of the coronary sinus[10].

Cor triatriatum sinister is indeed an extremely rare CHD. Some hypothesize that it may arise during cardiac embryogenesis when the common pulmonary vein fails to properly incorporate into the dorsal left atrial wall due to complex and currently poorly understood mutations, resulting in a fibromuscular membrane that divides this structure into two chambers[6,11]. Once this embryological remnant produces severe obstruction, the incidence of complications will be high, especially when associated to other CHD; the most frequent are: interatrial or interventricular septal defects, persistent left superior vena cava, stenosis of one or more PPVV and, to a lesser extent, mitral and aortic regurgitations[12,13].

The vast majority of cases exhibit a myriad presenting features of MS from early childhood onwards. On exceptional occasions and due to favorable conditions of obstacle permeability or absence of pulmonary venous return disorders, some patients reach adulthood with no symptoms at all, and therefore without being diagnosed[13,14].

Early diagnosis in asymptomatic patients is rather challenging and most of the times this CHD is found incidentally on routine examinations due to nonspecific and subtle symptoms, such as dyspnea on exertion, orthopnea, hemoptysis, atrial fibrillation and chest pain[8,9,15]. All of this coincides with the case presented in whom symptoms suggestive of MS prompted consultation for cardiac evaluation. The tests did not allow demonstrating associated CHD and, apparently, the intra-atrial structure evidenced in the transthoracic echocardiogram was not causing significant restriction to intra-atrial blood flow, as evidenced in the analysis of the spectral Doppler curve in the mitral valve (**Figure 3A**).

Echocardiography is recognized as the method of choice for diagnosis and hemodynamic evaluation of this CHD. Pulsed and color Doppler modalities are vital for chamber identification, as well as for assessing intra-atrial pressure gradients[16]. The introduction and greater availability of transesophageal and three-dimensional echocardiography and cardiac magnetic resonance imaging has made possible a more detailed and multilevel evaluation of the origin, extent, morphophysiology and CHD associated with this peculiar malformation[16-18].

The surgical approach commonly proposed is removal of the membrane, especially in symptomatic patients regardless of age. In the case of asymptomatic or oligosymptomatic adults, a controversial dilemma arises as to whether maintaining a conservative approach or proceeding with surgical treatment[19,20].

Li *et al.*[21] published an article in which two selected cases underwent a hybrid approach: intraoperative balloon dilatation of the membrane –based on the standard principles of percutaneous mitral valvuloplasty– followed by surgical resection, with the aim of exploring the feasibility of this type of strategy for the treatment of *cor triatriatum sinister*, and concluded that a percutaneous strategy is recommended only for isolated forms of *cor triatriatum sinister*, where all PPVV drain into the LA. Moreover, a number of anatomical features that may predict successful outcome of the procedure should be considered: membrane location, stenosis degree of PPVV, membrane calcification and presence of other CHD; as its long-term efficacy is yet to be confirmed. For the time being, surgical treatment remains the most successful for *cor triatriatum sinister* in symptomatic adult patients, especially when there are other CHDs requiring surgical correction[21]; however, structural heart surgery could eventually become a feasible low-risk solution for these patients, without requiring median sternotomy and cardiopulmonary bypass.

CONCLUSIONS

This is the first case diagnosed at the Clínica Girassol and, probably, in Angola. Hence, the finding should be thoroughly reviewed and published since, due to the infrequent incidence of this CHD, all details of this condition are not fully known. That could lead to misdiagnosis or even worse, to the assumption that it is nothing more than an intracardiac foreign body or a false image due to addition at the time of performing the echocardiogram; leading to an erroneous approach if the physician lacks experience, badly affecting the patient.

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