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**Case Report**

# Cor Triatriatum in the Adult with Aortic Stenosis and Mitral Stenosis

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**SUMMARY**

**Background** : Cor triatriatum is a rare congenital cardiac anomaly, in which the left atrium or right atrium is separated by an abnormal fibromuscular membrane with one or more restrictive orifices. This condition typically presents in infancy or early childhood and can be associated with other cardiac anomalies.

**Case presentation** : A 75-year-old woman was admitted for exertional dyspnea with moderate aortic and mitral stenosis. As cor triatriatum was revealed by a computed tomography and echocardiography, she was referred to our department for surgery. Aortic valve replacement, mitral valve replacement and excision of the membranous septum in the left atrium was performed. This report presents an incidental findings of cor triatriatum with aortic stenosis, moderate mitral stenosis in septuagenarian.

**Conclusion** : We encountered a rare case of cor triatriatum with aortic stenosis and mitral stenosis in septuagenarian. She was incidentally diagnosed by rheumatic aortic and mitral stenosis which had advanced to moderate level.

**Key Words** : aortic stenosis, mitral stenosis, cor triatiatum

## BACKGROUND

Cor triatriatum is a rare congenital cardiac anomaly, in which the left atrium or right atrium is separated by an abnormal fibromuscular membrane with one or more restrictive orifices. It occurs in approximately 0.1% of children who have congenital heart disease, with a male to female ratio of roughly 1 : 1[1]. This condition typically presents in infancy or early childhood and can be associated with other cardiac anomalies. Therefore, adult cases are very rare. This report presents an incidental findings of cor triatriatum with moderate aortic stenosis (AS) and moderate mitral stenosis (MS) in septuagenarian.

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## CASE PRESENTATION

A 75-year-old woman with exertional dyspnea was consulted to our hospital. Further investigation revealed moderate AS, moderate MS and Cor triatriatum. She was referred to our department for surgery.

On physical examination, her blood pressure was 130/60 mmHg with a heart rate of 73/min, normal. A grade 2/6 systolic murmur in the right upper sternal border and a grade 2/6 diastolic murmur in the apex were audible. Lungs were clear by auscultation and there was no cyanosis, clubbing, or edema of extremities. A brain natriuretic peptide (BNP) was 130.2 pg/mL. A chest X-ray showed a 49.5% cardiothoracic ratio with no pulmonary congestion and pleural effu-

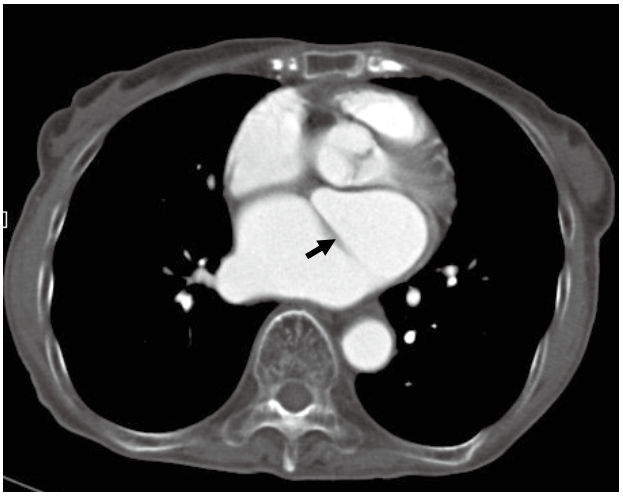

**Fig. 1** Enhanced computed tomography findings. Enhanced computed tomography shows a membrane structure (arrow) that divides the left atrium.

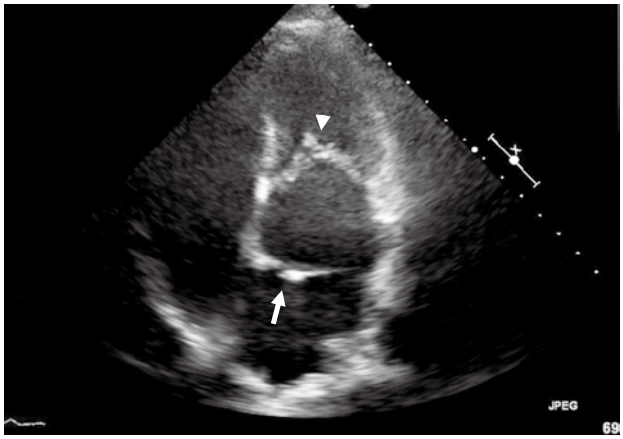


**Fig. 2** Echocardiography findings. Transthoracic echocardiogram (apical 4-chanber view) shows a membranous structure in the left atrium (arrow). The mitral valve cusps are thickening (arrowhead).

sion. Enhanced computed tomogram shows a membrane structure that divides the left atrium (LA) (Fig. 1). Transthoracic echocardiography revealed a normal left ventricle function (ejection fraction, 71.8 %) without asynergy. There was moderate AS (aortic valve area : 0.88 $cm^2$, mean pressure gradient : 12 mmHg) and moderate MS (mitral valve area : 1.20 $cm^2$, mean pressure gradient : 4 mmHg). The LA was mildly dilated and it was divided into two distinct chambers by a membranous septum (Fig. 2). There was no significant pressure gradient across the membranous septum. Similarly, there was no color flow acceleration noted across the membrane. Intra cardiac shunt at the atrial level was not revealed. Cardiac catheterization were performed with simultaneous pulmonary capillary wedge and left ventricular end diastolic pressure measurement, revealing a mean gradient of 7 mmHg. Pressure were as follows- aortic pressure 143/56 (98), left ventricular pressure 132/− (14), pulmonary arterial pressure 30/8 (15), pulmonary capillary wedge pressure (12), right ventricular pressure 33/1 (6), right atrial pressure (13) mmHg. Coronary artery angiography revealed no stenosis.

Surgical intervention of the membrane, aortic valve and mitral valve was recommended. We performed aortic valve replacement, mitral valve replacement and excision of the membranous septum in the left atrium. After median sternotomy, a cardiopulmonary bypass (CPB) was established with ascending aorta and bicaval canulation. In right-side left atrium approach, the membranous septum in the left atrium was revealed (Fig. 3a). It inserted into the interatrial septum, at the fossa ovalis region, and there was a large orifice at the lateral wall of the left atrium. The membrane was distinguished from a supravalvular mitral ring by its position superior to the left atrial appendage. It was removed (Fig. 3b). After that, due to the rheumatic AS and MS, the aortic valve and mitral valve were replaced with a 19-mm and 27-mm stented bioprosthesis (Carpentier-Edwards Perimount valve, Carpentier-Edwards Lifesciences, Irvine, CA, USA). The patient was weaned from CPB without incident (cardiopulmonary bypass time, 227 min ; aortic clamp time, 199 min). The postoperative course was uneventful.

## DISCUSSION

Cor triatriatum, first described by Church in 1868 is an uncommon congenital anomaly[2]. It occurs in approximately 0.1% to 0.4% of children who have congenital heart disease[3,4]. It has gender predilection with slight involvement 1.4 : 1 and is associated with other cardiac defects in up to 50% of cases[5]. Therefore, Cor triatriatum in adults is very rare.

The embryologic basis of Cor triatriatum is disputed. The three main theories are malseptation involv-

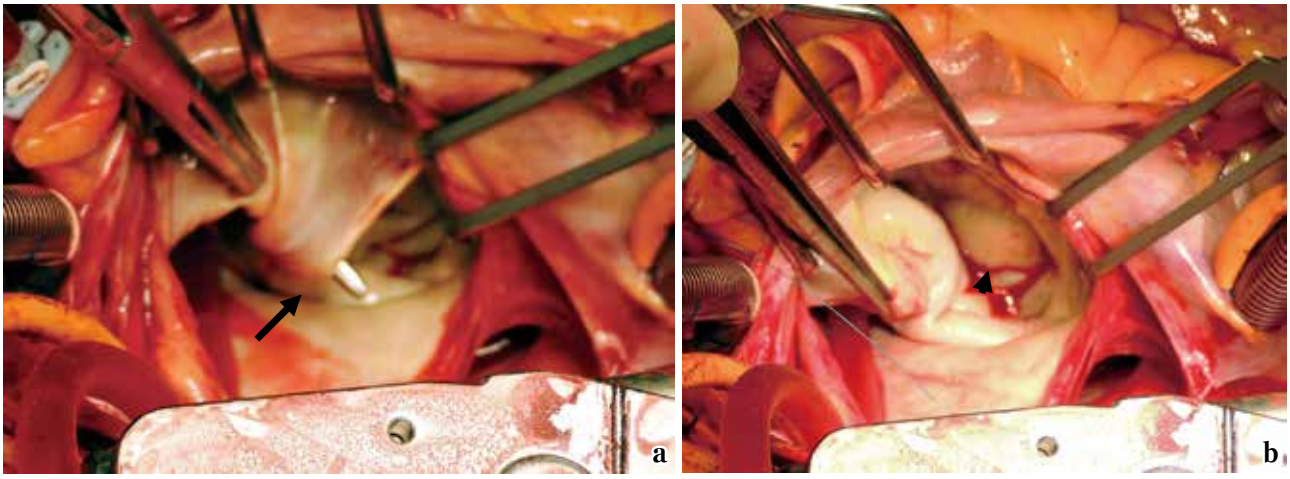

**Fig. 3** Intraoperative findings.
**a** : Operaive view of membrane. The left atrium is opened and the surgical instrument is inserted though the defect in the membrane (arrow).
**b** : Tthe membrane in the left atrium is excised. The mitral valve is revealing (arrowhead).

ing the septum primun, malincorporation of the common pulmonary vein, and entrapment of the common pulmonary vein[6,7]. Although the malincorporation hypothesis suggesting incomplete incorporation of the common pulmonary vein into the left atrium is the most widely accepted, it cannot explain all anatomic variants[8].

Cor triatriatum was classified into three groups according to the size of opening of the membrane : (1) no opening, accessory left atrium drains into the right heart ; (2) accessory left atrium drains into the true left atrium by either one or more very small orifices, resulting in high degree of obstruction ; (3) accessory left atrium and true left atrium communicate through a large orifice, the non-obstructive form, producing milder symptoms[9]. Classically, patients present symptoms during the neonatal or early infancy, although patients with group 3 cor triatriatum may remain undetected until late adulthood depending on the diameter of the orifice and the degree of obstruction. When the diameter is >1 cm, patients are symptom-free[9,10].

The most common symptoms present in adults are dyspnea, hemoptysis, and orthopnea, but cor triatriatum can be asymptomatic and the diagnosis can be incidental[6,11]. The hemodynamics consequences and symptoms of cor triatriatum resemble those of mitral stenosis. The differential diagnosis includes mitral stenosis, supravalvular mitral ring, left atrial tumor, thrombus, cyst, left atrial dissection, or pulmonary vein stenosis[10]. Cor triatriatum is often associated with other cardiac anomalies. In adults, mitral regurgitation, secundum atrial septal defect, and a left superior vena cava with unroofed coronary sinus are most common, although it has also been reported to be associated with aortic regurgitation with dissecting aneurysm, and anomalous pulmonary venous connection[1,6]. A definitive treatment for cor triatriatum is complete surgical excision of the membrane, which has shown excellent long-term results, with 85% survival without recurrence[10].

Our case was a group 3 anomaly with moderate large diameter orifice, and associated without other congenital heart disease. Therefore, she could remain asymptomatic and she was incidentally diagnosis as this disease.

## CONCLUSION

We encountered a rare case of cor triatriatum with moderate AS and MS in septuagenarian. She was incidentally diagnosed by rheumatic AS and MS that had advanced to moderate level. Therefore, aortic valve replacement, mitral valve replacement and excision of the membranous septum in the left atrium was performed.

**Disclosure Statment**

This study was not funded by any public, commercial or not-for-profit agency.

**Conflicts of interest statement**

None declared

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