Psuedo-Cor Triatriatum in an Elderly Patient With Dyspnea of Exertion: An Undescribed Condition Characterized by 3-Dimensional Transesophageal Echocardiography

Daniel Brooks Levin, MD¹, Gerald Charlton, MD², Richard Snider, MD², and Carlos A. Roldan, MD²

Abstract
A 66-year-old gentleman with no prior cardiac history presented with dyspnea on exertion and chest pain for 1 month. His workup included a transthoracic echocardiogram, which demonstrated findings suggestive of cor triatriatum (C-TAT) with uncertain degree of hemodynamic obstruction. In addition, mild left ventricular systolic dysfunction and segmental wall motion abnormalities suggestive of coronary artery disease were noted. The patient then underwent transesophageal echocardiography (TEE) to define better the structural characteristics and hemodynamic significance of the C-TAT, left and right heart catheterization to assess pressure gradients between the upper and lower left atrial chambers by simultaneous measurement of pulmonary capillary wedge and left ventricular end-diastolic pressures, and coronary angiography. Multiplane 2-dimensional TEE demonstrated an atrial membrane extending from the inferior portion of the interatrial septum to the superior aspect of the lateral atrial wall. This membrane exhibited a medial large oval opening with bidirectional flow and a ≤2 mm Hg gradient. Three-dimensional TEE imaging re-demonstrated this obliquely oriented membrane; however, of most importance, it revealed that the membrane divided the atria into a medial funnel and C-shaped cavity with a large distal oval shape opening and an even larger lateral atrial cavity. These findings were inconsistent with a true C-TAT and rather demonstrated what we defined as a pseudo-C-TAT membrane. Simultaneous right and left heart catheterization confirmed a minimal gradient of 3 to 5 mm Hg and coronary angiography demonstrated severe 3-vessel coronary disease as the primary cause of the patient’s clinical syndrome.

Keywords
cardiology, diagnostic testing, radiology, imaging

Cor triatriatum (C-TAT) is a rare congenital heart defect (0.1% to 0.4% in postmortem series) in which the left or rarely the right atrium is subdivided into 2 atrial chambers by a tissue membrane.¹ Typically, the tissue membrane subdivides the atrium into a more proximal or superior, and a more distal or inferior compartment. It is typically diagnosed in infancy as many of these membranes effect hemodynamics, presenting as a supravalvular mitral stenosis.

In this article, we present the case of an elderly patient with dyspnea on exertion and chest pain who was found to have a left atrial membrane suggestive of C-TAT. This membrane was investigated to define its structural and hemodynamic characteristics as there was concern for it causing the patient’s clinical syndrome.

A 66-year-old gentleman with no relevant prior cardiac history presented with dyspnea on exertion and chest pain. The dyspnea on exertion and chest pain started 10 hours prior to being seen in the emergency department. The pain was described as both dull and sharp and it was mainly right sided on presentation. In the emergency department, the patient had 2 sets of negative troponins, a negative chest X-ray, and a...
negative electrocardiogram. A D-dimer was drawn, which was elevated. Due to chronic kidney disease, the patient underwent a VQ scan that showed intermediate probability of pulmonary embolism and then was admitted to medicine for further diagnostic testing and care.

As part of his workup, the patient underwent a transthoracic echocardiogram (TTE). His TTE demonstrated findings suggestive of C-TAT with an uncertain degree of hemodynamic obstruction (Figure 1A and B) in addition, mild left ventricular systolic dysfunction, and segmental wall motion abnormalities suggestive of underlying coronary artery disease (CAD) were noted. To better determine the hemodynamic significance of patient’s apparent C-TAT and underlying CAD, the patient underwent a transesophageal echocardiogram (TEE) followed by right and left heart catheterization. Multiplane 2-dimensional TEE demonstrated an atrial membrane extending from the inferomedial to the superolateral aspect of the left atrium (LA) with a large medial opening (thin arrows). (E) This TEE 4-chamber view with color Doppler shows flow across the medial opening of membrane (thin arrow). (F and G) These TEE 4- and 2-chamber views with pulsed wave Doppler sampling across the medial opening of the membrane shows bidirectional flow with very low peak velocities up to 0.7 m/s, equivalent to a very low gradient of ≤2 mm Hg ($4 \times [0.7]^2$). (H and I) This TEE views with pulsed wave Doppler demonstrate the entrance of the left (H) and right (I) upper pulmonary veins into the medial aspect of the LA.

LV, left ventricle; LUPV and RUPV, left and right upper pulmonary veins, respectively.
membrane. Therefore, the patient underwent treatment for pulmonary embolism and CAD, while further testing occurred to further delineate a diagnosis. In the cardiac catheterization laboratory, simultaneous recording of the mean pulmonary capillary wedge pressure of 10 mm Hg and left ventricular end-diastolic pressure of 5 to 7 mm Hg (Figure 2E) verified a minimal differential gradient of 3 to 5 mm Hg. As this gradient was considered either physiologically normal or minimally elevated, it was determined that this membrane was not obstructive and therefore not contributing to the patient’s symptoms. Coronary angiography demonstrated severe 3 vessel CAD as the primary cause of patient’s clinical syndrome.

Due to the patient’s possible pulmonary embolism, he was treated with oral anticoagulation for a minimum of 3 months, after which the patient underwent uncomplicated coronary bypass surgery with left internal mammary artery to the left anterior descending artery and saphenous vein bypass grafts to 2 obtuse marginal branches and his right coronary artery. The patient was discharged home as he symptomatically and clinically improved. Expectedly, no intervention is currently planned for the patient’s atrial membrane mimicking C-TAT.

Cor triatriatum is a rare congenital heart defect that typically presents earlier in life with symptomatic heart failure. It has been rarely reported in the adult and elderly population. It is surmised that this patient had a curvilinear membrane extending from the inferomedial to the superior aspects of the atria dividing the atrium into a left or lateral and right or medial chambers with a large distal opening in the medial chamber and therefore leading to a physiologic to minimally elevated gradient of no hemodynamic consequences. These findings are inconsistent with a true C-TAT. To our knowledge, this is the first case of a pseudo C-TAT detected and best characterized by 3D TEE imaging. Therefore, adult or elderly patients with cardiovascular symptoms and an atrial membrane suggestive of C-TAT on TTE should undergo 3D TEE imaging to better define the anatomy and hemodynamic consequences of the membrane. This would better classify patients as having a true versus pseudo-C-TAT.

**Authors’ Note**

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Ethics Approval
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Informed Consent
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