

# Real-Time Four-Dimensional Echocardiography in the Diagnosis and Management of Cor Triatriatum



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## INTRODUCTION

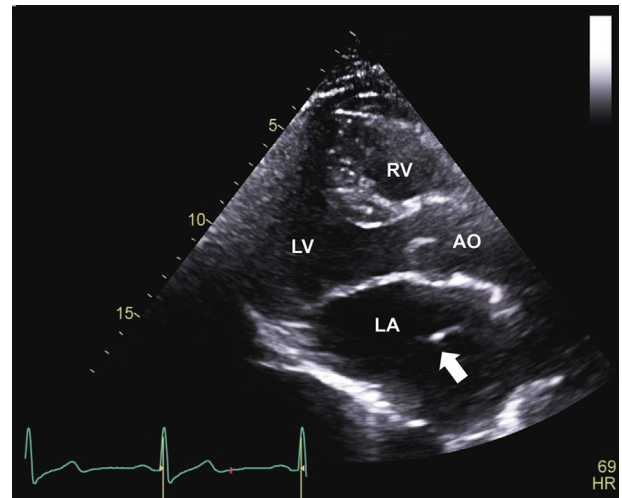
Cor triatriatum is a rare form of congenital heart disease, accounting for <0.1% of all congenital cardiac defects.<sup>1</sup> The first case was reported by W. S. Church<sup>2</sup> in 1868, initially described as a left atrium divided by an abnormal septum. The name “cor triatriatum” was given by Borst<sup>3</sup> in 1905. With the advent of two-dimensional transthoracic echocardiography, cor triatriatum was more easily recognized as a linear membrane that traverses the left atrium.<sup>1</sup>

## CASE PRESENTATION

A 58-year-old man presented to a cardiologist after an episode of dizziness while outside on a hot summer day; while on his lunch break, he felt lightheaded, sweaty, and weak. He noted no frank syncope, and his symptoms were not exertional; they resolved within several minutes without intervention. The results of physical examination were normal. Transthoracic echocardiography was performed to evaluate the etiology of his near syncope, and a linear echodensity was visualized in the left atrium that was suspicious for cor triatriatum (Figure 1, Video 1). Color-flow Doppler demonstrated an area of turbulent flow between the superior and inferior portions of the left atrium (Figure 2). To define this linear echodensity further, transesophageal echocardiography was performed next and confirmed the diagnosis of cor triatriatum sinister with a large fenestration in the center of the membrane and no other smaller communications between the two chambers (Figure 3, Video 2). Using four-dimensional real-time imaging, the insertion point of the membrane in relationship to the left atrial (LA) appendage (LAA) was visualized (Video 3), excluding the diagnosis of supra-ventricular mitral ring and also demonstrating the size and shape of the fenestration. The entire membrane was visualized clearly on four-dimensional real-time echocardiography.

## DISCUSSION

Cor triatriatum is a congenital membrane that divides the left atrium into a superior (proximal) chamber that receives the pulmonary veins and a distal (true) LA chamber that includes the LAA and



**Figure 1** Two-dimensional parasternal long-axis view of linear echodensity in left atrium. Transthoracic echocardiography demonstrates a linear echodensity in the left atrium (arrow). AO, Aorta; LA, left atrium; LV, left ventricle; RV, right ventricle.

the fossa ovalis. The two main objectives of echocardiography in cor triatriatum are (1) to define the abnormality in the left atrium and (2) once the diagnosis is made, to help guide decisions in medical management and surgery. Cor triatriatum has a clinical presentation that can be confused for mitral stenosis, supra-ventricular mitral ring, LA thrombus, or pulmonary vein stenosis. On transthoracic echocardiography, cor triatriatum may be mistaken for a supra-ventricular mitral ring or protruding LAA prominence. This is because these entities have the same linear appearance of a structure dividing the left atrium. LA prominence does not, however, divide the left atrium into two distinct chambers. Supra-ventricular mitral ring is distinguished from cor triatriatum by the insertion point of the membrane in relation to the LAA.<sup>4</sup> In cor triatriatum, the membrane will insert superior to the LAA. Transesophageal echocardiography with four-dimensional real-time imaging played a crucial role in establishing this diagnosis.

The Lam classification has several classes of cor triatriatum on the basis of anatomic features of the membrane and the presence of other congenital abnormalities.<sup>5</sup> In this scheme, our patient is categorized as Lam class A, in which the proximal (upper) chamber receives all pulmonary veins and the distal (lower) chamber includes the LAA and mitral valve; additionally, no atrial septal defect is visualized. In addition to these anatomic descriptions, the membrane also is described by the size and type of fenestration that allows blood flow between the two chambers. The opening(s) may be small, large, multiple, central, or eccentric. Generally, the diameter of the fenestration is <1 cm.<sup>4</sup>

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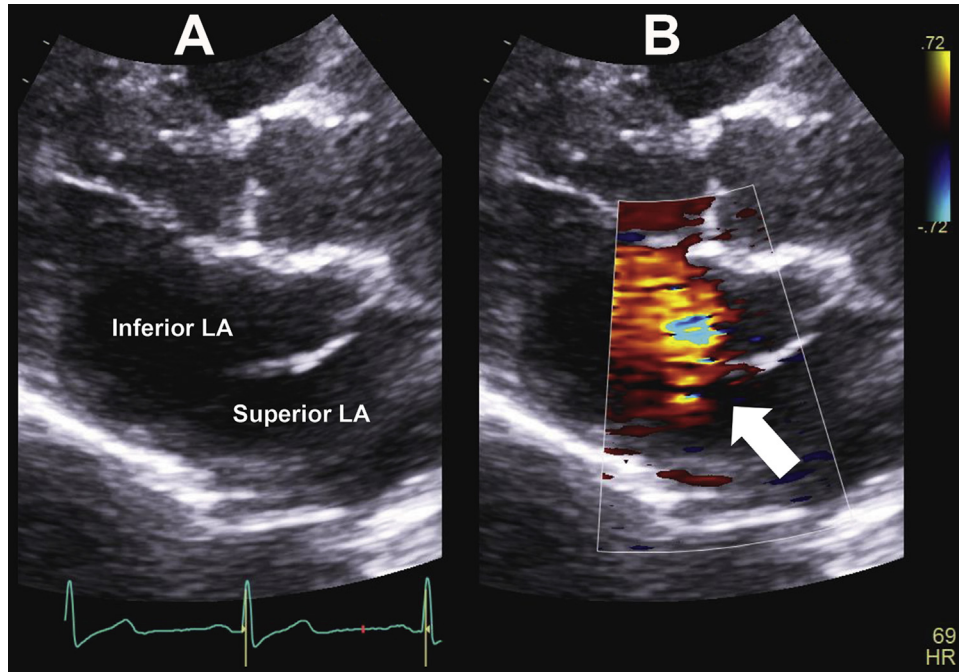
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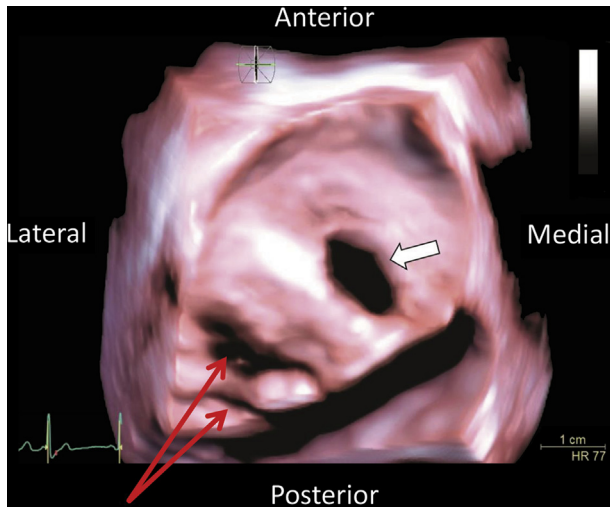
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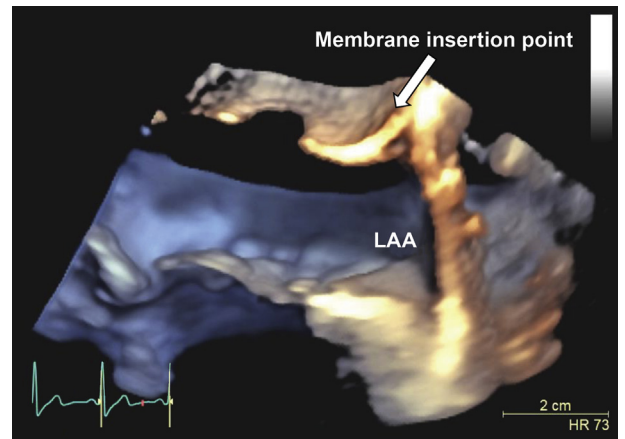
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**Figure 2** Simultaneous transthoracic echocardiography and color-flow Doppler through echodensity in left atrium. (A,B) Color-flow Doppler demonstrates turbulent flow between the superior and inferior portions of the left atrium (arrow). LA, Left atrium.



**Figure 3** Four-dimensional transesophageal echocardiogram of fenestration within the membrane. En face view, from superior LA side. Four-dimensional transesophageal echocardiographic image exhibits a large fenestration in the center of the membrane (arrow). Red arrows indicate artifacts, “echo dropout” from a relatively thin membrane, and do not represent additional fenestrations. LA, Left atrial.

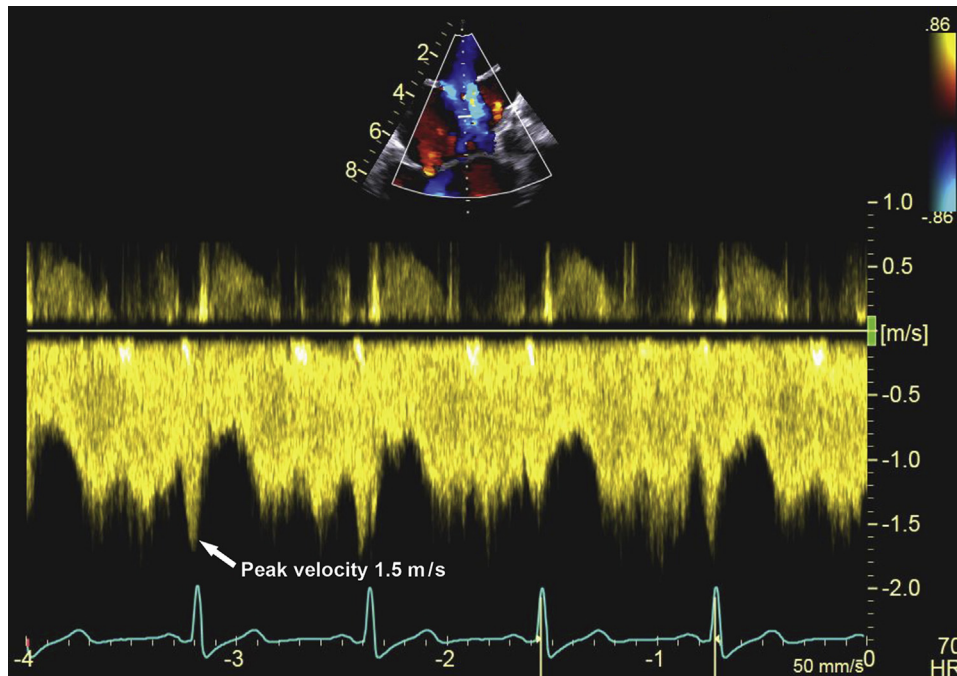


**Figure 4** Four-dimensional transesophageal echocardiographic image of membrane insertion point to LAA. Four-dimensional real-time imaging demonstrates the insertion point of the membrane in relationship to the LAA (arrow). LAA, Left atrial appendage.

The types of fenestrations can be divided into three groups on the basis of the number and size of the fenestrations. The first group is characterized by the absence of connection between the two chambers. Group 2 is defined by one or a few small fenestrations in the membrane, and group 3 is defined by one large fenestration. Group 3 typically is found in adulthood, whereas groups 1 and 2 are usually diagnosed in highly symptomatic infants or children.<sup>1</sup> When a cor triatriatum becomes symptomatic later in life, this could typically be

from fibrosis or calcification of the orifice, development of mitral regurgitation, or the onset of atrial fibrillation with rapid ventricular response.<sup>4</sup> The majority of cases that are diagnosed in adulthood are considered to be mildly obstructive.<sup>6</sup> Treatment typically is for symptomatic individuals and can be done either by surgically excising the membrane or balloon catheter dilatation of the fenestration.<sup>5,6</sup> In patients with obstructive cor triatriatum, surgery provides excellent outcomes with complete resection of the membrane, no residual obstruction, and repair of additional congenital abnormalities.<sup>5</sup>

In our patient, with real-time imaging, we also demonstrated the continuous flow of a cor triatriatum and ruled out significant obstruction (peak velocity 1.5 m/sec; Figures 4 and 5). Right ventricular systolic pressure, assessed from the tricuspid regurgitation signal,



**Figure 5** Continuous-wave Doppler signal through fenestration demonstrating no significant obstruction (peak velocity 1.5 m/sec; arrow).

demonstrated very mild pulmonary hypertension, at 37 mm Hg. With these echocardiographic findings and his clinical symptoms, the patient was diagnosed with vasovagal syncope and continues clinical follow-up for a mildly obstructive cor triatriatum sinister.

## CONCLUSION

Echocardiography is the method of choice in diagnosing cor triatriatum. Four-dimensional real-time imaging with transesophageal echocardiography was fundamental in determining the insertion point of the membrane and confirming the diagnosis of cor triatriatum sinister and also evaluating for other congenital abnormalities. Continuous-wave Doppler demonstrated a mildly obstructive lesion. This is typically the case when diagnosis is made in adulthood.

## ACKNOWLEDGMENTS

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## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.case.2017.04.005>.

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