

## Restrictive Cor Triatriatum in Asymptomatic Adult Patient

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### Introduction

*Cor Triatriatum* (CT), initially described in 1868, it is a congenital malformation resulting from a pulmonary vein resorption failure, common during the formation of the embryonic heart. This genetic error accounts for 0.1% of congenital heart diseases, with incidence of infant death in 75% of untreated symptomatic children. In classical way, it is characterized by the presence of a fibromuscular membrane in the left atrium, dividing this structure into two chambers: a proximal, as an accessory that receives the pulmonary veins, and distal, corresponding to the real left atrium, where the atrial appendage orifice and the mitral valve are<sup>1-5</sup>. Severity and age at which disease symptoms manifest are determined primarily by the number and size of the fenestrations. Patients with fenestrations with a diameter larger than 1 cm are usually asymptomatic<sup>6,7</sup>. Patients with small fenestrations usually develop symptoms resulting from pulmonary hypertension (exertional dyspnea, orthopnea, hemoptysis, etc.) with physiology similar to that of mitral valve stenosis. *Cor triatriatum* is frequently associated with other congenital anomalies such as atrial septal defect, a persistent left superior vena cava, anomalous pulmonary venous return, ventricular septal defect, or even more complex heart defects such as tetralogy of Fallot<sup>8</sup>.

### Case Report

We reported a rare case of an adult patient, RJA, 39 years, with *cor triatriatum* diagnosed in routine examination, prompted by cardiologists to perform physical activities, with orifice of 0.4 cm diameter, without further cardiac changes and asymptomatic, forwarded to the echocardiography service for transthoracic echocardiogram. At doctor's office, after the first evaluation, the patient did not complain; normal physical tests, and electrocardiogram without significant changes. The two-dimensional transthoracic echocardiogram identified a membrane dividing the left atrium at the level of the left superior

pulmonary vein and left atrial appendage, with perforation of approximately 0.4 cm in diameter slightly restrictive, with an average gradient in the 10 mmHg orifice area. There was a slight dilatation of the left atrium without evidence of significant increase in systolic pulmonary pressure. Thereafter, he began to monitor annually at our facility with two-dimensional transesophageal echocardiography (Figures 1 and 2). In the previous exam (August 2013), no evolution was observed in the gradient or the size of the orifice, remaining asymptomatic (maximum gradient of 16.6 mmHg average and 10.1 mmHg) (Figure 3). The moving images can be observed in the Movies 1 to 3.

### Discussion

The echocardiogram is the first-line test for the diagnosis of *cor triatriatum*. However, other methods of image as the angiotomography and cardiac magnetic resonance, for its high spatial resolution to detect changes of cardiac anatomy can also be useful in the diagnosis<sup>9</sup>. Although uncommon, when it is not accompanied by other cardiac malformations, the *cor triatriatum* surgical correction is easy, and the early diagnosis is important in symptomatic cases. Rare in adults, our case was described in a 39-year-old patient and, still asymptomatic, although having an orifice of only 0.4 cm in diameter. In the case described, there was no further structural birth defects. The noninvasive diagnosis can be made by Transthoracic Echocardiography (ETT). The Doppler color flow mapping allows the recognition of perforation(s) between the two chambers, especially when there's pressure gradient between them, as in the present case. With the pulsed Doppler, this gradient can be measured. The Transesophageal Echocardiography (ETE) is indicated if needed for greater precision of anatomic defect for programming surgical corrections or determining the presence of associated injuries, which in this case, were discarded. It is worth mentioning that in this particular case, it is possible that the three-dimensional echocardiography could add important information regarding the anatomy of the defect, as it is not common for a so small orifice to cause symptoms. We should point out that, perhaps, the orifice may show a more complex form, and it cannot be correctly identified by a two-dimensional study.

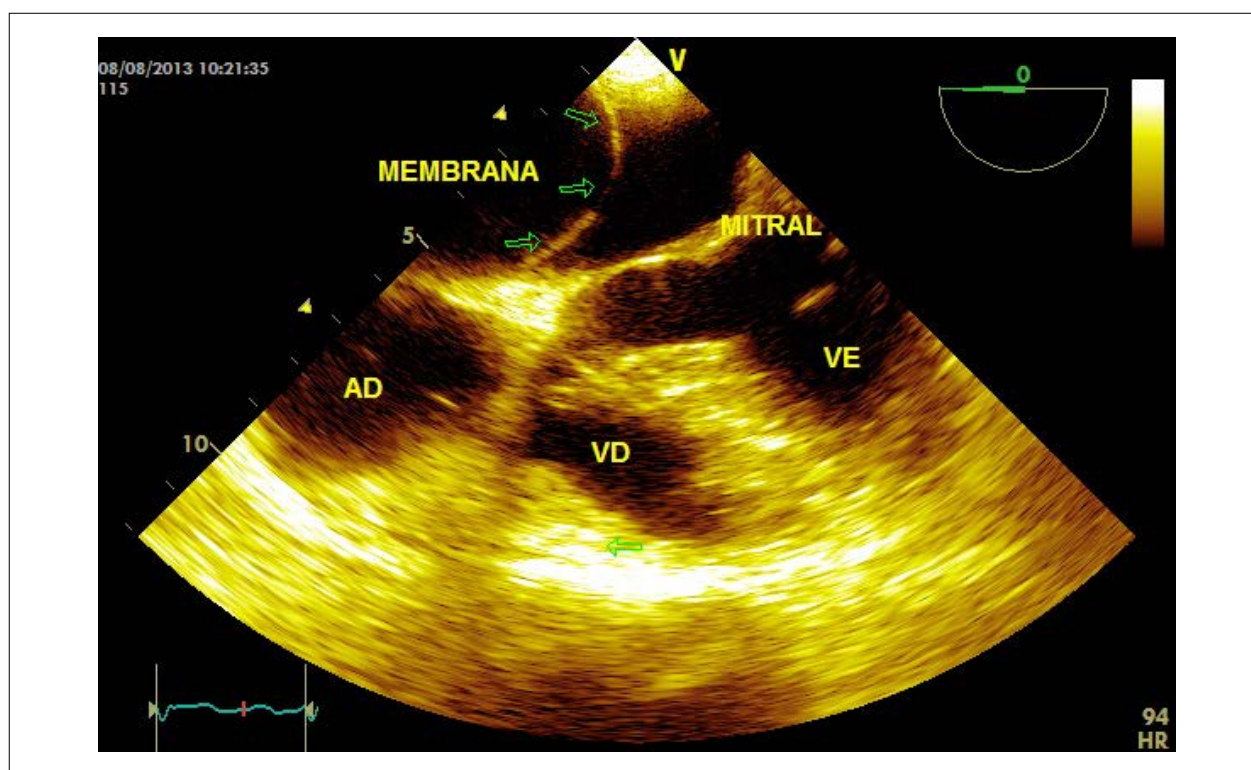
The treatment of *cor triatriatum* in symptomatic patients with gradients greater than 10 mmHg is generally surgical<sup>6</sup>. As our patient was asymptomatic and gradient borderline, we opted for conservative treatment, and he was released for light physical activities under periodic medical supervision.

### Keywords

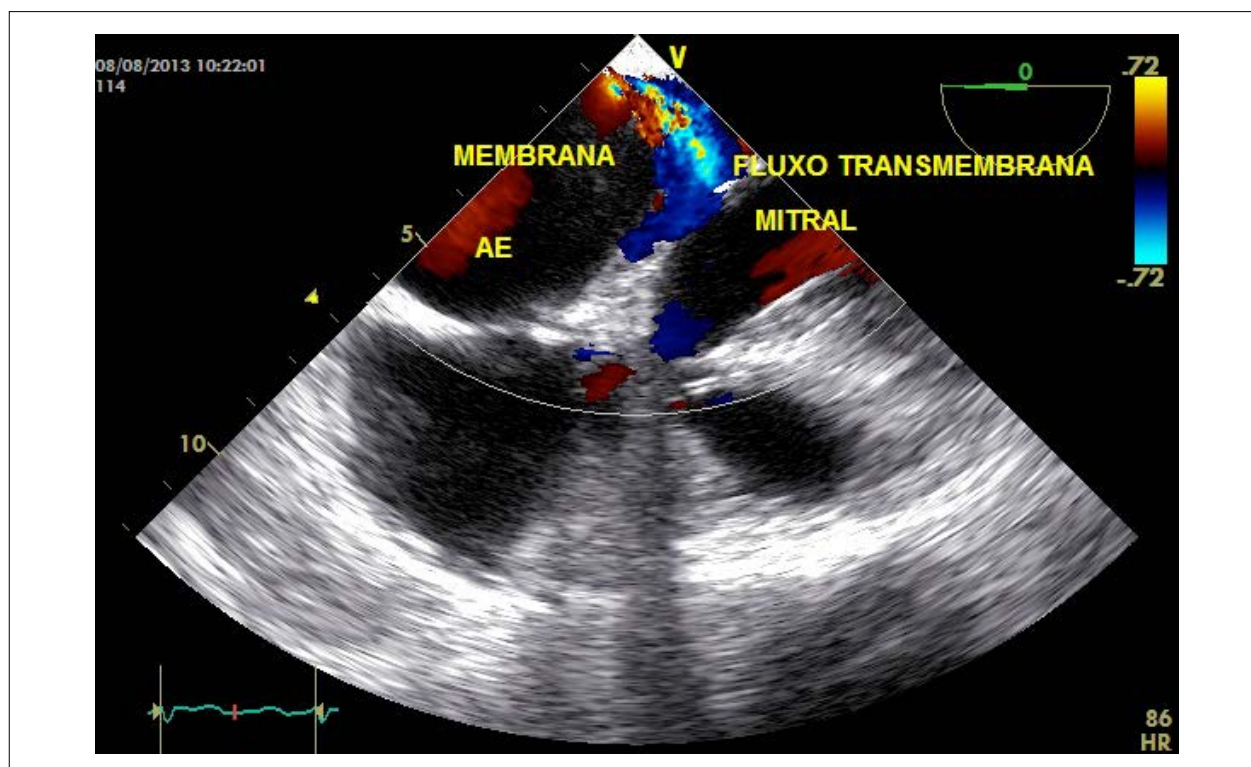
Cor triatriatum; Adult; Heart atria; Echocardiography; Computer-assisted image processing; Atrial septal defect.

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**Figure 1** – Four-chamber cut to the transesophageal echocardiography showing the membrane inside the left atrium. RA = right atrium, RV = right ventricle, LV = left ventricle.



**Figure 2** - Four-chamber cut transesophageal echocardiography with color flow mapping showing the jet through the membrane orifice. LA = left atrium.

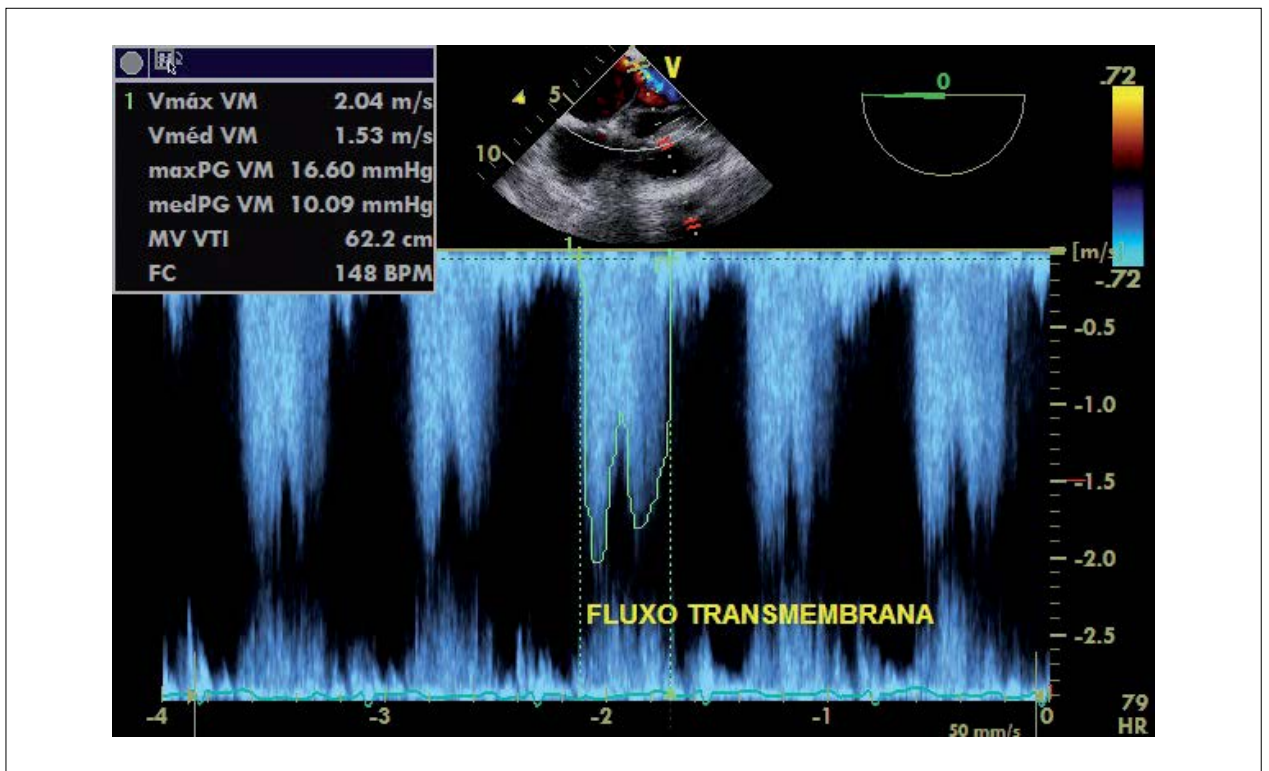
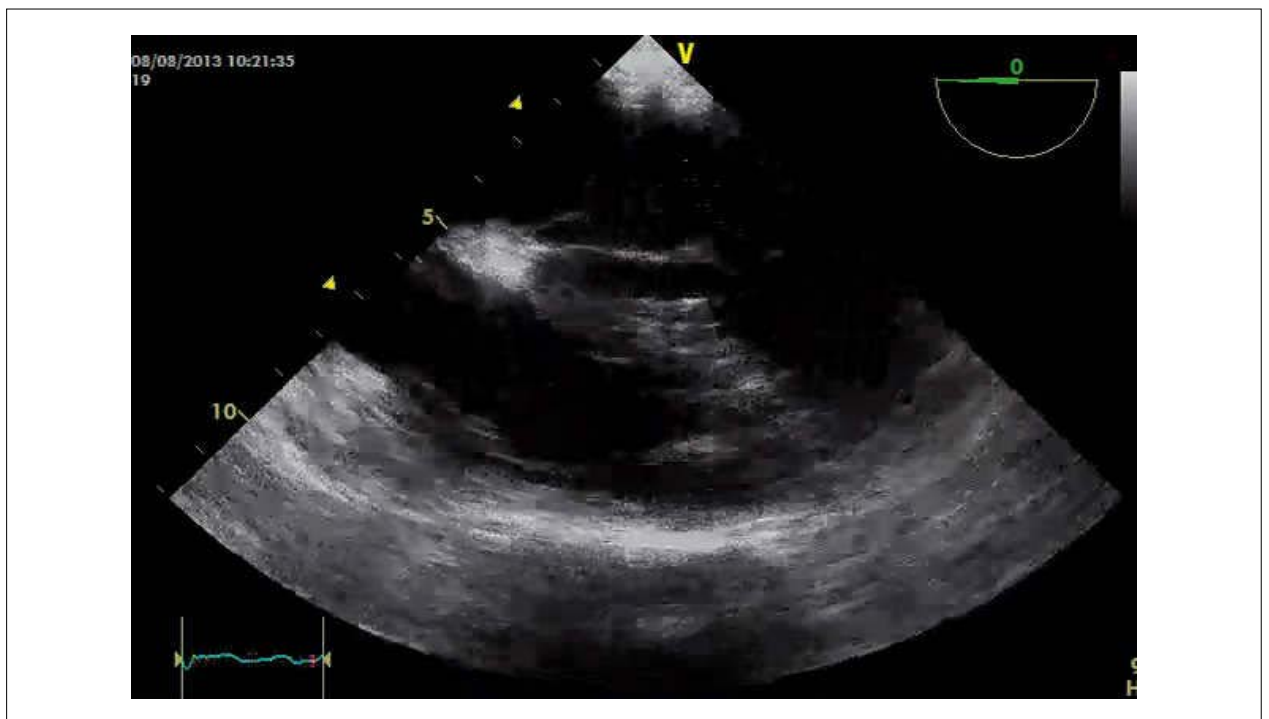


Figure 3 - Study of the transmembrane flux to the pulsed Doppler by quantifying the maximum gradient in 16.6 mmHg and medium in 10.1 mmHg.

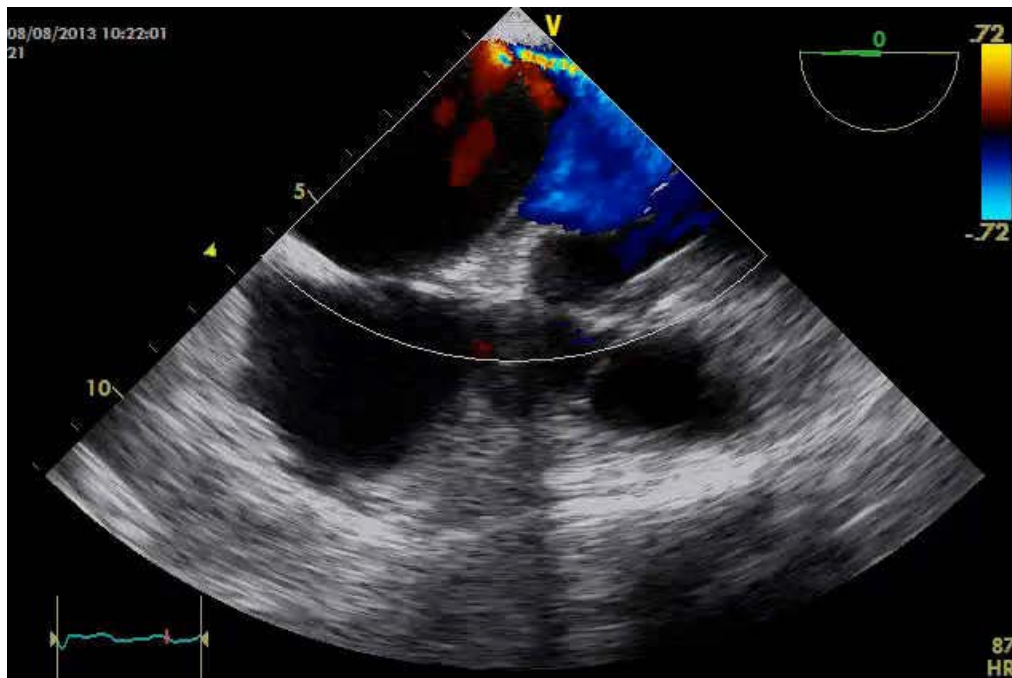


Video 1

## Case Report



Video 2



Video 3

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