

# Cor triatriatum sinister: two cases diagnosed in adulthood and a review of literature

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[Received 6 July 2012; Accepted 11 August 2012]

*Cor triatriatum sinister is a rare condition caused by a membrane within the left atrium that separates the pulmonary veins from the mitral valve. While the condition is usually diagnosed in childhood, a rare presentation during adulthood is observed when the membrane is incomplete. We report two cases of incomplete cor triatriatum sinister diagnosed during adulthood and review the literature for this rare anomaly. (Folia Morphol 2012; 71, 4: 275–279)*

**Key words:** cor triatriatum sinister, left atrium, three dimensional echocardiography, transesophageal echocardiography, cardiac computerised tomography

## INTRODUCTION

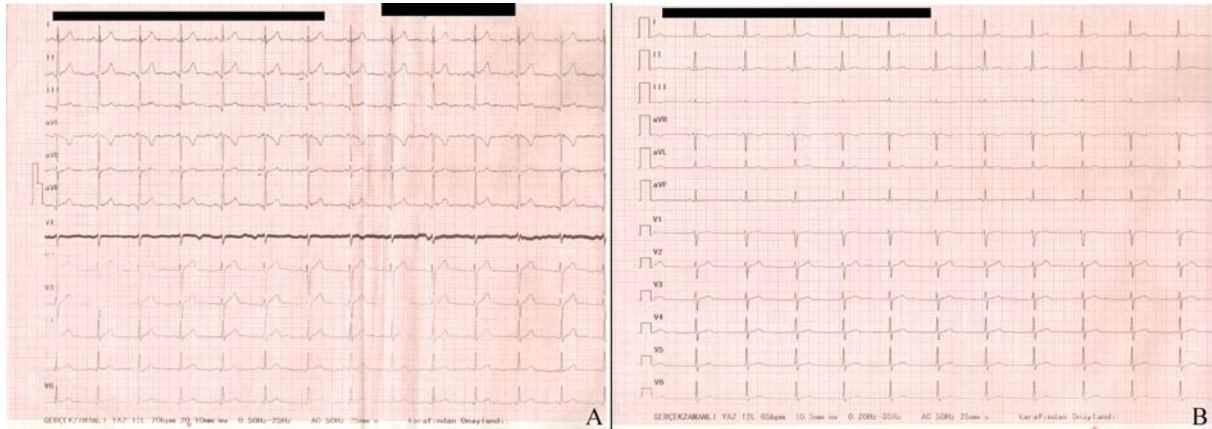
Cor triatriatum sinister (CTS) is a rare congenital anomaly that is caused by a fibromuscular membrane dividing the left atrium (LA) into two chambers. Communication between the pulmonary veins and anterior chamber is provided by fenestrations on the membrane. As a result of atrial flow obstruction, symptoms of congestive heart failure develop during infancy and childhood. When the membrane is incomplete or has large fenestrations, the condition is diagnosed at adulthood due to lack of symptoms.

In this article, we report two cases of incomplete CTS that are diagnosed in adulthood and review the literature for this congenital anomaly.

## CASE 1

A 31-year-old female patient with a complaint of shortness of breath during exercise was admit-

ted to our institution. She had given birth to one child, and her previous medical records did not show any chronic medical condition. During her pregnancy, she had experienced mild shortness of breath that began on the 12<sup>th</sup> gestational week, and these symptoms had abated spontaneously after labour. Her family history was unremarkable. On physical examination, her blood pressure was 100/60 mm Hg and heart rate was 70 bpm. No abnormal findings were present during auscultation of cardiac and lung fields. Likewise, there was no jugular distention, hepatomegaly or pretibial oedema. Her electrocardiogram showed normal sinus rhythm with a rate of 70 bpm. Amplitude, duration, and axis of P waves and duration of PR interval were within normal ranges, and there were no signs of right ventricular enlargement (Fig. 1A). Her chest X-ray was also normal. As she had unexplained dyspnoea, transthoracic echocardiography was performed. Two-

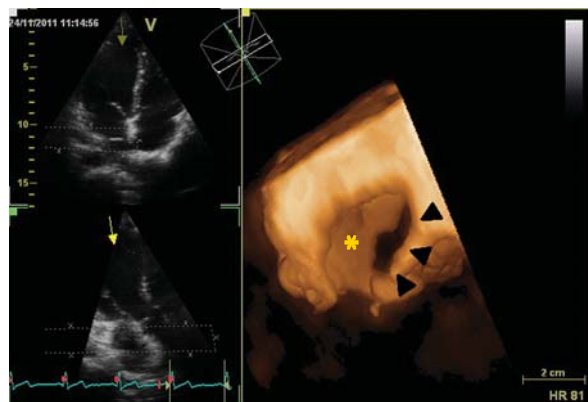


**Figure 1.** Electrocardiograms of Case 1 (A) and Case 2 (B). Both cases had no specific findings for cor triatriatum. An early repolarisation pattern is noted in (A).

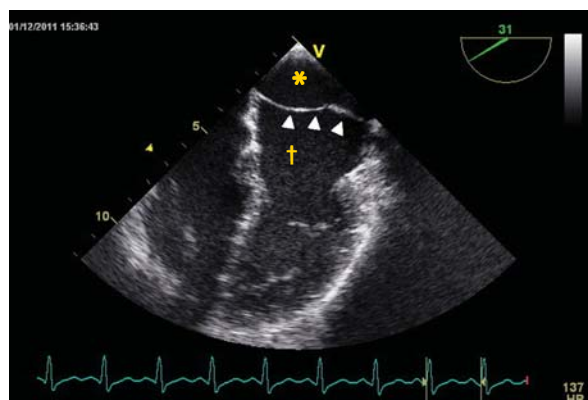
-dimensional (2D) echocardiography showed a partial membrane in LA, and colour Doppler recordings showed that flow was present at the fenestrated part. Three-dimensional (3D) echocardiographic reconstruction of LA revealed a membrane in LA that separated the pulmonary veins from the mitral valve. Continuity of the membrane was complete at the posterior aspect, while a large fenestration was present on the anterior side, which allowed communication between two separate chambers (Fig. 2). Two-dimensional transesophageal echocardiography allowed measurement of the gradient between the two chambers separated by the membrane (Figs. 3, 4). As the pressure gradient was negligible and the patient had minimal symptoms during treadmill exercise test, surgical intervention was not considered.

### CASE 2

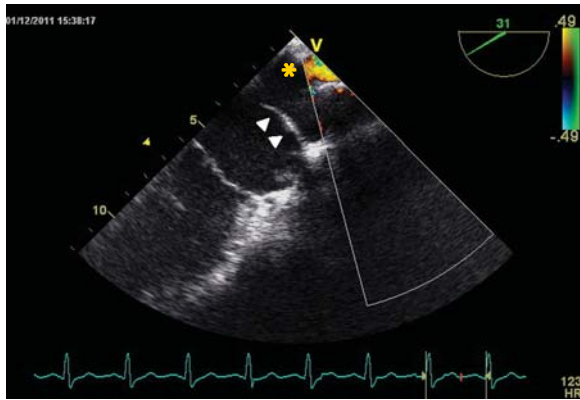
A 27-year-old male patient admitted to our institution with a complaint of chest pain that began one month previously. He was physically active, had no chronic disorders, and he had been smoking one pack/day for ten years. His family history was unremarkable. On physical examination, his blood pressure was 110/70 mm Hg and heart rate was 65 bpm. Auscultation of cardiac and lung fields showed normal findings, and no signs of right ventricular failure were noted. His electrocardiogram showed sinus rhythm with normal P waves and PR duration with a normal QRS axis, and signs of right ventricular enlargement were not present (Fig. 1B). Two-dimensional echocardiography revealed a membrane in the LA, dividing it into two chambers. As his apical window was inadequate for a complete 3D



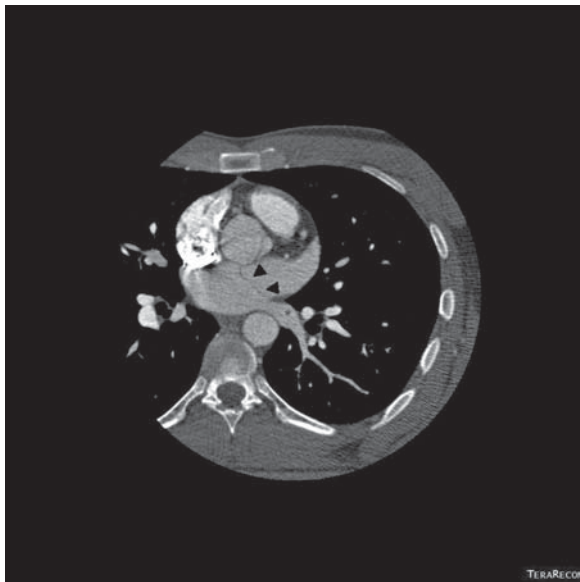
**Figure 2.** Three-dimensional transthoracic echocardiography view of the first patient, showing fibromuscular membrane (asterisk). Black arrowheads point to the opening (foramen) on the membrane. The membrane is shown as seen from the left ventricle. The mitral valve is cropped for a clear view of the membrane.



**Figure 3.** Two-dimensional transesophageal view of the first patient in lower oesophageal four chamber view. The membrane is seen from the long axis of the left ventricle (arrowheads). Posterior chamber is shown with an asterisk, while the anterior chamber is shown with a dagger sign (†).



**Figure 4.** Two-dimensional transesophageal view of the first patient at mid oesophagus showing the relationship between the membrane (arrowheads) and the orifices of the left superior and inferior pulmonary veins. Doppler flow shows pulmonary flow in left inferior pulmonary vein (asterisk).



**Figure 5.** Axial maximum intensity projection computed tomography image demonstrating the membrane (arrowheads) dividing the left atrium into two distinct chambers and the left upper pulmonary vein (asterisk) draining into the posterior chamber near the communication of the membrane to the atrial wall.

echocardiographic reconstruction, cardiac computed tomography (CT) was planned. On cardiac CT, an incomplete membrane separating the atrium into a posterior and an anterior chamber was observed (Fig. 5). All pulmonary veins were in communication with the posterior chamber, while continuation of the membrane was interrupted at the anterior aspect of the LA. As he had no symptoms compatible with outflow obstruction, a treadmill exercise test was performed for hidden dyspnoea. However, dyspnoea was not observed at maximal exercise, and the test

was terminated at the 12<sup>th</sup> minute due to fatigue. As he remained asymptomatic after the treadmill test and had an incomplete membrane, surgery was not considered and follow-up was planned.

## DISCUSSION

Cor triatriatum sinister is a rare congenital cardiac anomaly [10]. This condition is a result of failure of embryologic common pulmonary vein incorporation into the back of the LA [5, 17]. As a result, the LA is divided into two chambers by a fibromuscular membrane [11]. The two cavities, positioned posterior superior and anterior inferior, are anatomically and functionally separated [5, 11].

The incidence of cor triatriatum is 0.1–0.4% of patients with congenital heart disease [11]. This entity can present in classical or atypical form [9]. While the classical form is an isolated thin membrane within the LA, the atypical form is accompanied by other cardiac anomalies [9].

In 1868 Church first described cor triatriatum [1]. After the postmortem description of Church, Miller et al. [14] described this entity by angiography at the Mayo Clinic in 1964. In 1984 Ostman-Smith et al. [15] demonstrated cor triatriatum by echocardiography for the first time.

The classical morphology has been well described in recent years. The volume of the proximal chamber is larger than the distal chamber. Also, the wall of the proximal chamber is thicker, and the pressure is higher in the proximal chamber than the distal chamber [10]. The posterosuperior chamber receives the pulmonary veins and the anteroinferior chamber is continuous with the mitral valve [18].

In cor triatriatum the right and left pulmonary veins join the posterior-superior part of the LA forming an accessory chamber [7, 10]. This chamber, which leads pulmonary venous obstruction, communicates with the LA through a restricted opening [7, 17]. Cor triatriatum is an anatomical barrier of pulmonary venous drainage and a rare, surgically correctable cause of pulmonary arterial hypertension [7].

Persistent left superior vena cava (LSVC) is reported as the most common coexisting abnormality observed with cor triatriatum [9, 10]. Kaneko et al. [9] hypothesised that CTS is caused by a failure of LSVC to disappear during foetal life. As LSVC drains to the coronary sinus, dilation of the coronary sinus could lead to a bulging of the posterior atrial wall. This bulging of the atrial wall is considered as the cause of obstruction of the LA outflow [9].

One of the variations coexisting with CTS is anomalous pulmonary venous return [10]. Anomalous connection of pulmonary veins can be total or partial. Partial anomalous pulmonary venous return can present as right-sided or left-sided anomalous pulmonary venous return [10, 17]. In right-sided partial anomalous pulmonary venous return, right-sided veins join with each other to drain the anomalous proximal chamber, while left-sided pulmonary veins drain into the LA. In left-sided partial anomalous pulmonary venous return, left-sided pulmonary veins join to form a left vertical vein. This left vertical vein connects to the left innominate vein. The remaining pulmonary veins connect into the anomalous proximal chamber as a component of this entity. In some variants, partial anomalous venous return consists of right pulmonary veins draining into the proximal chamber and left veins connecting to the coronary sinus [10].

Other cardiac anomalies that accompany CTS are ventricular septal defect, coarctation of aorta, tetralogy of Fallot, atrioventricular septal defect, and mitral stenosis [10, 13, 18]. Park et al. [16] reported an extremely rare clinical presentation as cerebral infarct due to mitral stenosis in a patient with CTS. Extracardiac manifestations that may be observed with CTS include asplenia and polysplenia [13].

Depending on the severity of obstruction, CTS may be symptomatic or asymptomatic. Symptoms are a result of outflow obstruction and include dyspnoea, orthopnoea, cyanosis, haemoptysis, and chest discomfort. When the obstruction is severe, CTS is usually diagnosed during infancy or adulthood. Some cases may remain asymptomatic or minimally symptomatic during lifetime [16, 18]. Rarely, obstructive symptoms may become manifest during adulthood [8]. In our first case, symptoms attributable to CTS developed during pregnancy, while the patient was minimally symptomatic at diagnosis. An increase in blood volume was considered as responsible for her symptoms during pregnancy, as previously reported by Davlourous et al. [2]. Our second case was asymptomatic.

Electrocardiography is often normal in CTS patients, especially when obstruction is not present. When there is significant obstruction, electrocardiogram may show signs of right ventricular hypertrophy, such as right axis deviation and prominent R or R' wave in V1, which may develop due to associated pulmonary hypertension [19]. In adults, CTS might present with atrial fibrillation (AF), and re-

moval of the membrane could abolish the arrhythmia [20]. Whether this is a direct consequence of the membrane or a result of atrial stretch is unknown. Well-known causes of AF, such as accompanying mitral regurgitation, could also play a role in the development of AF when present [21]. Other reported arrhythmias include atrial tachycardia and ectopic atrial rhythm with abnormal P waves [22, 23]. Interestingly, in the latter case, abnormal P waves disappeared after the operative removal of the membrane, and the authors demonstrated HCN4 + cells with electrical automaticity in the removed material.

Cor triatriatum sinister is a complicated cardiac malformation due to associated anomalies and obstruction at the level of the LA. Haemodynamic parameters and anatomical structures of the heart can be well visualised by 2D transthoracic and transesophageal echocardiography [3]. Three-dimensional echocardiography gives more reliable information about anatomical features than 2D echocardiography [6]. Combined use of 3D echocardiography, transesophageal echocardiography, and Doppler assessment of stenosis severity allows a complete evaluation of CTS [6]. Additional modalities that could be used for diagnosis of CTS include cardiac CT and cardiac MRI [4, 12]. Both techniques allow a complete anatomic evaluation of the lesion when echocardiographic assessment is unavailable or inadequate. In our first case, 3D transthoracic echocardiography was able to completely visualise the membrane, while additional haemodynamic data was obtained with transesophageal echocardiography. In the second case, cardiac CT was used to evaluate atrial anatomy as transthoracic echocardiography views were inadequate.

Surgical removal of the membrane abolishes obstructive symptoms in patients. Urgent operation is needed when the membrane is complete and the proximal chamber communicates with the right atrium via an atrial septal defect [10]. Operation is also indicated when the opening is restrictive and obstruction is manifested, as high death rates are observed in the infancy period with CTS. In adulthood, surgical removal is generally indicated if obstructive symptoms are present. However, additional complications including AF or mitral regurgitation could be observed in adulthood, and could even present symptom of CTS [18]. Our cases were asymptomatic or minimally symptomatic at peak exercise, so surgical removal was not considered.

## CONCLUSIONS

Cor triatriatum sinister can be observed in adulthood when the dividing membrane is incomplete. Echocardiography and CT can readily demonstrate the membrane, while 3D reconstruction allows complete anatomical evaluation. Surgical removal is indicated when obstructive symptoms are present, while management for asymptomatic individuals remains unknown.

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