

# Cor Triatriatum Sinistrum With Partial Atrioventricular Septal Defect In An Adult Nigerian: A Case Report

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## Abstract

Cor triatriatum is a congenital anomaly in which the left atrium (cor triatriatum sinistrum) or right atrium (cor triatriatum dextrum) is divided into 2 parts by a fold of tissue, a membrane or a fibromuscular band.<sup>1</sup> It is a rare cardiac malformation comprising about 0.1 to 0.4% of congenital heart disease.<sup>3</sup> Its presentation is either classical or atypical.<sup>4</sup> We present a case of an atypical cor triatriatum sinistrum with partial atrioventricular septal defect.

## CASE REPORT

A 35 year old nurse presented with cough, dyspnea, and leg swelling of 5 days duration. Dyspnea was initially on moderate exertion but progressed to being at rest with associated paroxysmal nocturnal dyspnea, orthopnea, and effort intolerance. Cough was productive of mucoid sputum without hemoptysis but she admitted to intermittent low grade fever. The patient was told she had a congenital heart disease 4 years earlier when a chest radiograph was done during pre-employment evaluation that showed cardiomegaly and a follow up echo detected a 'hole in her heart'. The patient has had recurrent episodes of cough in the past which responded to treatment with antibiotics. Examination revealed a young lady in respiratory distress, centrally cyanosed with grade 3 finger clubbing, and pitting leg edema. She had tachycardia of 120 bpm, blood pressure of 110/80 mmHg, distended neck veins, and a displaced and diffuse apex. There was left parasternal heave, loud P2 and a grade 3 pansystolic murmur radiating to the axilla. She also had a tender hepatomegaly and fine bibasal crepitations at the lung bases.

A chest radiograph showed massive cardiomegaly with prominent upper lobe vessels. (Fig. 1) Her electrocardiogram showed sinus tachycardia, right axis deviation, right atrial enlargement, right bundle branch block and right ventricular hypertrophy. (Fig. 2) An echocardiogram done showed a markedly dilated right atrium and a ventricle with a huge atrial secundum defect measuring about 6.55 cm. (Fig. 4)

The mitral valve was dysplastic and had a poorly developed subvalvular apparatus and the left atrium was noted to have a band extending from the atrioventricular junction to the lateral wall. This band divided the left atrium into 2 chambers- a superior and an inferior chamber. (Fig. 3) The pulmonary outflow tract and inferior vena cava were dilated and tricuspid regurgitation was noted with an estimated RVSP of 70.6 mmHg. (Fig. 5 and 6) We made the diagnosis of cor triatriatum with partial atrioventricular septal defect and severe pulmonary hypertension in heart failure. She was commenced on anti- heart failure medication as well as sildenafil citrate for the severe pulmonary hypertension. She recovered and was discharged home and asked to continue her follow-up in the clinic.

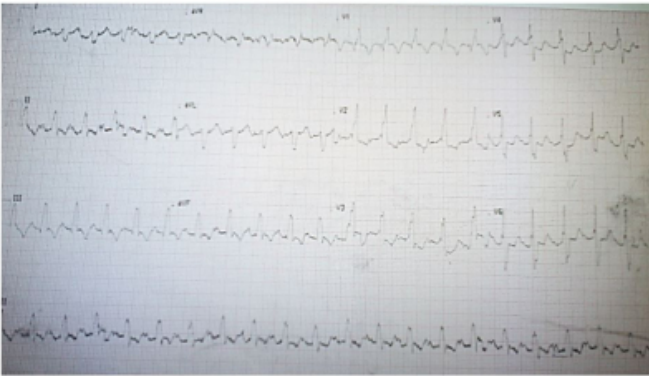
**Figure 1**

Chest radiograph showing massive cardiomegaly.



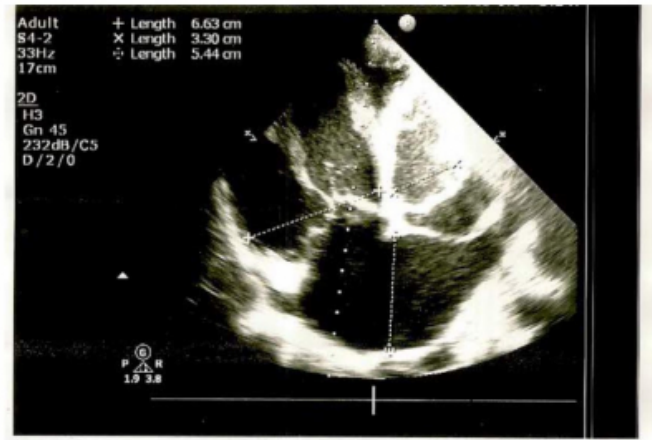
**Figure 2**

Electrocardiogram showing right axis deviation, right ventricular hypertrophy and right bundle branch block.



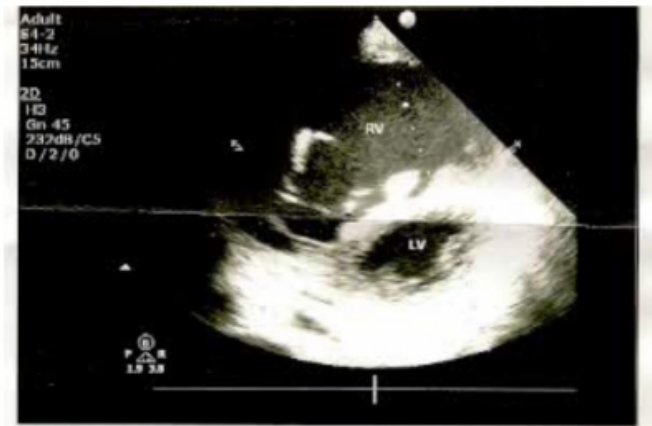
**Figure 3**

2 dimensional transthoracic apical 4-chamber view showing the atrial septal defect, the fibrous band and the dilated right atrium and ventricle.



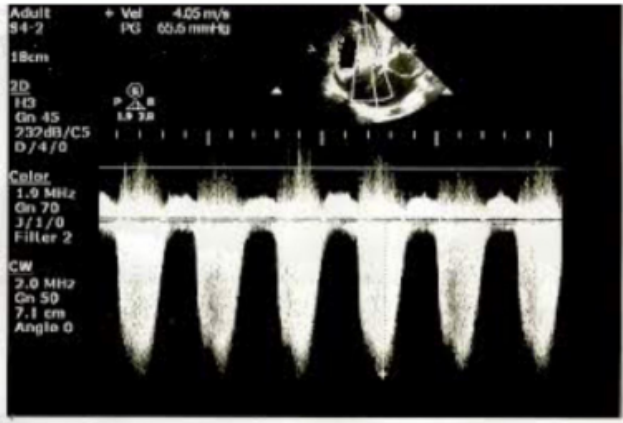
**Figure 4**

2 dimensional parasternal short axis view showing the dilated right ventricle (RV) relative to the left ventricle (LV).



**Figure 5**

Continuous wave Doppler across the tricuspid valve showing the severe tricuspid regurgitation.



**Figure 6**

2 dimensional subcostal view showing dilated inferior vena cava.



## LITERATURE REVIEW

Cor triatriatum (or triatrial heart) is a congenital heart defect where the left (cor triatriatum sinistrum) or right atrium (cor triatriatum dextrum) is subdivided by a thin membrane, resulting in three atrial chambers. Cor triatriatum is a rare congenital anomaly with a ratio of men to women of 1.5:1. *This condition is a result of failure of embryologic common pulmonary vein incorporation into the back of the left atrium. As a result, the left atrium is divided into two chambers by a fibromuscular membrane. The two cavities positioned posterior superior and anterior inferior, are anatomically and functionally separated.*<sup>[ii]</sup> Cor triatriatum (CT) represents 0.1-0.4% of all congenital cardiac malformations and usually refers to the left atrium.<sup>[iii]</sup> This entity can present in classical or atypical form. While the classical form is an isolated thin membrane within the left

atrium, the atypical form is associated with other cardiac anomalies.<sup>[iv]</sup> Association of CT and atrioventricular septal defect (AVSD) is rare.<sup>[v]</sup> Patients with AVSD are prone to develop pulmonary hypertension and often lead to inoperable pulmonary vascular occlusive disease. Pulmonary venous obstruction with CT, which is also rare, is a correctable cause of pulmonary arterial hypertension.<sup>[vi]</sup> Other cardiac anomalies that accompany CT are ventricular septal defect, coarctation of the aorta, tetralogy of Fallot and mitral stenosis. Very few cases have been reported in sub-Saharan Africa such the case by Oyediji et al.<sup>[vii]</sup> Extracardiac manifestations that may be observed with CT include asplenia and polysplenia.<sup>[viii]</sup> Depending on the severity of the obstruction, CT may be symptomatic or asymptomatic. Symptoms are a result of outflow obstruction and include dyspnea, orthopnea, cyanosis, hemoptysis, and chest discomfort. When the obstruction is severe CT is usually diagnosed during infancy or adulthood. Some cases may remain asymptomatic or minimally symptomatic at diagnosis.<sup>[ix]</sup> Our patient has been asymptomatic until now and the earlier diagnosis of congenital heart disease was from a chance finding during pre-employment screening where cardiomegaly was found on her chest radiograph necessitating further investigations. Electrocardiography is often normal in CT patients especially when obstruction is not present. When there is significant obstruction, electrocardiogram may show signs of right ventricular hypertrophy, such as right axis deviation and prominent R or R' wave in VI, which may develop due to associated pulmonary hypertension as was present in our patient. In adults, CT might present with atrial fibrillation, and removal of the membrane could abolish the arrhythmia.<sup>[x]</sup> Other reported arrhythmias include atrial tachycardia and ectopic atrial rhythm with abnormal P waves.<sup>9</sup> Radiologically, the heart is moderately enlarged and the lungs may reveal some degree of congestion with fluid. The diagnosis is usually confirmed by echocardiography. M-mode and 2-dimensional echocardiography demonstrates a membrane in the mid left atrium while Doppler echocardiography is useful to assess the degree of flow obstruction.<sup>[xi]</sup> Additional modalities that could be used for diagnosis of CT include cardiac CT and cardiac MRI. Both techniques allow a complete anatomic evaluation of the lesion when echocardiographic assessment is unavailable or inadequate.<sup>[xii]</sup>

Surgical removal of the fibromuscular membrane abolishes obstructive symptoms in patients especially in those in which the membrane is complete and the proximal chamber

communicates with the right atrium through an atrial septal defect.[xiii] We decided to manage our patient conservatively with sildenafil citrate because of the severe pulmonary hypertension. The only therapeutic option for her is lung transplantation with surgical correction of the heart defect that however is associated with very high operative risk.

## CONCLUSION

Cor triatriatum sinistrum, although extremely rare, may remain asymptomatic into adult life. It is an important entity to recognize because it may be easily surgically corrected when hemodynamically significant. Echocardiography is the best imaging modality in diagnosis but cardiac CT and cardiac MRI could also help for a more complete anatomical evaluation of the lesion.

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