

## A Rare Case of Cor Triatriatum Dexter with Secundum Atrial Septal Defect in a 73-Year-Old Patient who Presented with Acute Decompensated Right Heart Failure and Atrial Fibrillation

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### Abstract

Cor triatriatum is a rare congenital cardiac anomaly, in which the left or right atria are subdivided into three chambers and represents about 0.1% of all congenital heart diseases. There are 2 types: Cor Triatriatum Sinistrum which involves the left atrium, and the even rarer Cor Triatriatum Dexter which involves the right atrium. This rare anomaly is seen in young children and is extremely rare in the elderly population. We report a rare case of Cor Triatriatum Dexter with Secundum Atrial Septal Defect in a 73-year-old male who presented with signs and symptoms of Acute Decompensated Right Heart Failure and Persistent Atrial Fibrillation as his initial presentation of the condition to a medical facility. To our knowledge and per the literature review, this is a rare presentation in an elderly individual.

**Keywords:** Case Report; Cor Triatriatum Dexter; Secundum Atrial Septal Defect; Right Heart Failure; Transesophageal Echocardiography; Percutaneous ASD Closure

### Introduction

Cor Triatriatum Dexter is a very rare cause of cyanotic congenital heart disease, with a reported incidence of 0.025% (25 in 100000 cases) [1]. Its incidence in elderly adults is an extremely rare presentation. It is caused by the persistence of the right valve of the sinus venosus. In this condition, the right atrium is divided into two chambers by a thin membrane forming a triatrial heart, hence the name. Typically, the right atrial partition is due to exaggerated fetal Eustachian and thebesian valves, which together form an incomplete septum across the lower part of the atrium. This septum may range from a reticulum to a substantial sheet of tissue [2,3].

### Study setting

This case has been reported at the Emergency Department (ED) of Mediclinic City Hospital (MCH) situated in Dubai Health Care City (DHCC), Dubai, United Arab Emirates. MCH is a 230 bedded multidisciplinary tertiary care hospital and a Level 1 Cardiac Centre in the region. We see around 45,000 patients per year in our ED, of which 2400 patients present with cardiac symptoms. On average, the Cardiac Catheterization Lab carries out 300 Coronary Angioplasty procedures per year.

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## Case Report

A 73-year-old gentleman presented to the Cardiology Out-Patient Department with a 2-week history of progressive shortness of breath, bilateral lower limb swelling, intermittent palpitations, and abdominal discomfort. He also had a mild cough. All these symptoms started 2 weeks after a long-haul air travel. He was a frequent flier but had never experienced these symptoms before. He is usually physically active and goes for regular walks without any symptoms. No significant past medical or surgical history. No significant family history of note.

His initial cardiovascular assessment confirmed an irregular heart rate of 114 beats per minute, Blood Pressure of 134/88 mm Hg, Oxygen saturation of 100% in room air. The patient had clinical evidence of decompensated right heart failure and atrial fibrillation with a soft apical systolic murmur.

His resting ECG confirmed fast atrial fibrillation with a ventricular rate of 118. There were no signs of acute ischemic changes seen on the ECG. Troponin I was 0.017 ng/mL (< 0.034). D-Dimer was 2.7 ug/mL FEU (age-adjusted cut-off is < 0.73). B-Type Natriuretic Peptide (BNP) was 2236.30 pg/mL (< 100). Sodium 114 mM (136 - 145), Chloride 79 mM (98 - 107), Bicarbonate 13 mM (23 - 31), Potassium 4.4 mM (3.5 - 5.1). Total Bilirubin was elevated at 41.6 μM (3.4 - 20.5) with a Direct Bilirubin of 21.8 μM (< 8.8), Total Serum Protein was 61 g/L (64 - 83) with a normal Albumin of 35 g/L (35 - 48). Urea and creatinine were within normal limits. Full Blood Count (FBC) and Thyroid Function Tests were normal.

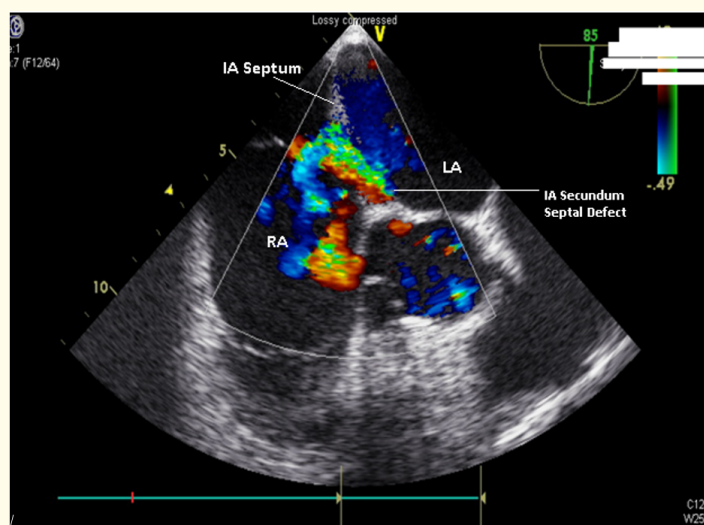
Chest X-Ray confirmed cardiomegaly and evidence of a moderate bilateral pleural effusion with collapse/consolidation in both lower lobes.

A duplex scan of both extremities did not reveal any evidence of deep vein thrombosis.

Transthoracic Echocardiogram confirmed a significantly dilated right heart with poor right ventricular systolic function, severe pulmonary hypertension with an estimated Pulmonary Artery Systolic Pressure (PASP) of 95 mmHg. Paradoxical septal motion abnormality was noted with a left to right shunt suggestive of an Atrial Septal Defect (ASD). Left Ventricular Systolic function was well preserved with normal left ventricular size.

Computed Tomography (CT) Chest with contrast showed no evidence of Pulmonary Embolism, however it revealed a large interatrial septal defect with a left to right shunt.

Given the above-mentioned findings, a Trans Esophageal Echocardiogram (TEE) was performed. This confirmed the presence of a Secundum Atrial Septal Defect in the presence of an adjacent septum in the right atrium with a large defect suggesting Cor Triatriatum Dexter (Figure 1 and 2).



**Figure 1:** Echocardiography showing the secundum atrial septal defect.

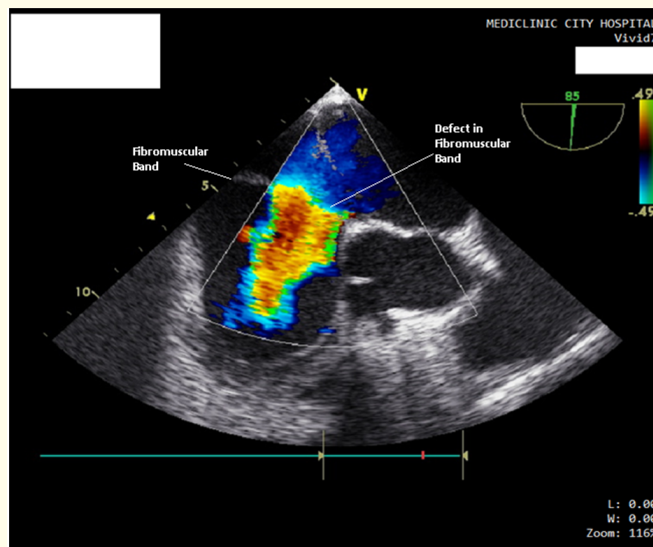


Figure 2: Echocardiography showing defect in the fibromuscular band.

Abdominal Ultrasound confirmed the presence of ascites, congested liver, and enlarged prostate but no other abnormalities.

Investigations were done to rule out vasculitis. He was found to be negative for Anti Neutrophil Cytoplasmic Antibodies (ANCA), double-stranded DNA (dsDNA), and Anti-Nuclear Antibody (ANA) profile. CA 19-9 and CEA were normal. His total Prostatic Specific Antigen (PSA) was elevated at 11.459 ng/mL (< 4.0 ng/mL) hence he was referred to the Urologist who diagnosed Benign Prostatic Hyperplasia.

Following the initial investigations, the patient was admitted to the Intensive Care Unit (ICU) for further management. He was treated with Enoxaparin, Furosemide, Digoxin and Spironolactone. For his pleural effusion, he required a right-sided chest drain and a total of 1200 mL of transudative pleural fluid was drained. However, no sinister etiology for the pleural effusion was noted.

A size 34 mm Amplatzer Septal Occluder Device was successfully used to treat the Secundum ASD percutaneously. As expected, the remaining defect in the fibromuscular band in the right atrium allowed for the necessary right intra-atrial shunt (Figure 3).

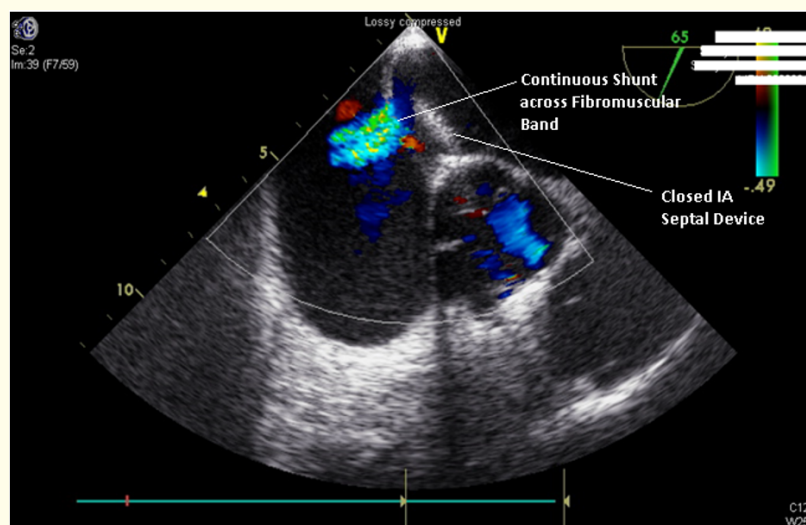


Figure 3: Echocardiography showing continuous shunt across the fibromuscular band defect and closed inter atrial septal defect with the device.

The patient's clinical condition progressed very well through his hospital course. He had a good response to offloading therapy, supportive measures like correction of electrolyte imbalances, nutritional support, broad-spectrum intravenous antibiotics, and physiotherapy. The patient was discharged home in a stable condition. He was reviewed in the Cardiac Clinic 2 weeks later and remained completely asymptomatic with excellent exercise tolerance. Clinical examination confirmed no evidence of acute cardiac decompensation with good rate control.

## Discussion

Cor Triatriatum is a very rare congenital cardiac anomaly with an incidence of 0.1% [4,5]. It is subdivided into two types, the Cor Triatriatum Sinistrum which involves the left atrium, and the much rarer Cor Triatriatum Dexter, which involves the right atrium. The basic underlying pathophysiology is the remnant fetal Eustachian and thebesian valves resulting from poor regression of the right horn of the sinus venosus, which eventually forms the incomplete septum along the lower part of the right atrium [6,7]. Cor Triatriatum Dexter may present with cyanosis if there is right ventricular outflow tract obstruction. They are often accompanied by other congenital cardiac defects like right ventricular hypoplasia, tricuspid valve hypoplasia, or pulmonary venous defects, but the exact association is unknown due to the small number of cases being reported [8,9].

The diagnosis is done by echocardiography, contrast echocardiography, or cardiac Magnetic Resonance Imaging (MRI). Most of the patients who are asymptomatic or with mild symptoms are not treated unless they require cardiac or thoracic surgery of any type. In the past, the mainstay of treatment was surgical resection of the fibromuscular band and ASD closure. Recent advances like the percutaneous disruption of the membrane or fibromuscular band and closure of ASD, are replacing the surgical modalities with less morbidity and mortality of such patients [10].

## Conclusion

Cor Triatriatum is an exceedingly rare congenital cardiac anomaly that is seen in neonates and children and quite uncommon in the adult population. There are only a few cases reported in adults with Cor Triatriatum sinister presenting with atrial fibrillation but are quite rare in adults with Cor Triatriatum Dexter. We report an exceedingly rare case of a 73-year-old adult male who presented with Cor Triatriatum Dexter with Ostium Secundum ASD, who had his initial clinical presentation with acute decompensated cardiac failure and atrial fibrillation. He underwent percutaneous correction and closure of the ASD with a good outcome.

## Conflict of Interest

The authors certify that we have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

## Consent

Written informed consent could not be obtained from the patient for publication as the patient could not be contacted. The entire case report has been sufficiently anonymized as per the ICMJE guidelines. Consent waiver has been issued by Mediclinic Research and Ethics Committee, and a copy of the waiver is available for review by the Editor-in-Chief of this journal on request.

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