

Cor Triatriatum Sinister

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Cor triatriatum is a very rare congenital cardiac anomaly with no known associated genetic abnormalities. Cor triatriatum is generally an isolated anomaly but may be associated with anomalous pulmonary venous return in approximately 10% of patients, patent ductus arteriosus, left superior vena cava, ventricular septal defects, Shone syndrome, tricuspid atresia, Ebstein malformation, atrioventricular canal, and tetralogy of fallot.¹ Here, we describe a male with cor triatriatum who presented with progressive dyspnea upon exertion. A 24-year-old active male presented with progressive dyspnea upon exertion and palpitation from 2 months ago (Functional Class 2 late, New York Heart Association). There were no significant symptoms earlier in life. He had a history of severe common cold one month ago that resolved spontaneously without any treatment. Upon physical examination, he had sinus tachycardia, normal blood pressure, clear lung, accentuation of second heart sound, apical holosystolic murmur grade 2/6 with radiation to the axilla, and no central cyanosis. Other systems were normal. Electrocardiogram showed left atrial enlargement with nonspecific ST-T changes. Chest x-ray showed mild increase in cardiac size with pulmonary venous congestion, left atrial and left atrial appendage enlargement. Transthoracic echocardiography showed mild left ventricular enlargement, mildly reduced ejection fraction, and discrete membrane in the left atrium that subdivided this chamber to 2 separate parts. Mitral valve and left atrial appendage were in the distal chamber and foramen ovale and pulmonary veins in the proximal chamber. There was mild mitral valve thickening and regurgitation and tricuspid regurgitation in echocardiography (Figure 1). Doppler study showed a typical turbulent flow across the membrane with a 20 mm Hg gradient and a systolic pulmonary artery pressure of approximately 55 mm Hg. Transesophageal echocardiography was refused by the patient. Cardiac catheterization showed moderate pulmonary hypertension with 20 mm Hg gradient between wedge pressure and left ventricular end diastolic pressure. Dye injection in the pulmonary artery with late opacification (Levophase) showed no abnormalities in pulmonary venous return, and a delicate and discrete membrane inside the left atrium with 2 separate chambers. No other associated congenital cardiac anomalies were found. Left ventricular size was enlarged mildly with

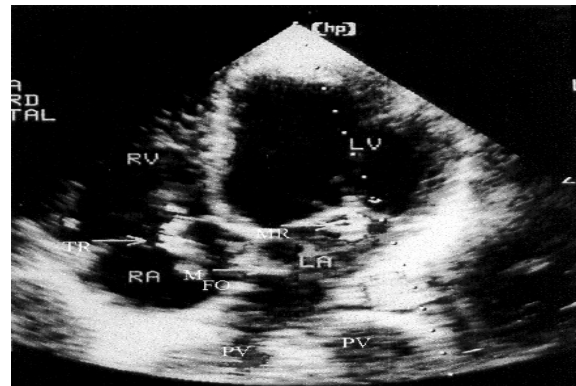


Figure 1 - Echocardiogram before surgery. Note to the mitral regurgitation (MR) and tricuspid regurgitation (TR) during systole. Echocardiogram shows the entrance of two pulmonary veins into the proximal chamber (PV), foramen oval (FO), membrane (M).

good ejection fraction and mild mitral regurgitation (one degree). Surgery was planned. Median sternotomy was performed. Open correction was performed on cardiopulmonary bypass through atrial incision with complete resection of the diaphragm. This was a single perforation in the center of the diaphragm. There was no significant residual regurgitation on mitral valve. Foramen ovale was in the proximal chamber, but the left atrial appendage was in the distal. Tissue sample was sent for pathological evaluation. He was disconnected from the pump easily with satisfactory hemodynamic. After 6 days, he was discharged from the hospital in good condition. Echocardiography, one month after surgery exhibited normal left ventricular size with a good ejection fraction, disappearance of stenotic diaphragm, mild mitral regurgitation and normal pulmonary pressure. Currently, he is in good condition without any problem. The membrane is composed of a histological structure similar to the wall of pulmonary vein (bases). The membrane contains an outer thin endothelial lining (smooth wall) with its sub endothelial layer of loose connective tissue. Below this membrane, there are myocardial muscle fibers running in isolated but parallel direction. The nuclei were shrunken in size, but there is no evidence of myocardial fiber degeneration and infiltration of inflammatory cells.

Cor triatriatum is a very rare congenital malformation, occurring in 4% of patients with cardiac defects. There are no known risk factors or associated genetic abnormalities. This anomaly is generally considered to be a result of faulty incorporation of the common pulmonary vein into the left atrium so that pulmonary veins do not directly join to the left atrium. The foramen ovale and the left atrial appendage usually communicate with the distal left atrium (true

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left atrium) distinguishing cor triatriatum from a supra valvular mitral ring. The observation that a left superior vena cava is frequently associated with this lesion has led some to propose impingement of the left superior vena cava on the developing left atrium as a potential pathogenesis.¹ This fibro muscular membrane subdivides the left atrium into a venous portion, sometimes called the proximal chamber, and a true left atrium portion, referred to as the distal chamber. In almost all cases, the left atrial appendage is distal to the cor triatriatum membrane. The location of the fossa ovalis is variable. In the typical form, the windsock funnel-shaped membrane is perforated by a single hole (90%) or less commonly, multiple fenestrations (10%). Marine Garcin et al classified cor triatriatum into 3 types: fibro muscular diaphragm, hourglass, and elongated tubular accessory left atrial connection.² Cor triatriatum is differentiated from the supra valvular mitral ring by the fact that the membrane is usually shelf-like and located just superior to the mitral annulus. Whereas, in cor triatriatum, the left atrial appendage is located in the true left atrium below the membrane. This is an important anatomic feature that distinguishes cor triatriatum from mitral valve stenosis and supra valvular ring. In supra mitral valve ring, the left atrial appendage is situated in the distal chamber and the membrane merely consists of fibrous tissue. The Supra mitral valve ring is usually associated with a structural defect in the mitral valve.² Cor triatriatum appears to have a slight male predilection with a male-to-female ratio of 1.4:1. Most patients present early in life. Presentation later in life is less typical, but evidence of pulmonary congestion predominates.² Occasionally, it may be noted incidentally on echocardiography. Although, presentation in their eighth or ninth decade of life was reported rarely. The age at clinical presentation usually depends on the degree of obstruction in the left atrial blood flow, and in the presence of associated left to right shunts or other cardiac defects. Upon physical examination, the pericardium is quiet unless pulmonary arterial pressure is elevated. The chest roentgenogram in cor triatriatum usually shows redistribution of blood flow with increased pulmonary vascular markings suggestive of pulmonary venous congestion. The heart size is normal or mildly enlarged. The ECG may be normal; however, right axis deviation, right atrial or biatrial enlargement, and right ventricular hypertrophy are more commonly seen.² The echocardiography is the best tools for diagnosis, as it can demonstrate no mobile membrane transverse the left atrium.^{2,3} Transesophageal echocardiography was used for the evaluation of entrance of pulmonary

veins and delineation between the membrane and left coronary artery fistula.⁴ Care must be taken to trace the entrance of all 4 pulmonary veins, especially if surgery is to be carried out without catheterization.⁵ At catheterization, generally elevation in pulmonary artery and capillary wedge pressure has been reported. The Levophase of a pulmonary artery injection in the cranially angulated anteroposterior projection usually demonstrates the associated anomalies and delicate membrane into the left atrium. The natural history of the defect is dependent on the size of the ostium. Usually infants are critically ill and die at a young age.³ If the communication is larger, patients may present later in life with a clinical picture very similar to mitral valve stenosis. Our patient had symptoms similar to mitral stenosis. Surgical management is the only treatment of choice and usually involves excision of the intra-atrial membrane. Dilatation of the membrane intraoperatively or percutaneously by balloon has been associated with a high chance of restenosis. Reported mortality for all ages of patients is approximately 16-50% depending on associated anomaly and the general condition of patients. No significant late mortality has been reported from arrhythmias or other problems.¹

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