Cor Triatriatum Sinister Diagnosed In An Elderly Male During Work-Up For New-Onset Atrial Fibrillation
A Doku, D Ahadzi, A Yakubu, J Akamah

INTRODUCTION
A diagnosis of a congenital heart anomaly in adulthood may occur as an incidental finding on cardiac imaging with no hemodynamic consequences to the patient. On the other hand, these lesions may be revealed as the cause of new-onset cardiac symptoms in adulthood. Cor triatriatum sinister is a rare congenital heart anomaly resulting in a septation in the left atrium[1]. It is rarely diagnosed in adults. Pathophysiologically, this lesion can mimic mitral stenosis in instances where the opening in the septation is significantly narrowed, resulting in obstruction to proximal intra-atrial flow. Symptomatic adults may present with atrial fibrillation and cardioembolic phenomena as seen in our patient[2]. This case demonstrates the unmasking of an unusual and possibly curable cause of atrial fibrillation using echocardiography.

CASE PRESENTATION
A 61-year-old male with a recent diagnosis of hypertension a month prior reported with sudden intense numbness in his right arm with associated inability to speak. He reported for evaluation soon after the onset of his symptoms. However, by the time he reported, his symptoms had already begun to resolve. He noted progressive improvement and subsequent complete resolution of his symptoms within 72 hours. He had however observed new-onset exertional dyspnea and palpitations around this index event with no significant limitation in his activities of daily living. He reported compliance on his antihypertensives (oral Amlodipine 10mg daily and oral Lisinopril 10mg daily) with adequate control of his blood pressures. Physical examination was remarkable for an irregularly irregular heart rate of 114 beats per minute. An electrocardiogram (ECG) showed atrial fibrillation with a rapid ventricular response. An emergent plain head computed tomography (CT) scan done at the time of presentation was normal.

A transthoracic echocardiogram (TTE) revealed a horizontal septation, dividing the left atrium into a superior chamber into which the pulmonary veins drained, and an inferior chamber abutting the mitral valve and left atrial appendage. A transoesophageal echocardiogram revealed similar findings (Figure 1) with turbulent flow across an ostium in the septation on Colour Doppler interrogation. The peak velocity of flow across the ostium was 2.4m/s with a peak pressure gradient of 24mmHg (Figure 2). The ostium in the septation on transesophageal echocardiography (TEE) measured approximately 1.3cm in diameter (Figure 3). Colour Doppler revealed obstruction to the forward flow of blood from the superior to the inferior chamber mimicking...
mitral stenosis. The jet was directed toward the anterior mitral valve leaflet causing mild deformity of the anterior leaflet. There were no other structural abnormalities on echocardiographic imaging.

**Figure 1**
TEE showing septation of the left atrium (LA) by a membrane (arrow). The left atrial appendage (LAA) has no visualized clot and opens into the outer chamber of the left atrium that has the mitral orifice (star). LV=left ventricle

**Figure 2**
TTE. Apical 4 chamber view showing inlet flow jet across the membrane (arrow). LV=left ventricle

The patient was recently diagnosed with hypertension. This put him at risk for structural heart changes which could be responsible for his atrial fibrillation. However, given the absence of significant left ventricular hypertrophy and the presence of left atrial dilatation limited to the accessory chamber, we theorized that the cause of the patient’s atrial fibrillation was likely due to the pathological changes caused by the presence of the abnormal septation in the left atrium. He was managed for Cor Triatriatum Sinister complicated by atrial fibrillation, cardioembolic stroke and hypertension. Beta-blocker therapy was added on to his antihypertensive therapy (oral bisoprolol 2.5mg daily) for rate control. Anticoagulation was initiated with Oral Warfarin 5mg and the dose was titrated to maintain his INR in a therapeutic range of 2 - 3. On account of his cardioembolic phenomena, persistent atrial fibrillation and mild exertional dyspnea the patient was counseled on surgical excision of the abnormal left atrial septation as a therapeutic option. He however declined a surgical intervention and opted for a conservative management strategy. Over a period of approximately two years, the patient has remained in atrial fibrillation with adequate rate control. He has achieved optimal anticoagulation with warfarin with no recurrence of clinically overt thromboembolic phenomena. He still has mild exertional dyspnea with no significant limitation of his activities of daily living.

**DISCUSSION**
Cor triatriatum sinister is a rare congenital heart anomaly with an estimated incidence of 0.1% of all congenital heart
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3. Thakrar A, Shapiro MD, Jassal DS, Neilan TG, King MEE, Abbara S. Cor triatriatum: The utility of cardiac imaging modalities, the diagnosis of cor triatriatum sinister may become more common. The clinical presentation however varies and largely depends on the presence and size of an ostium in the abnormal septation. In asymptomatic or minimally symptomatic patients a conservative approach may be acceptable whereas surgical excision should be considered in symptomatic patients with documented complications resulting from the lesion.

CONCLUSION

With the advent of widespread utilization of highly sensitive cardiac imaging modalities, the diagnosis of cor triatriatum sinister may become more common. The clinical presentation however varies and largely depends on the presence and size of an ostium in the abnormal septation. In asymptomatic or minimally symptomatic patients a conservative approach may be acceptable whereas surgical excision should be considered in symptomatic patients with documented complications resulting from the lesion.

References

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Author Information

Alfred Doku
Department of Medicine and Therapeutics, University of Ghana Medical School, Korle-Bu Teaching Hospital; Department of Medicine, Korle-Bu Teaching Hospital
Accra, Ghana

Dzifa Ahadzi, MD
Department of Medicine, Korle-Bu Teaching Hospital; Tamale Teaching Hospital
Accra, Ghana; Tamale, Ghana

Abdul-Subur Yakubu
Department of Medicine, Korle-Bu Teaching Hospital; Tamale Teaching Hospital
Accra, Ghana; Tamale, Ghana

Joseph Akamah
Department of Medicine and Therapeutics, University of Ghana Medical School, Korle-Bu Teaching Hospital; Department of Medicine, Korle-Bu Teaching Hospital
Accra, Ghana