One Heart, Five Chambers, Sixty Years: A Rare Presentation of Cor Triatriatum in an Elderly Patient

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Introduction

Cor triatriatum sinistrum (CT) is a rare congenital abnormality of the heart (0.1-0.4% incidence), characterized by a membrane separating the left atrium into postero-superior and antero-inferior chambers, receiving pulmonary veins and holding the mitral apparatus respectively. Over 75% of patients die within the first year of life and survivors are usually detected before middle-age.

Case Report

A 62 year old veteran with a history of coronary artery disease with multiple stents, atrial fibrillation on chronic anticoagulation and history of ASD that was repaired at age 9 presented to us with shortness of breath and chest pain. Initial exam was significant for an irregularly irregular rhythm and a flow murmur. Patient underwent serial trans-thoracic and trans-esophageal echocardiograms over 5 years revealing a class 3 CT with a 2.5cm orifice and a moderate mitral and tricuspid regurgitation. There was a progressive increase in left atrial size (5.5 to 7.2cm) and pulmonary pressures (35 to 45mmHg) over 5 years of follow up. A chest xray and computed tomography scan showed cardiomegaly with features of left atrial enlargement and compensated congestion.

Patient was managed entirely medically for his above problems and is currently asymptomatic with stable disease. Echocardiographic images are presented (figure 1).

Discussion

Almost all patients with symptomatic CT with a narrow orifice (< 1cm) are managed surgically or with percutaneous balloon correction if co-morbidities preclude surgery (1). However, as in this case, in an elderly patient with a wide orifice between chambers, symptomatic medical management is a suitable option.

After a comprehensive literature review, the most common presenting feature of CT was found to be a misdiagnosis as mitral stenosis (2) or as an incidental finding (3). Other less common presenting features include strokes/TIAs (4) , hypotension (5), shortness of breath (6) and pulmonary disease (7) . Apart from an ASD (8), common associations of CT include patent foramen ovale (9), tetrology of fallot (10) and other valvular abnormalities (11). Common complications include atrial fibrillation (12), pulmonary hypertension (13) and thrombosis (14). The most commonly used tool for diagnosis is a trans-thoracic echocardiogram though three dimensional echocardiography and magnetic resonance imaging are more sensitive.(15)

Conclusions

This case is unique in multiple ways; firstly, the patient is in his seventh decade of life with CT which has not been reported prior to this in an American population. Secondly, the patient presented with heart failure which is unusual in CT and thirdly, the patient was managed medically throughout the course of the disease and was followed up with serial echocardiograms over a period of 5 years. Evidently, medical management of CT can be considered by clinicians depending on symptom control and age at presentation for patients with large orifices.

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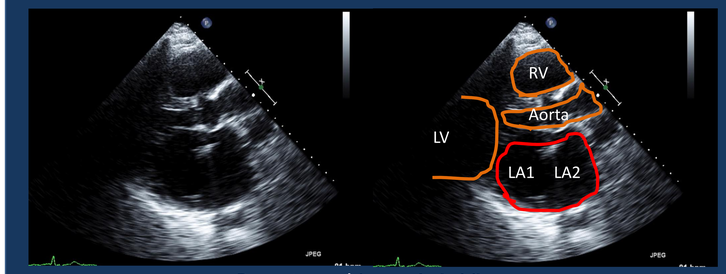
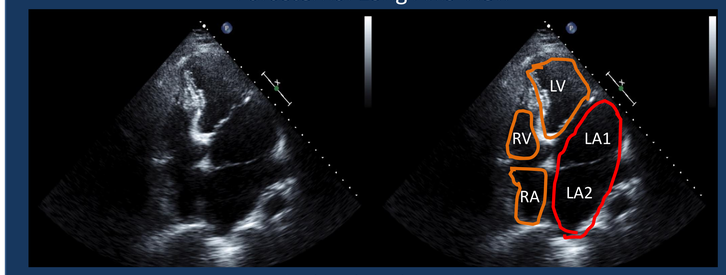
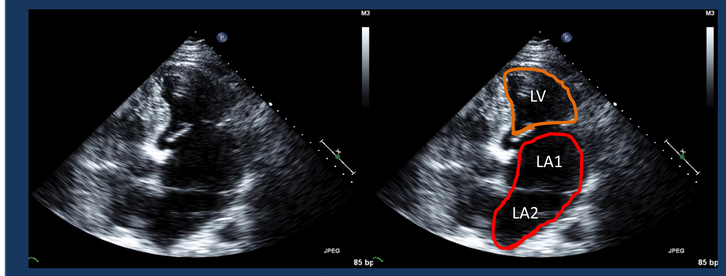


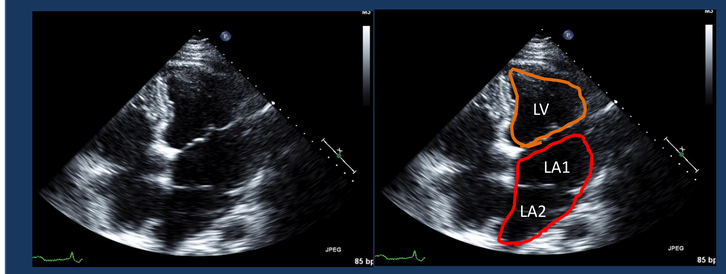
Parasternal Long Axis View



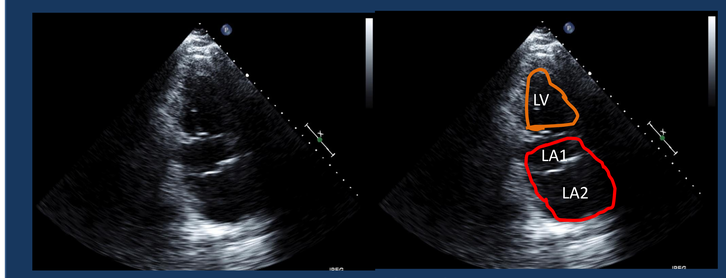
Apical 4 chamber view



Valves opening



valves closing



Valves closed

Doppler