# Fenestrated cor triatriatum sinistrum: a case report

## Fenestre kor triatriatum sinister: Olgu sunumu

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***Summary–*** We present a 20-year-old male patient with cor triatriatum sinistrum with fenestrations and long-standing dyspnea on exertion, fatigue, and palpitation. An apical early to mid-systolic murmur of grade I-II/VI was heard on cardiac auscultation. Electrocardiography showed sinus rhythm and an incomplete right bundle branch block. Transthoracic echocardiography showed a membrane dividing the left atrium into two compartments. Transesophageal echocardiography showed a membrane with fenestrations originating from the left upper pulmonary vein, extending to the interatrial septum, and dividing the left atrium into two compartments as proximal and distal. Cardiac MR imaging for further detailed anatomical assessment demonstrated similar findings without additional anomaly. Medical follow-up was conducted due to no pressure gradient across the membrane.

***Özet–*** Bu yazıda, fenestrasyonlu kor triatriatum sinisteri bulunan 20 yaşında erkek hasta sunuldu. Hasta uzun zamandır var olan eforla ilişkili nefes darlığı, yorgunluk ve çarpıntı yakınmalarıyla başvurdu. Kardiyak muayenesinde I-II/VI dereceden apikal erken-orta sistole uzanan bir üfürüm saptandı. Elektrokardiyografide sinüs ritmi ve inkomplet sağ dal bloğu vardı. Transtorasik ekokardiyografide sol atriyumu ikiye bölen bir membran izlendi. Transözofageal ekokardiografide sol üst pulmoner venden başlayan interatriyal septuma uzanan üzerinde fenestrasyonlar bulunan sol atriyumu proksimal ve distal olarak iki kısma ayıran bir membran görüldü. Daha detaylı anatomik değerlendirme için yapılan kardiyak MR görüntülemesinde benzer bulgular dışında ek bir anomali saptanmadı. Membran üzerinde basınç gradiyenti olmadığı için hasta medikal olarak izleme alındı.

Cor triatriatum sinistrum (CTS) is a rare congenital heart disease with an incidence of approximately 0.1% in adults.[1] In this disease, the left atrium is divided into two spaces by a fibromuscular membrane, and these spaces are composed of pulmonary veins proximally and the left atrial appendix distally. In most cases, these spaces are linked to each other through one or more orifices. The majority of patients present with clinical signs depending on the size of the opening on the fibromuscular membrane. However, most of these cases (75%) are lost in infancy.[1]

**Abbreviations:**

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| *CTS* | *Cor triatriatum sinistrum* |
| *MR* | *Magnetic resonance* |
| *NYHA* | *New York Heart Association* |
| *TEE* | *Transesophageal echocardiography* |
| *TTE* | *Transthoracic echocardiography* |

In this article, we present a case of a 20-year-old male having fenestrated CTS.

## CASE REPORT

A 20-year-old male patient was admitted to our cardiology clinic with dyspnea on exertion, fatigue, and palpitation. He had been suffering these symptoms for several years, while his medical history was unremarkable. According to the New York Heart Association (NYHA) classification, his functional capacity was class 2. His arterial blood pressure was 120/60 mmHg, his temperature was 36.6°C, his heart rate was 88 bpm with a regular rhythm, and his respiratory rate was 16/min on physical examination. Both lungs were equally participating in breathing and his respiratory sounds were normal. An apical early to mid-systolic

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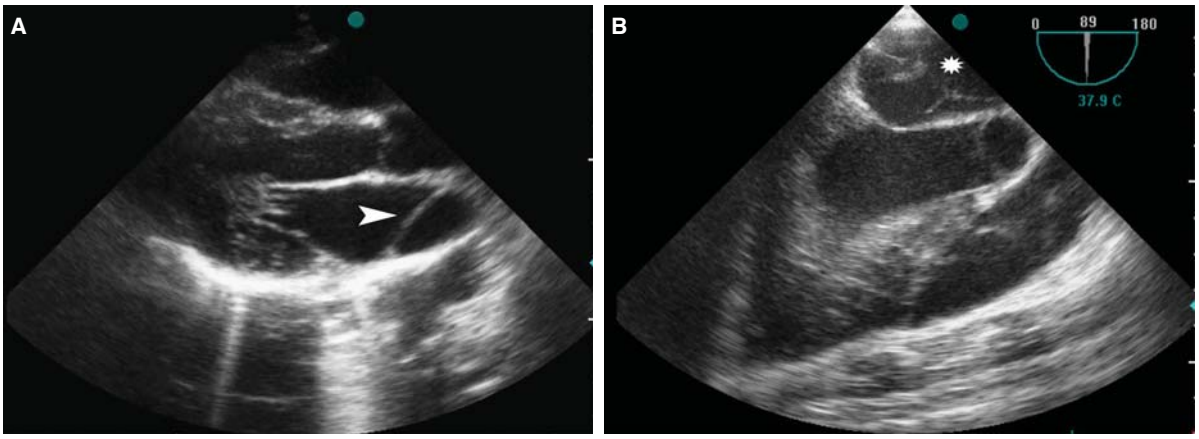


**Figure 1.** **(A)** Cor triatriatum sinistrum dividing the left atrium into two compartments in parasternal long-axis view (arrow). **(B)** Transesophageal echocardiography showing the membrane with fenestrations (star).

murmur of grade I-II/VI, nearly extending into the late systole, was heard on cardiac auscultation. ECG showed signs of an incomplete right bundle branch block with normal sinus rhythm. Transthoracic echocardiography (TTE) (Vivid 3, GE Medical System) showed a membrane dividing the left atrium (Fig. 1A, see supplementary video file 1). Cardiac chambers were normal in width. The left ventricular ejection fraction was 65%. No defect was seen in either the inter-atrial or inter-ventricular septum. For further anatomical diagnosis, transesophageal echocardiography (TEE) was performed (Vivid 3, GE Medical System). Anatomic localization of the pulmonary veins was normal, and the inter-atrial septum was intact. The membrane originating from the left upper pulmonary vein, extending to the interatrial septum, and dividing the left atrium into two compartments as proximal and distal was seen. Fenestrations were observed in this membrane where it adheres to the interatrial septum (Fig. 1B, see supplementary video file 2). Color flow transition between the proximal and distal portions in this region was imaged by color-Doppler echocardiography. We did not obtain a pressure gradient from this region. Cardiac magnetic resonance (MR) imaging was performed as a more advanced imaging method, with which we were able to see fenestrations of the membrane dividing the left atrium (see supplementary video file 3). The patient was followed medically.

## DISCUSSION

CTS was first described by Church in a postmortem study in 1868.[2] In this very rarely seen congenital malformation, the left atrium is divided by an abnormal fibromuscular membrane into a postero-superior chamber receiving the pulmonary veins and an antero-inferior chamber giving rise to the left atrial appendage. The incidence of this malformation is between 0.1% and 0.4%.[3] The severity of clinical symptoms is associated with the size of the defect on the fibromuscular membrane, the resistance to pulmonary venous return, and pulmonary hypertension. In the majority of cases, the diagnosis is made during infancy. However, asymptomatic cases having large defects without any difference in pressure gradients between the proximal and distal spaces have been described. Associated abnormalities such as atrial septal defect, patent foramen ovale, partial anomalous venous return, and persistent left superior vena cava have been reported.[4] Typical symptoms of mitral stenosis may be seen. Either a systolic or diastolic murmur may be heard or the patient may have a completely normal physical examination. A functional capacity of NYHA class 2 and an apical systolic murmur were present in our case. The mechanism of low-grade systolic murmur may be due to flow through the membrane during the filling of the left atrium. It is possible to diagnose the disease by TTE, but in most cases, TEE is required to confirm and to evaluate additional cardiac anomalies.

Multi-slice computed tomography and MR imaging often confirm the diagnosis of CTS. In our study, we confirmed the patient's diagnosis with TTE and TEE. It is important to note that the presence of fenestrations may be misdiagnosed as. To rule out the very rare anomaly of cor polyatriatum, we performed MR imaging on our patient,[5] but this anomaly was not

seen. However, although rare, this anomaly must be kept in mind.

***Supplementary video files associated with this article can be found in the online version.***

***Conflict-of-interest issues regarding the authorship or article: None declared***

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