



COR TRIARIATUM: A NARRATIVE REVIEW

MEGHANA CHANAMOLU, KRITHIKA SUNDARAM, WHITENY BAXTER, SHASHANK CHANAMOLU

INTRODUCTION

Cor triatriatum is a rare congenital heart defect that has an incidence of 0.1% in all patients with congenital heart problems. In the United States, cor triatriatum occurs in 0.1%-0.4% of infants who have congenital heart disease. This condition has no genetic association and has a male to female ratio of 1.5:1.

Cor triatriatum is caused by abnormal fibromuscular membrane septation that splits either the left or right atrium into three chambers. Cor triatriatum sinister (CTS) is the most common form of this condition, resulting from a thin membrane splitting the left atrium into two chambers. When this occurs in the right atrium it is called cor triatriatum dexter (CTD) which is a rarer heart defect than CTS. The severity of both conditions varies as they can present early in infancy or go undetected until adulthood.

ANATOMY & PATHOPHYSIOLOGY

CTS the left atrium is divided into two chambers, proximal and distal (**Figure 1**). The distal chamber receives all the blood coming from pulmonary veins, while the proximal chamber contains the mitral valve and left appendage. The membrane that divides the atria can be classified as complete, incomplete, or fenestrated. There are three different anatomic variations in the membrane: diaphragmatic, hourglass, or tubular.

Malincorporation theory is the failure of pulmonary veins to fuse into the left atrium resulting in a narrow pulmonary venous ostium that later gives rise to the atrial appendage. The atrial appendage functions like a septum-like structure that divides the left atrium into two chambers. Malseptation theory states how the fibromuscular membrane is an abnormal outgrowth of the septum primum. The entrapment theory describes how the membrane develops from the trapping of the common pulmonary vein in the embryonic sinus venosus, thus preventing its fusion into the left atrium.

CTD is a cardiac malformation that causes division of the right atrium due to the failure of the right-sided sinus venous valve's embryonic remnant to regress (**Figure 2**). Sinus venosus fails to relapse, it gives rise to a network of fenestrated fibers called Chiari's network. These fibers then form a membrane in the right atrium leading to CTD. The most common location of the membrane is at the right superior vena cava, coronary sinus, and inferior vena cava.

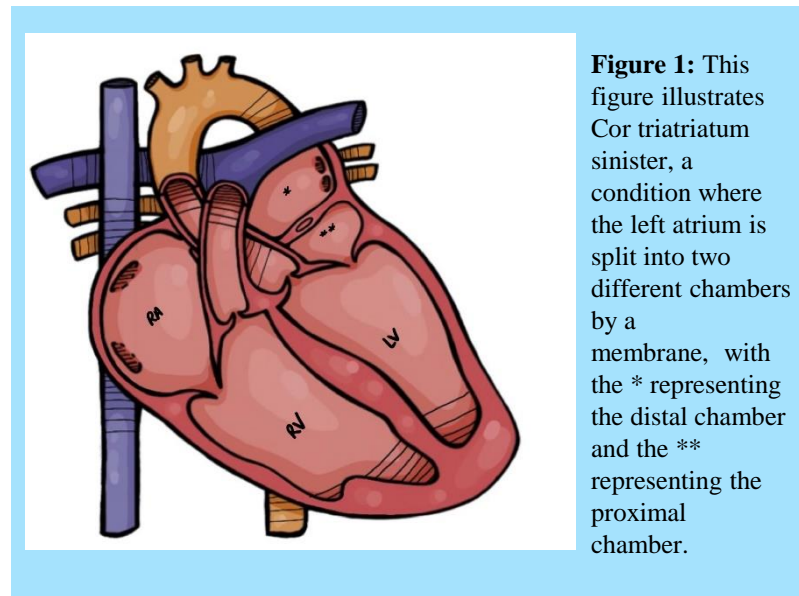


Figure 1: This figure illustrates Cor triatriatum sinister, a condition where the left atrium is split into two different chambers by a membrane, with the * representing the distal chamber and the ** representing the proximal chamber.

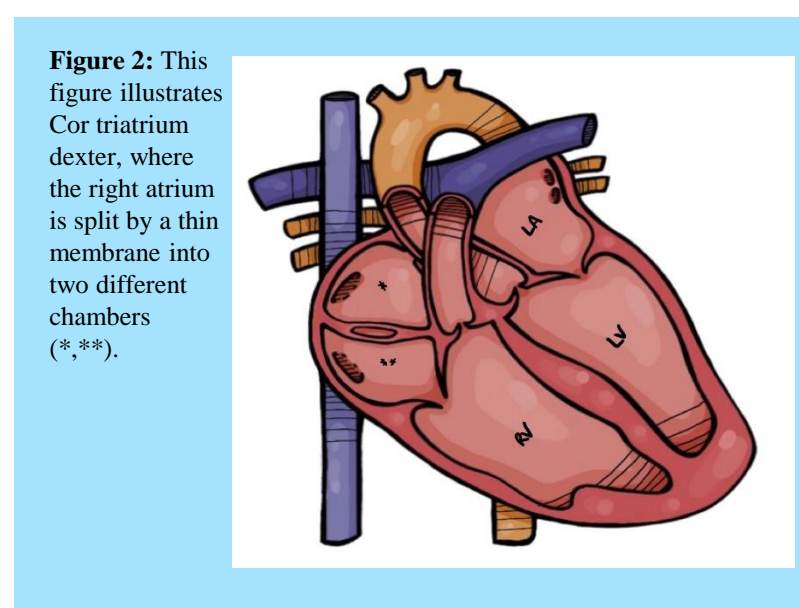


Figure 2: This figure illustrates Cor triatriatum dexter, where the right atrium is split by a thin membrane into two different chambers (*,**).

CLINICAL PRESENTATION

Child

The age of presentation of cor triatriatum sinister is dependent on the size and number of fenestrations. Membrane with larger size holes reports symptoms later in life than those with a smaller size. General symptoms include tachypnea, wheezing, murmurs, coughing, and pulmonary congestion, leading to cardiomegaly and congestive heart failure. Cardiac symptoms include congestive heart failure, low cardiac output syndrome, respiratory complaints, hepatomegaly, cardiomegaly, pulmonary edema, partial anomalous pulmonary venous connection (PAPVC), ASD, PDA, VSD, tetralogy of Fallot, double outlet right ventricle, pulmonary hypertension, and secondary pulmonary hemosiderosis.

Adult

Adults are primarily asymptomatic because they lack an intra-atrial pressure gradient due to the large size of the foramen, which causes minor obstruction. Symptoms appear after wear and tear of the membrane, causing the membrane to become sclerosed and calcified, leading to more obstruction and creating a prominent intra-atrial pressure gradient. Patients can present with dyspnea, orthopnea, hemoptysis, and palpitations, hypercoagulable state increasing the risk of thromboembolism formation, and congestion of blood leading to pulmonary or systemic hypertension.

DIAGNOSIS

Diagnosis of cor triatriatum requires many imaging methods, including echocardiography, angiography, chest X-ray, ECG, and cardiac CT. Echocardiography is the preferred method for diagnosis not only because of its accuracy, but also its ability to construct a three-dimensional model of the heart identifying the actual location of the membrane, aiding in the creation of a surgical approach. Cardiac CT is now emerging as a more preferred test to diagnose cortriatrium as it is a non-invasive way of identifying heart disease due to its high temporal and spatial resolution.

DIFFERENTIAL DIAGNOSIS

CTS has similar presentations with other congenital heart diseases and should be distinguished clearly from other conditions when diagnosing this disorder. Some of these diseases include Cor triatriatum supravulvular mitral stenosis, mitral stenosis, idiopathic pediatric pulmonary arterial hypertension, pulmonary venous hypertension, total anomalous pulmonary venous return, partial anomalous pulmonary venous return, atrial septal defect, ventricular septal defect, idiopathic pulmonary hypertension, tricuspid stenosis, and atrial myxoma.

TREATMENT

Neonates undergo cardiopulmonary bypass with mild to moderate hypothermia in which the surgical procedure involves resection of the fibromuscular membrane that divides the left atrial cavity usually done through the transeptal path.

Catheter-based ablation is widely used to treat tachyarrhythmias that might develop after long-term surgical resection of the membrane, but correction of other anatomical variations via surgery is the main treatment in adults with a 10-year survival rate of 83%. The surgical approach reduces flow obstruction and if the surgery is performed in young individuals with congenital heart disease, they have a greater possibility of adverse outcomes with a lower survival rate.

CONCLUSION

Due to the rarity of this condition, physicians must learn to distinguish this condition from other heart diseases..