

## **Cor triatriatum sinister with left superior vena cava, unroofed coronary syndrome and valvular pulmonary stenosis**

### **Cor triatriatum sinistrum con vena cava superior izquierda, síndrome coronario destechado y estenosis de la válvula pulmonar**

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#### **Abstract**

A 25-year-old woman with a history of recent dyspnea and palpitation was admitted to our center. Transthoracic echocardiography (TTE) showed an echogenic and septal muscular ridge along the left atrium which were indicative of cor triatriatum sinister (CTS). Further evaluation with transesophageal echocardiography (TEE) showed that a lower chamber of divided left atrium receiving lower right inferior pulmonary vein, mitral valve, left superior vena cava (LSVC) and unroofed coronary sinus (CS). Also, the lower chamber had an unrestrictive communication with the right atrium. The upper accessory chamber receiving one left and one right upper pulmonary vein and communicated with the right atrium by a small atrial septal defect (ASD). However, upper and lower pulmonary venous systems separated directly from each other by a muscular ridge without the presence of any window or hole to allows blood flow between these two accessory chambers. Although the absence of septum associated with the presence of LSVC and unroofed CS makes our case a unique or very rare type of this complex anomaly in an adult case. In our case, surgical removal of dividing muscular membrane with redirection of LSVC and unroofed CS to the right atrium are warranted.

**Keywords:** Cor triatriatum. Echocardiography. Heart disease.

#### **Resumen**

Se trata de una paciente de sexo femenino, de 25 años de edad, con antecedentes recientes de disnea y palpitaciones que ingresó a nuestro centro. Un ecocardiograma transtorácico (ETT) evidenció un reborde ecogénico muscular del tabique de la aurícula izquierda indicativo de cor triatriatum sinistrum (CTS). En un estudio más a fondo con ecocardiografía transesofágica (ETE) se evidenció que una cámara inferior de la aurícula izquierda dividida recibía la vena pulmonar inferior derecha, la válvula mitral, la vena cava superior izquierda (VCSI) y el seno coronario (SC) destechado. Además, la cámara inferior tenía comunicación libre con la aurícula derecha. La cámara superior secundaria recibía una vena pulmonar superior izquierda y una derecha y se comunicaba con la aurícula derecha a través de una comunicación interauricular (CIA) pequeña. Sin embargo, los sistemas venosos pulmonares superiores e inferiores se encontraban separados entre sí por un reborde muscular sin la presencia de alguna ventana u orificio que permitiera el flujo de sangre entre estas dos cámaras secundarias.

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*La ausencia del tabique junto con la presencia de la VCSI y el SC destechado hacen que nuestro caso sea un tipo único o extremadamente inusual de esta anomalía compleja en un caso en un adulto. En este caso, se justifica la extirpación quirúrgica de la membrana muscular divisoria con redireccionamiento de la VCSI y el SC destechado a la aurícula derecha.*

**Palabras clave:** *Cor triatriatum. Ecocardiografía. Cardiopatía.*

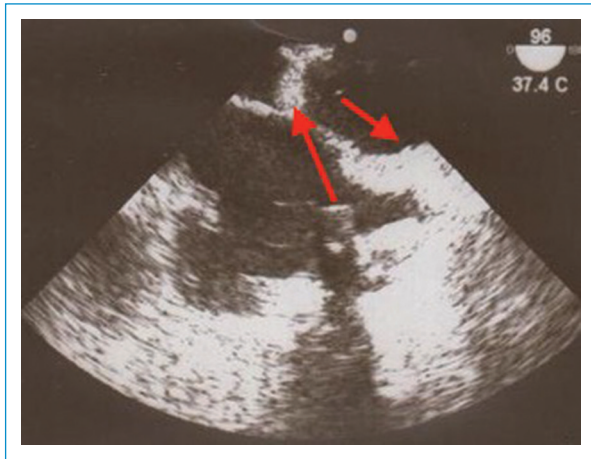
## Introduction

*Cor triatriatum sinister* (CTS) is a rare congenital anomaly detected in children less than 10 years old. In this anomaly, the left atrium is separated into two smaller chambers by the presence of a fenestrated or non-fenestrated muscular septum<sup>1</sup>. The time of CTS presentation mutually depending on the presence of communication between two or each accessory chamber with the right atrium. In the presence of a membrane between two accessory chambers and with the absence of communication with the right atrium, the size of the existing hole between two accessory chambers has an important effect on the amount of flow<sup>2</sup>. However, there are no characteristics of clinical features that differentiate CTS from atrial septal defect (ASD) or valvular anomaly but, breathlessness has been found as the most common symptom at the time of presentation in adults. Two important precipitating diseases in the early presenting of symptoms are atrial fibrillation (AF) or the presence of mitral regurgitation<sup>2</sup>. This exceptional case report demonstrates the unique anatomic variations in CTS and the echocardiographic challenges for the cardiologist and making an unplanned surgery for a surgeon that may encounter when presented with this unusual cardiac anomaly.

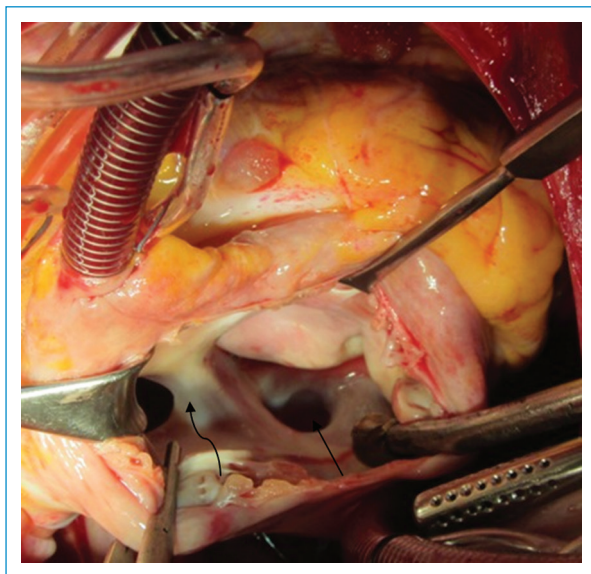
## Clinical case

A 25-year-old girl was referred by her primary family physician for evaluating her dyspnea. The patient complained of weakness in walking, lightheadedness lower extremity muscular pain, chest discomfort, dyspnea or palpitations. No history of taking any drugs such as statin was reported. On physical exam, her blood pressure was 120/70 mmHg, heart rate 100 beats per minute (bpm), respiratory rate 21 breaths per minute, and oxygen saturation 95%. Her femoral and pedal pulses in both lower extremities were normal. He had not any apparent respiratory distress but she complained from exertional dyspnea. She also reported palpitation that was noted to be AF rhythm, rales were found, but a harsh murmur was heard on the left precordial area. Peripheral pulses, especially pedal pulses, were full and strong. An electrocardiogram revealed AF with a rapid

ventricular response of 100 bpm with the absence of ST-segment changes or T wave morphology indicative of ischemia. Laboratory exams which including liver and renal function tests, thyroid function tests, complete blood count, fasting blood sugar, and fasting lipid profile were unremarkable. Subsequently, a transthoracic echocardiogram (TTE) was ordered to detect the possible structural cardiac disease as a cause for the recent onset dyspnea and AF. The TTE showed normal aortic and mitral valve function, left ventricular ejection fraction, enlarged right ventricular size, and small left atrium with the presence of moderate size of ASD (2 × 2 cm) similar to a venous type of ASD and no evidence of lower chamber was detected. TTE also showed large left superior vena cava (LSVC) darning to the right atrium. No echogenic linear muscular structure in the left atrium was detected. The estimated mean pulmonary arterial pressure was 40 mmHg. Due to the complicated results of TTE, a preoperative TTE was requested to be done by an expert cardiologist that CTS was confirmed (Fig. 1). The patient was scheduled for operation with a mid-sternotomy, bi-cava, and aortic cannulation, cardiopulmonary bypass (CPB) was instituted. Intraoperative inspection revealed CTS anatomy so upper accessory chamber that receiving upper left and right pulmonary vein was not communicated with lower accessory chamber and drained its inflow blood circulation from upper pulmonary veins via ASD to the right atrium (Fig. 2). The lower accessory chamber was widely communicated to the right atrium such a common atrium. It, not only receiving a large left superior vena cava (LSVC) opening but also an unroofed coronary sinus demonstrated itself as a small opening to the mid-portion of huge LSVC. The mitral was in a normal position. The lower right and left pulmonary veins drained to the lower accessory chamber. The muscular septum between two accessory chambers was widely opened and with ascertaining the proper location of LSVC location, a large fresh pericardial patch was prepared and sutured to the roof of the lower accessory chamber so LSVC and its associated CS directed to drain into the right atrium. The ASD of the upper chamber was repaired primarily with prolene 4/0 suture. Pulmonary valve stenosis is repaired by triple commissurotomy of the valve and the use of a small



**Figure 1.** Preoperative transesophageal echocardiogram: showing the left atrium divided into proximal (+) and distal (\*) chambers by an intact muscular membrane (arrow).



**Figure 2.** Shows intraoperative inspection of anomaly upper (curve arrow) and lower (vertical arrow) left atrial chambers clearly exhibited.

pericardial patch (Fig. 3). The patient weaned from CPB uneventfully and new left atrium anatomy was examined by TEE. The intraoperative TEE showed the absence of gradient across the iatrogenically fenestrated membrane. TEE was also showed no evidence of any left to right shunt across the new pericardial patch or in redirected LSVC (Fig. 4). A TEE with Doppler mode confirmed an iatrogenically performed large communication between the upper and lower accessory chamber. The

mitral valve was structurally normal with no evidence of regurgitation. With the presence of symptomatic AF, use of a large pericardial patch, medical management with warfarin was started and follow-up by regular international normalised ratio (INR) checking with an INR goal of 2.5.

## Discussion

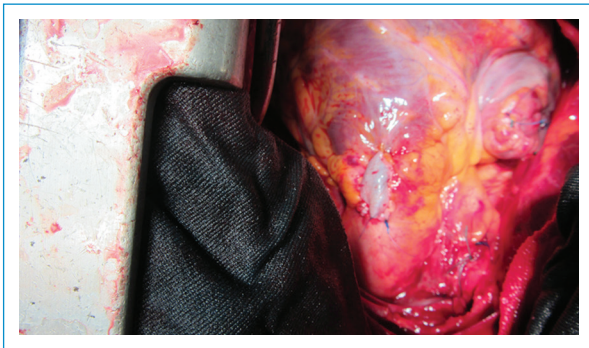
The classic CTS is a rare congenital anomaly characterized by division of the left atrium into an upper or proximal accessory chamber that receives its inflow pulmonary venous blood from all four pulmonary veins<sup>3</sup>. Anatomic characteristics of this anomaly were first explained by Church and showed that CTS consists of 0.1% of all congenital cardiac anomalies<sup>4</sup>. The most important factors in the early detection of the anomaly are related to the obstructing effect of dividing membrane that leads to obstruction of pulmonary venous blood flow. Another factor in precipitating early sign and symptom explained by associated congenital defect as in our case, although detection of severe cases in adult cases have not been increasing despite improvements in modality imaging.

The occurrence of this anomaly during embryonic growth was explained by multiple embryologic hypotheses. Fowler supposed that the mechanism of occurrence of this malformation related to the presence of a defect in normal growth of the embryonic septum primum<sup>5</sup>, but Parsons proposed that anomaly was created by the defective growth of primitive embryonic pulmonary veins bud and its incorporation into the left atrium<sup>6</sup>. Van Praagh supposed that the encroachment of the primitive common pulmonary venous system by the rapid growth of the left appendage of the sinus venous tissue, may preventing its attaching and its incorporation into the future left atrium<sup>7</sup>. Another mechanism described by Gharagozloo showed that the presence of diminutive right SVC leads to the perseverance of the LSVC that influence on the evolving left atrium<sup>8</sup>. However, no logical explanation with expert consensus has been described for the embryogenic cause of this rare cardiac anomaly. There is multiple classification regard to CTS that none of them is coinciding with an anatomic picture of our case. A simple classification of Loeffler that has been planned base on the number of fenestration or size of the defect in the membranous septum, does not explain the rare type of CTS that the lower accessory chamber is completely unroofed<sup>9</sup>. In Loeffler classification, CTS is divided into three types: type one is evident by the presence of a non-fenestrated





**Figure 3.** Postoperative TEE shows iatrogenically created septotomy between two accessory chamber (black arrow).



**Figure 4.** Shows repair of pulmonary valve ring by commissurotomy and small pericardial patch.

septum between two accessory chambers and the presence of an ASD in one of the accessory chambers for outflow draining of pulmonary venous blood flow. Type two is defined by the presence of an obstructing hole or defects between two accessory chambers that obstructing the free flow of pulmonary blood flow to the mitral valve. In type three, a redundant membrane with a wide opening exists between two accessory chambers<sup>9</sup>. In Marin classification, the configuration of the pulmonary venous chamber defines the type of proposed CTS<sup>10</sup>. In Lam classification, the site of inflow of the pulmonary veins to each accessory chamber defined type of CTS<sup>11</sup>. In Lupinski classification, the existence of an ASD to the accessory chambers specified type of CTS<sup>12</sup>, and in the Thilenius classification, the existence of abnormal pulmonary venous return and their location with the left atrial chambers defined type

of CTS<sup>13</sup>. The three common concomitant anomalies with CTS in children including ASD, partial anomalous pulmonary venous connection, and PDA with an incidence of 77%, 53%, and 28% subsequently, most of them were diagnosed by aforementioned anomalies. In opposition to children diagnosis of CTS in the majority of the adult is mainly based on, presence of ASD or mitral insufficiency. The time of presentation of CTS in the adult is multifactorial and the majority of cases presented after increasing mitral valve regurgitation or obstruction of fenestration by age-induced fibrosis process that simulating signs and symptoms of mitral stenosis (MS) and the existence of concomitant anomalies. On the other hand, increasing severity of mitral regurgitation by myxomatous valve degeneration with age can lead to the occurrence of atrial fibrillation and reducing cardiac output and symptom of congestive heart failure. In the adult patient presence of a diastolic murmur, a harsh P2 sound shows the existence of stenotic fenestration and pulmonary hypertension however the absence of an opening snap could differentiate CTS from MS. Some adult cases presented with hemoptysis as a consequence of the obstructing effect of fenestration between two accessory chambers. The physiologic sequel of CTS are mimicking the MS complication, it has been found that MS-like physiologic consequences are an important precipitating factor in such cases that may be presenting with diverse interrelated conditions such interstitial lung fibrosis, atrial fibrillation, stroke, massive thrombus in the pulmonary artery, and rare case of esophageal varices. However, echocardiography is the gold standard imaging for CTS diagnosis, but it has major limitations especially in delineating the proper location of the upper accessory chamber and related structures in adults so, it could not define complete delineation of the muscular membrane of the flow gradient across the hole in the membrane. Then, TEE should be used for better delineation of the muscular septum, intra septal defect, detecting a gradient across the membrane, the existence of ASD, and identification of other pathologic conditions such as MS or supra-valvular mitral ring. In conclusion, treatment of CTS related to the hemodynamic effect of the atrial membrane on the inflow tract of the mitral valve so surgical criteria for surgical intervention in MS are also surrogates for CTS surgery. Careful medical literature revealed that most cases of CTS in children presenting with severe obstruction in inter accessory chambers membrane so almost of them have been operated in early life. But given this fact that less severe obstructions to blood flow across the membrane present in elderly cases,

children are managed with a surgical approach more often than elderly cases in whom with the presence of non-restrictive fenestration warranted that a medical approach to be implemented. CTS in the adult is a rare anomaly and has a wide variety of clinical manifestations that may contribute to a false diagnosis of MS or AF. TEE is a gold standard imaging tool to delineates anatomy and verify concomitant anomaly and helps as a guide to planning an appropriate approach<sup>14</sup>.

In opposition to previously reported cases, our patient had a non-fenestrated septum between two accessories left atrial chambers while upper and lower accessory chambers communicating with the right atrium by small and large ASD subsequently. Other characteristics feature of our case related to the presence of unroofed CS in the lower accessory chamber and drainage of LSVC through a large opening into the lower chamber.

However, echocardiography could detect most cases of CTS, by detecting a gradient between the lower and upper chambers, but our case had not had a fenestrated hole between chambers so the anomaly was not detected preoperatively, while the upper accessory chamber with restrictive ASD to the right atrium was diagnosed as ASD. We did not perform cardiac angiography in cases less than 35 years old age than so presence of lower accessory chamber was missed by unavailable results of chambers' blood saturation values.

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## Conflicts of interest

The authors have no conflicts of interest.

## Ethical disclosures

**Protection of humans and animals.** The authors declare that no experiments on humans or animals have been performed for this research.

**Confidentiality of data.** The authors declare that they have followed their center's protocols on the publication of patient data.

**Right to privacy and informed consent.** The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

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