

## Cor triatriatum sinistrum in adults: surgical treatment of two cases

### *Erişkin iki vakada cor triatriatum sinister*

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### Introduction

Cor triatriatum sinistrum is a rare congenital cardiac malformation, constituting about 0.1-0.4% of congenital heart disease (1, 2). It is characterized by the presence of a fibromuscular membrane that subdivides the left atrium into two chambers in the classical form. Most patients with classic cor triatriatum present during the neonatal period or early infancy but adult case of cor triatriatum is very rare. In this report, we present successful surgical treatment of two cases with adult cor triatriatum.

### Case Reports

#### Case 1

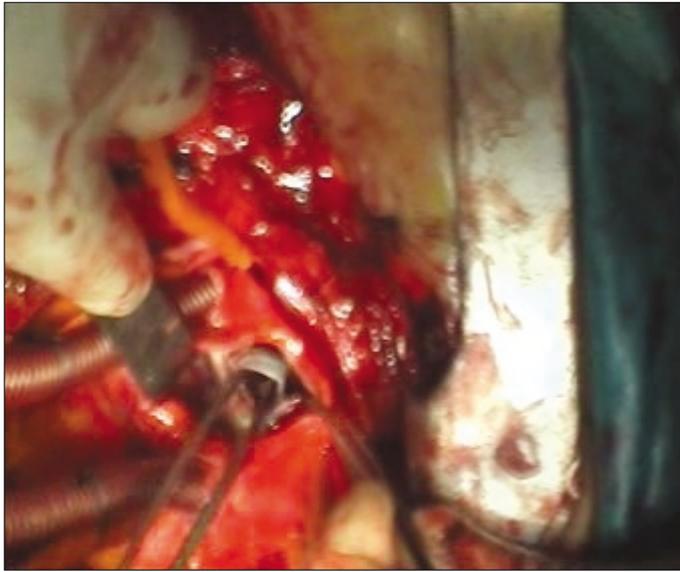
A 32-year-old man was admitted to our hospital with a history of dyspnea on exertion and increasing fatigue. At his medical history, he also had atrial septal defect (ASD) with inferior vena cava type that had been operated by direct suture at age of 11.

On his physical examination, his blood pressure was 110/70 mmHg, heart rate 73 bpm, regular. Cardiac auscultation revealed a fixed split second sound. Other systems were normal. The chest X-ray showed moderately enlarged configuration of the heart with vascular redistribution to both lower lobes. Electrocardiography revealed sinus rhythm, right-axis deviation, and right atrial enlargement. Transthoracic echocardiography (TTE) revealed the fibromuscular membrane located above the mitral ring and left atrial appendage, and it was associated with ASD between the left atrial accessory chamber and the right atrium. Mean pulmonary artery pressure was 50 mm Hg. The ratio of pulmonary blood flow to systemic blood flow (Qp/Qs) was 1.4. Transesophageal echocardiogram (TEE) was performed and the diagnosis cor triatriatum was confirmed. We diagnosed this case as A1 type of cor triatriatum according to Rodefeld classification, depending on the presence and localization of ASD. The patient was operated with the diagnosis of cor triatriatum and secundum

atrial septal defect. He underwent electively surgical operation, through a median sternotomy. After aortic and bicaval venous cannulation, following the initiation of cardiopulmonary bypass (CPB) and after application of the aortic cross-clamp, under mild systemic hypothermia, the heart was arrested with cold blood cardioplegia. The ASD was determined and cor triatriatum membrane was reached via a transeptal approach (Fig. 1). The membrane was successfully resected and the atrial septal defect was then closed with Goro-Tex patch. The postoperative course was uneventful, and the patient was discharged on postoperative day 7. The postoperative cardiac echocardiogram revealed satisfactory results, he is doing well now.

#### Case 2

A 38-year-old man was referred to our hospital for evaluation of a murmur. He had history of progressive dyspnea on exertion over one year. On examination, the patient was found to be normotensive with a regular pulse of 75 beats / min. On auscultation, the S1 was normal; the S2 was found to be widely split with a grade II/VI ejection systolic murmur best heard over the second to third left intercostals space. Electrocardiography showed sinus rhythm, right axis deviation, right ventricular hypertrophy. Chest radiography displayed significant cardiomegaly with pulmonary venous congestion. Echocardiography revealed the presence of the accessory atrial chamber communicating with right atrium. Transesophageal echocardiography clearly demonstrated cor triatriatum, and ASD (Fig. 2). The diagnosis of A1 type of cor triatriatum according to Rodefeld classification was established. He underwent electively surgical operation, through a median sternotomy with initiation of cardiopulmonary bypass (CPB), application of the aortic cross-clamp, under mild systemic hypothermia and cold blood cardioplegia. The cor triatriatum membrane was resected through access via a transeptal approach. The ASD closure was performed using Goro-Tex patch. The postoperative cardiac echocardiogram showed satisfactory results and postoperative course was uncomplicated. The patient was discharged on postoperative day 4.



**Figure 1. Operating view of cor triatriatum. Surgical forceps are inside the obstructing membrane**

## Discussion

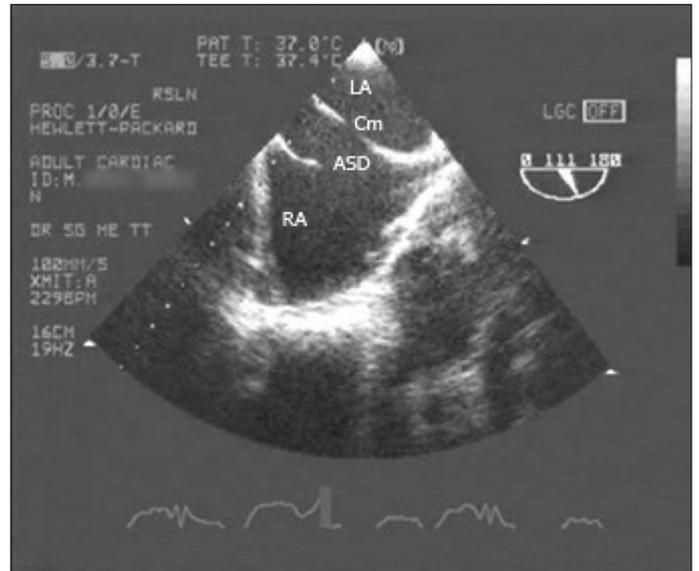
Traditionally cor triatriatum was encountered most often in infancy and childhood; diagnosis in adulthood is extremely rare. We found only about 50 cases of cor triatriatum in adult in the literature between January 1995 and 2005 years.

Rodefeld presented morphologic classification of cor triatriatum as follows: type A1 - ASD proximal to the obstructing membrane; type A2 - ASD distal to the obstructing membrane, type B, dilated coronary sinus that receives all four pulmonary veins; type C - accessory chamber that receives no pulmonary veins (3). Our cases 1 and 2 had A1 type of cor triatriatum according to Rodefeld classification.

The clinical picture and survival of patients with cor triatriatum in adult depends on the degree of obstruction to pulmonary venous flow and the associated intracardiac defects (1, 4).

Marin-Garcia and Thelenius reported that ASD or patent foramen ovale was present in 70-80% of patients with cor triatriatum (5, 6). When the ASD exists between the left atrial accessory chamber and the right atrium, the patients apply to hospital with symptoms of associated elevated pulmonary venous and arterial pressures because of blood is shunted from left to right (7). In our series, the patients had history of dyspnea on exertion.

Although cross-sectional echocardiography with Doppler has been shown to be of great value in diagnosis of patients with cor triatriatum, biplanar TEE provides a more complete and detailed data of this anatomy (8, 9). Currently computed tomography scan, magnetic resonance imaging, and 3D-TEE are used for the assessment of cor triatriatum. In the current series all patients were diagnosed with TTE.



**Figure 2. Transesophageal echocardiographic view of discrete membrane in left atrium**

ASD- atrial septal defect, Cm- membrane of cor triatriatum, LA- left atrium, RA- right atrium

Surgery is an unique treatment of choice for cor triatriatum. Surgical correction of cor triatriatum in adult is a relatively simple and long-term results are very satisfactory. Our patients also underwent successful surgical operation and they are doing well at the 18th- month follow-up visit.

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