A Tale of Three Chambers: Cor Triatriatum Sinistrum

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INTRODUCTION

Cor triatriatum sinistrum (CTS) is one of the rarest congenital abnormalities of the heart (0.1%-0.4%) that is defined by the presence of an abnormal fibromuscular septum dividing the left atrium (LA) into two chambers.[1]

CASE PRESENTATION

A 40-year-old patient with a history of sinus bradycardia who had undergone dual chamber pacemaker implantation at 37 years of age, with morbid obesity, type 2 diabetes mellitus, and hyperlipidemia, presented with palpitations, exertional dyspnea, and chest pain. The patient also reported having "three chambers in the heart" found at the age of 18 years during evaluation of a syncopal episode. Interestingly, the patient did not follow up with any cardiologist, although they did undergo two transthoracic echocardiograms (TTE) during hospital admissions for noncardiac reasons that showed possible cor triatriatum and a left ventricular ejection fraction (EF) of 50% in 2014 (Figure 1). On presentation to our hospital, the patient was diagnosed with heart failure with reduced EF of 30% and started on guideline-directed medical therapy, which resulted in improved EF (Figure 2). The patient was morbidly obese and referred for bariatric surgery, resulting in significant reduction in weight. They also had recurrent episodes of symptomatic paroxysmal atrial fibrillation and were referred to an electrophysiologist. The patient underwent direct-current cardioversion two times as well as radiofrequency pulmonary vein isolation ablation. Transesophageal echocardiography (TEE) was done to further evaluate cardiac anatomy and revealed CTS with a small opening of 0.8 cm^2 (Figures 3-5, Videos 1 and 2). The interatrial septum was normal. There was no anomalous pulmonary venous return. There was concern for mitral stenosis–like physiology caused by the CTS membrane, so the patient was referred to an adult congenital heart disease clinic. Right heart catheterization showed pulmonary artery pressure of 50/30 mm Hg, pulmonary capillary wedge pressure of 30.0 mm Hg, and pulmonary venous resistance of 0.8 Wood unit. The cardiac index was 3.5 L/min/m^2. Coronary angiography showed no significant coronary artery disease. The patient underwent surgical correction of CTS. Intraoperative findings included a small patent foramen ovale and a large LA membrane with a tiny aperture near the septum. Surgical LA membrane resection, patent foramen ovale closure, and biatrial maze and LA appendage ligation was performed. Antiarrhythmics were continued, and anticoagulation was stopped after 3 months. The patient continued to do well at 6-month follow-up, with no further atrial fibrillation or symptoms of congestive heart failure.

DISCUSSION

CTS is a congenital heart disease in which the LA is divided into two compartments by a fold of tissue, a membrane, or a fibromuscular band. The exact pathophysiology is unknown, but several theories have been proposed, including malseptation (abnormal growth of septum primum that bisects the atrium), entrapment (embryonic sinus venosus traps the common pulmonary vein and prevents its incorporation into the LA), and the most widely accepted theory, malincorporation, in which the pulmonary vein fails to incorporate into the LA, forming a septumlike appendage that divides the LA into two chambers. Classically, the true atrial chamber is in contact with the atrioventricular valve and contains the atrial appendage as well as the true atrial septum that bears the fossa ovalis, while the accessory portion of the atrium receives venous blood. The membrane that separates the affected atrium into two parts has been documented to be of various sizes and shapes and may be with or without any openings. This determines the severity of symptoms and complications of the disease. The Loeffler classification divides CTS into three groups: group 1, absence of connection between the superior and inferior (true atrium) chambers; group 2, one or a few small openings in the intra-atrial membrane; and group 3, wide opening between superior and inferior chamber.

Although the first two groups cause severe symptoms in childhood and result in high mortality, group 3 usually presents in adulthood, as it may remain asymptomatic for years. The spectrum of symptoms largely depends on the severity of obstruction caused by the membrane. Patients are likely to be asymptomatic when the aperture or fenestration is larger than 1 cm.[2] In a systematic review of literature, Rudienė *et al.*[3] found only 170 reported cases of CTS in adults, with a mean age at diagnosis of 43 years. Forty-one percent of the patients had restrictive physiology and resulting complications of CTS, including atrial fibrillation (32.8%), congestive heart failure (44.3%), and pulmonary hypertension (27.1%). Pulmonary edema during labor has also been reported in young adult women.[4]

It is unclear why patients with Loeffler group 3 CTS who are asymptomatic for years later develop complications. It is thought that there might be electrical and mechanical cardiac remodeling of the tissue predisposing to atrial fibrillation or mitral stenosis–like physiology. Another explanation is the degenerative changes and calcification that can occur over years to create a restrictive physiology of the

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VIDEO HIGHLIGHTS

Video 1: Two-dimensional TEE, midesophageal view, without (*left*) and with (*right*) color flow Doppler, demonstrates flow acceleration across the opening in the cor triatriatum membrane.
Video 2: Three-dimensional TEE, midesophageal view, demonstrates the CTS membrane.

View the video content online at www.cvcasejournal.com.

membrane. However, this theory was disputed by a retrospective study of patients with CTS who were followed for 6 years and showed no significant increase in membrane gradient in patients who were managed conservatively.[5] Regrettably, there are no available studies following patients with CTS for a longer duration to confirm the findings of Fuchs *et al.*[5]

Presenting symptoms of complicated CTS may include syncope, neurologic deficits, chest pain, and shortness of breath. Subtle cardiac auscultatory findings suggestive of CTS may be present on physical examination. Cardiac murmur is typically diastolic with a loud P2 when pulmonary hypertension is present. The absence of an opening snap or a loud S1 marks the difference between CTS and mitral stenosis. Signs of right heart failure, pulmonary hypertension, and pulmonary congestion may also be present.

Diagnostic evaluation begins with TTE. On the four-chamber view, the membrane of CTS is seen as a thin linear structure bisecting the atria distally to the mitral valve and the LA appendage. In the long-axis view, the superior part of the membrane is usually parallel to the aortic wall, while its inferior part connects to the LA posterior wall. One or more foramina may be seen within the membrane, with an increased velocity on color flow Doppler. TTE is also useful in identifying other associated congenital cardiac anomalies, such as atrial septal defects, ventricular septal defects, and tricuspid or mitral stenosis. Spectral Doppler interrogation is used to measure the mean and maximal transmembrane pressure gradients and maximal velocity as well as the pulmonary artery pressure. It has been reported that severe obstruction is indicated by a maximum Doppler velocity >2 m/sec. Although three-dimensional TTE may be useful to delineate the size and anatomy of the orifice, TEE is often required for this purpose. TEE also helps determine the relation of the membrane to other cardiac structures, especially to differentiate CTS from supravalvular ring, the latter being located below the LA appendage. Three-dimensional matrix-array TEE can also provide detailed visualization of the orifice or fenestrations of CTS. A retrospective analysis showed a 50% higher detection rate of CTS on preoperative computed tomographic imaging compared with echocardiography. In 80% of patients, complex cardiac anomalies were either missed or misdiagnosed on echocardiography.[6] Thus, echocardiography alone may not be sufficient to describe the anatomic presentation of the defect and an integrative diagnosis may be helpful. Cardiovascular magnetic resonance (CMR) can be helpful in the assessment of hemodynamics, valve disease, and chamber size and function. Sakamoto *et al.*[7] described two cases of CTS in adults in which the use of CMR provided additional anatomic information that was not evident on TTE, particularly regarding the location of atrial communication and anatomy of the pulmonary veins.

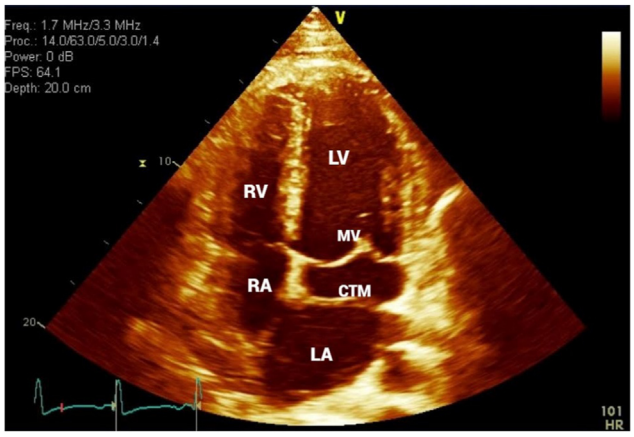

Figure 1 Two-dimensional TTE, apical four-chamber view, systolic phase performed 8 years before the current presentation, demonstrates the cor triatriatum membrane (CTM) dividing the left atrium (LA) into true and accessory chambers. *LV*, Left ventricle; *MV*, mitral valve; *RA*, right atrium; *RV*, right ventricle.

According to the updated 2018 American Heart Association and American College of Cardiology guidelines for the management of adult congenital heart diseases, adults presenting with CTS should be evaluated for other congenital abnormalities, particularly atrial septal defect, ventricular septal defect, and anomalous pulmonary venous connection (Class 1). In adults with previous repair of CTS and recurrent symptoms, they recommend evaluating for pulmonary vein stenosis (Class 2a).[8]

The management of CTS requires a multidisciplinary approach. Early involvement of the congenital heart disease team, electrophysiologists, and the cardiac surgery team will help accurately diagnose and understand the extent of the condition, tailor treatment plan on the basis of age, overall health, and severity of disease, as well as provide ongoing follow-up and monitoring for complications. Cardiopulmonary exercise testing can be useful for baseline functional assessment and serial testing in adult congenital heart diseases (2018 American Heart Association/American College of Cardiology Class 2a recommendation), but there is scarce evidence of its use in patients with CTS.[9] Exercise stress echocardiography can also be useful to determine the functional severity of the membrane.[10]

An incidental echocardiographic finding of LA extra membrane with no pressure gradient in an asymptomatic patient requires monitoring only. When exertional dyspnea and signs of pulmonary congestion occur, diuretics and preload reduction are the mainstays of medical therapy.

Atrial arrhythmias should be adequately treated with ventricular rate control and appropriate anticoagulation, as they may quickly lead to cardiac decompensation. As with other congenital heart diseases, it is not uncommon for patients with CTS to have other complex interatrial arrhythmias aside from atrial fibrillation.[11] When performing catheter ablation for atrial fibrillation, additional electrophysiologic testing (e.g., rapid atrial pacing) should be considered to assess for atrial flutters and atrial tachycardias. Successful management of atrial fibrillation in patients with CTS with catheter ablation is reported, but long-term outcomes have not been reported.[12] However, catheter ablation can be challenging in these patients with an altered LA anatomy. First, when performing transseptal access to the LA, there needs to be specific focus on entering the "accessory atrium" receiving blood from the pulmonary veins. As CTS can

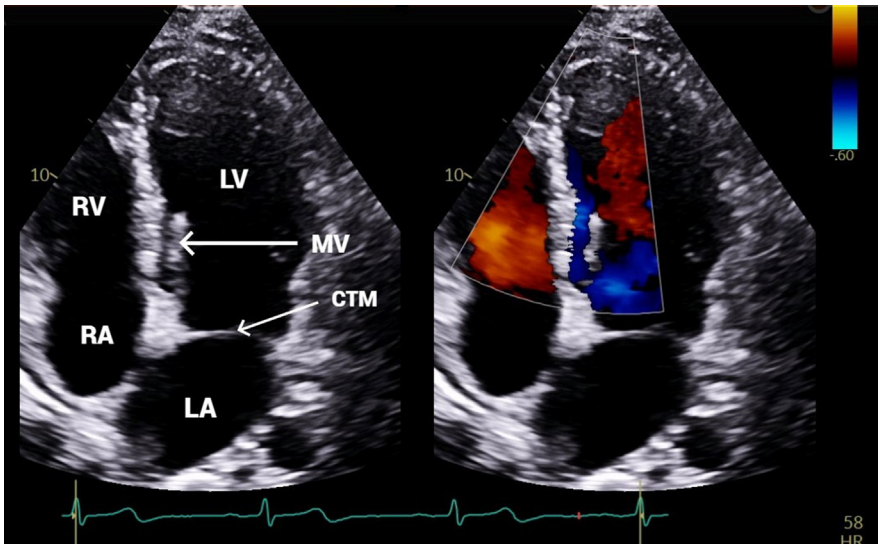

Figure 2 Two-dimensional TTE, apical four-chamber view, without (*left*) and with (*right*) color flow Doppler, diastolic phase, demonstrates the CTM dividing the LA into true and accessory chambers. The left atrial volume index was 52 mL/m^2 as measured using the biplane method.

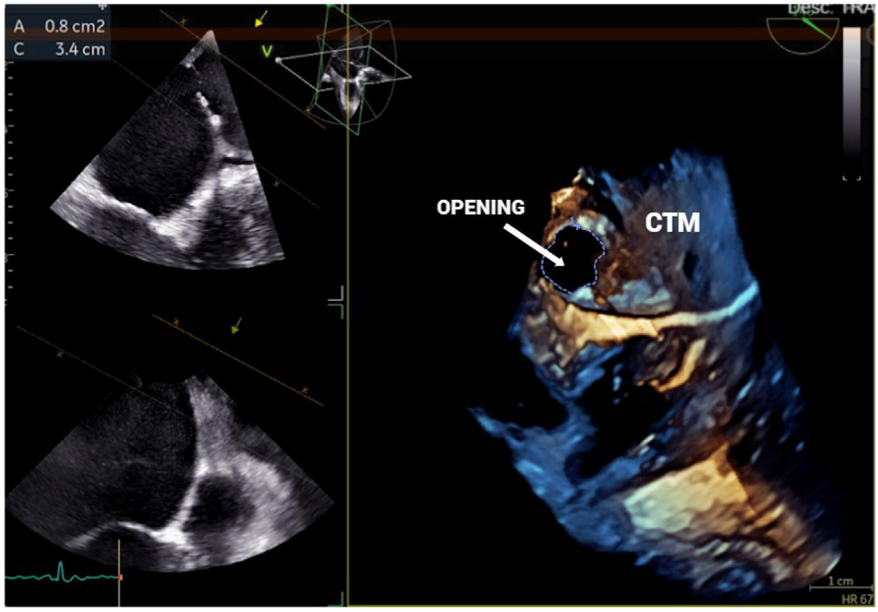

Figure 3 Three-dimensional TEE, midesophageal (*left*) and reconstructed en face (*right*) view, systolic phase, demonstrates the opening through the CTM.

involve the interatrial septum, there may be less interatrial septum available for transeptal access. Second, once access into the "accessory atrium" is achieved, catheter manipulation may be constrained because there is a smaller than usual space in which to perform catheter ablation of the pulmonary veins.[13] In our patient, the CTS membrane was noted to originate from the posterolateral ridge but was mostly out of the way.

According to the 2018 American Heart Association and American College of Cardiology guidelines, surgical repair is indicated for adults with CTS for symptoms attributable to the obstruction or a substantial gradient across the membrane ≥8 mm Hg (mean, 17 mm Hg; range, 8-40 mm Hg; Class 1).[8] Complete resection of the membrane and closure of the atrial septum with a pericardial patch is a common approach. Associated congenital defects need to be corrected at the same time. Surgical outcomes are usually favorable at well-experienced centers, with nearly all patients becoming asymptomatic at follow-up and an overall reported survival 10 years of 83%.[14]

Percutaneous strategies have also been explored as a treatment modality. There are no guidelines for selecting patients for percutaneous treatment. However, Li *et al.*[15] suggested that balloon

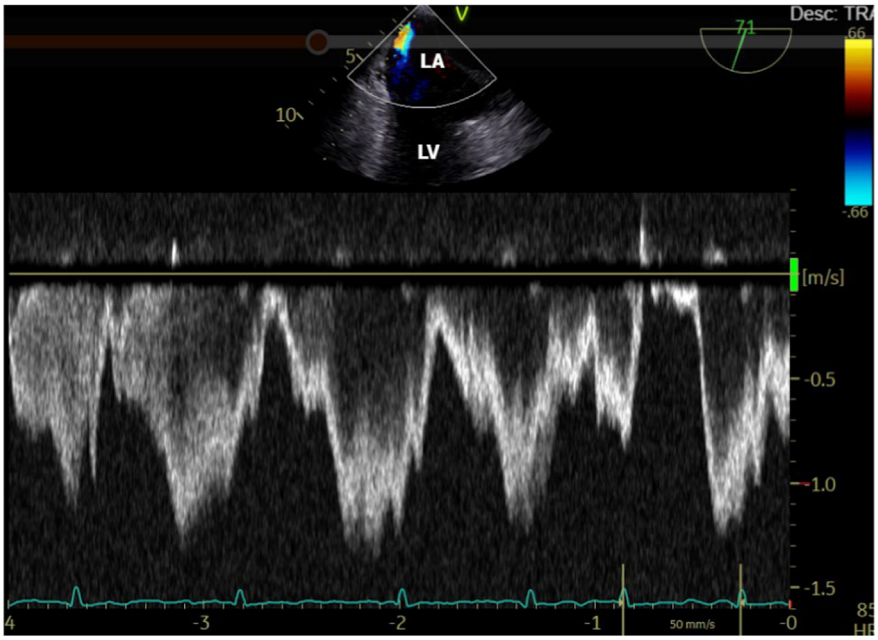

Figure 4 Two-dimensional TEE, midesophageal view, with pulsed-wave Doppler, demonstrates the opening in the CTM.

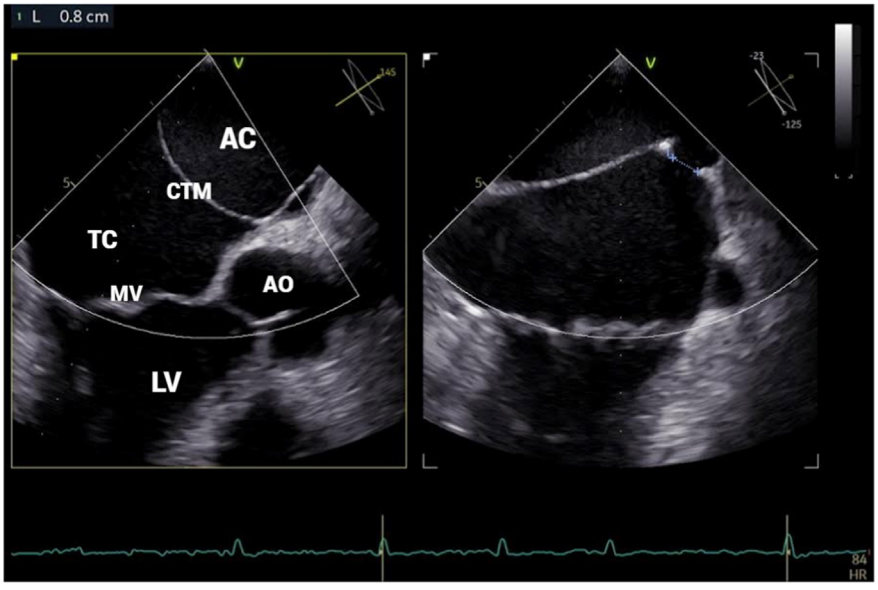

Figure 5 Two-dimensional TEE, midesophageal view of the LA in early systole, demonstrates the CTM and 0.8-mm opening. *AC*, Accessory chamber; *AO*, aorta; *TC*, true chamber.

angioplasty can be considered as a bridge to surgery or as definitive therapy in patients with acute heart failure who are unfit for surgery at the time or symptomatic pregnant patients who have compatible anatomic variants of CTS. They concluded that anatomic characteristics, determined using advanced imaging such as CMR, should be considered to predict the technical success of catheter-based interventional strategies and that percutaneous strategies can be recommended only for isolated forms of CTS in which all pulmonary veins ultimately drain into the LA. Given that the long-term efficacy of percutaneous strategies remains to be confirmed, surgery is still the mainstay of treatment.

CONCLUSION

CTS is an important differential diagnosis in young patients with systolic heart failure, even if they have multiple comorbidities. Early multidisciplinary involvement is important for timely diagnosis and intervention.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

FUNDING STATEMENT

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

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SUPPLEMENTARY DATA

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