

Cor triatriatum sinister: a case series

Kor triatriatum sinister: Olgu serisi

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ABSTRACT

Objective: As a highly rare congenital defect, cor triatriatum sinister represents only 0.1% of congenital cardiac anomalies. Depending on the degree of obstruction and the accompanying symptoms, cor triatriatum can be diagnosed at any age. This case series described 5 patients with cor triatriatum sinister who underwent operation.

Methods: Five patients with cor triatriatum sinister were seen at our institution between 2007 and 2013. The demographic characteristics and surgical results of these patients are outlined in this retrospective review.

Results: The surgical approach consists of left or right atriotomy, excision of the obstructing membrane, and repair of the associated intracardiac anomalies. After an uneventful postoperative hospital stay, all patients were discharged 5–10 days postoperatively. There were no instances of recurrent constriction after surgical treatment of the cor triatriatum membrane. Patients were followed up for a median of 4 years and were symptom free.

Conclusion: In the surgical management of this easily and fully treatable congenital cardiac anomaly, it is difficult to determine which atriotomy approach is comparatively more advantageous. However, in the management of cor triatriatum sinister, priority should be given to confirmation of the diagnosis and full resection of the membrane. Thus, the surgeon should not hesitate to perform additional incisions if deemed necessary.

As a highly rare congenital defect, cor triatriatum sinister represents only 0.1% of congenital cardiac anomalies.^[1] In cor triatriatum, the pulmonary veins enter a posterior proximal left atrial chamber that is separated from the anterior distal left atrial chamber containing the mitral valve and left atrial ap-

ÖZET

Amaç: Son derece nadir görülen kor triatriatum sinister doğumsal kalp anomalilerinin sadece %0.1'ini oluşturmaktadır. Darlığın derecesine ve eşlik eden belirtilere bağlı olarak her yaşta kor triatriatum tanısı konulabilir. Bu olgu serisinde kor triatriatum sinister tanısıyla ameliyat edilen beş hasta sunuldu.

Yöntemler: Kor triatriatum sinister tanısı konan beş hasta 2007–2013 tarihleri arasında hastanemizde ameliyat edildi. Bu hastaların demografik özellikleri ve cerrahi sonuçları bu geriye dönük incelemede özetlendi.

Bulgular: Cerrahi yaklaşım tıkaçıcı membranın kesilip çıkarılması ve ilişkili kalp içi anomalilerin tamirini içeren sol veya sağ atriyotomiden oluşmaktadır. Sorunsuz ameliyat sonrası izlemin ardından tüm hastalar 5-10 gün arasında taburcu edildi. Kor triatriatumun cerrahi tedavisi sonrası tekrarlayan daralma görülmedi. Hastalar ortalama 4 yıl boyunca semptomsuz olarak takip edildi.

Sonuç: Kolayca ve tamamen tedavi edilebilir bu doğumsal kalp anomalisinin cerrahi yönetiminde, hangi atriyotomi yaklaşımının nispeten daha avantajlı olduğunu belirlemek zordur. Ancak, bu anomaliye yaklaşım öncelikle tanının doğrulanması ve membranın tamamen kesilip çıkarılması şeklinde olmalıdır. Bu nedenle, cerrah gerekli gördüğü takdirde ek kesi yapmada tereddüt etmemelidir.

pendage by a diaphragm in which there are 1 or more restrictive ostia.^[1]

Cor triatriatum can be diagnosed at any age, depending on the severity of the obstruction and con-

Abbreviations:

ASD Atrial septal defect
CT Computed tomography
MRI Magnetic resonance imaging
TTE Transthoracic echocardiogram

Received: April 20, 2015 Accepted: June 11, 2015

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comitant symptoms. In symptomatic patients, treatment consists of a resection of the diaphragm and correction of the associated congenital heart defects. Although cor triatriatum can be an isolated lesion, it is associated with other congenital cardiovascular anomalies.^[2]

In this study, we investigated patients with cor triatriatum sinister who underwent operation.

METHODS

Five patients with cor triatriatum sinister were seen at our institution between 2007 and 2013. The clinical presentation, diagnostic evaluation, and surgical results are outlined in this retrospective review. Operation is the treatment of choice for this rare congenital cardiac defect. Resection of the obstructing atrial membrane was performed using cardiopulmonary bypass through a median sternotomy with standard aortobicaaval cannulation in all cases.

RESULTS

The demographic characteristics and postoperative clinical data of these patients are given in Table 1. Two cases presented with tachycardia, and 3 cases

presented with signs and symptoms of congestive heart failure. There were 3 boys and 2 girls, aged 4 months, 6 months, 4 years, 10 years, and 18 years. The anatomy was delineated with transthoracic echocardiogram (TTE) in all cases; however, other techniques such as catheter angiography and computed tomography (CT) were used to confirm the diagnosis (Figure 1). The morphologic classification of cor triatriatum sinister first described by Lam et al.^[3] in 1962 was used (Table 2). Left atriotomy was performed in 1 patient, and right atriotomy was performed in the remaining 4. Associated anomalies included atrial septal defect (ASD) in 2 patients and partial anomalous pulmonary venous return in 1 patient. The cases with ASD were repaired with an autologous fresh pericardial patch. In the case with partial anomalous pulmonary venous return, the right upper pulmonary vein was returned anomalously to the left innominate vein. Direct anastomosis between the left atrium and the anomalous right upper pulmonary vein was successfully performed. There were no complications related to operative and postoperative mortality. The postoperative course was excellent in all 5 surviving patients; all remained asymptomatic throughout the follow-up period.

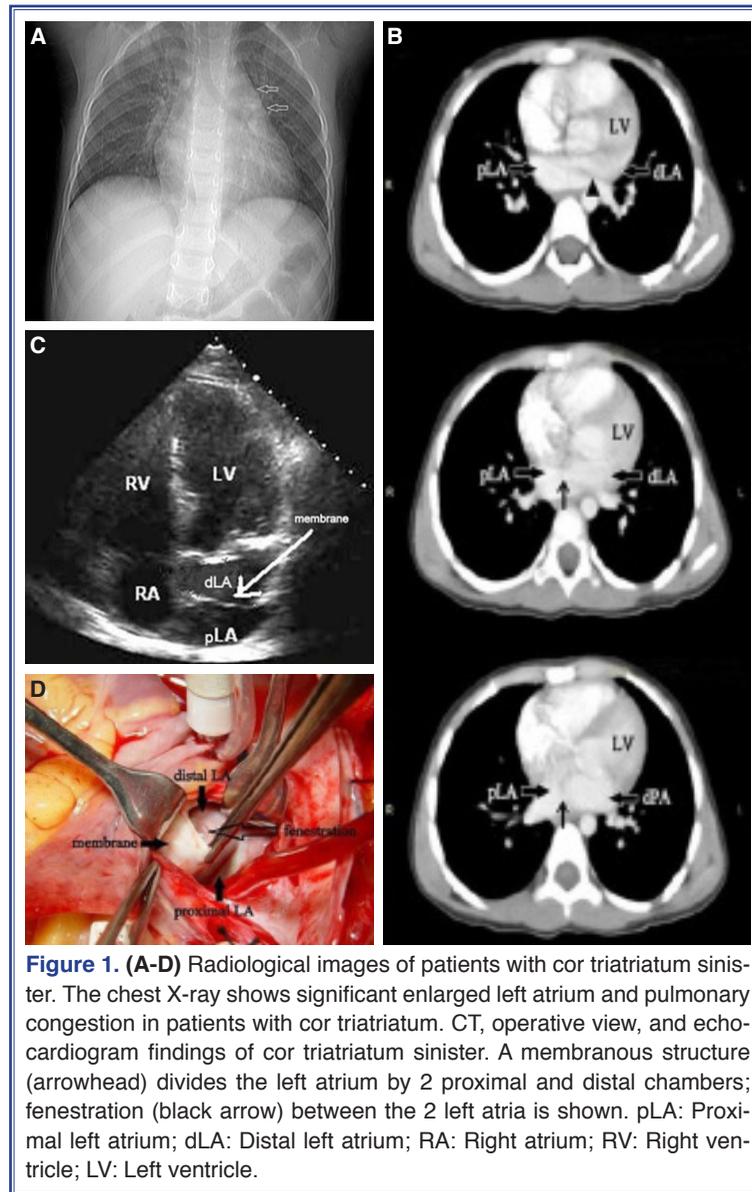
Table 1. Demographic and clinical characteristics of patients

Patient	Age	Gender	Weight (kg)	ASD	TPG (mmHg)	Lam classification	Hospital stay (days)	Follow-up (years)
1	4 months	Male	4	+	13	A2	5	1
2	6 months	Female	6	+	10	A1	10	3
3	4 years	Male	13	+	8	A1	6	5
4	10 years	Female	21	–	10	A	6	4
5	18 years	Male	48	–	12	A	5	7

TPG: Transmembrane pressure gradient; ASD: Atrial septal defect.

Table 2. Lam classification of cor triatriatum^[4]

Class	Description
A	Proximal chamber receives all the pulmonary veins; distal chamber contains the left atrial appendage and the mitral valve. There is no atrial septal defect.
A1	Atrial septal defect between right atrium and proximal chamber.
A2	Atrial septal defect between right atrium and distal chamber.
B	Pulmonary veins drain into the coronary sinus.
C	No anatomic connection between the pulmonary veins and the proximal chamber.



DISCUSSION

Cor triatriatum sinister is a highly rare and surgically treatable congenital defect. It is slightly more common among men than women, with a men:women ratio of 1.5:1.^[4] No genetic predisposition has been linked to this particular anomaly. The clinical features on presentation can mimic those of mitral stenosis, supralvalvular mitral ring, or pulmonary venous stenosis. These entities share a common hemodynamic pathophysiology of obstruction between the pulmonary venous system and left heart chambers. The most common presenting symptoms are dyspnea, hemoptysis, orthopnea, and chest pain.^[5] The defect generally

manifests during infancy and early childhood. However, some cases will not exhibit any symptoms well into adulthood. Previous studies have suggested that the defect might be caused by the gradual narrowing and fenestration of the atrial membranes.^[5]

Several techniques have been used to establish the diagnosis, such as TTE, transesophageal echocardiography, catheter angiography, CT, and magnetic resonance imaging (MRI). The use of CT presents the risk of radiation, while transesophageal echocardiography brings discomfort associated with intubation. When compared with echocardiography and catheter angiography, MRI was found to have a higher detection rate.

Echocardiography provides an effective non-invasive method for diagnosing physiological and anatomical cardiac abnormalities, including anomalies associated with the intra-atrial area.^[6] Though MRI is appropriate for adults, it may not be the most appropriate choice for sick newborns, infants, and children, as they usually require intubation and general anesthesia. It has been previously shown that methods such as angiography or catheterization are not sufficient for ensuring the proper diagnosis of obstruction in the left atrial inflows.^[7] However, they are seldom necessary when there is suspicion of irreversible pulmonary vascular disease. While the anatomy was delineated with TTE in all cases, CT scan was performed in 2 patients (Patients 1 and 2) for differential diagnosis.

Since the first surgical approach in 1956 by Lewis et al., surgery is the treatment of choice in symptomatic patients with cor triatriatum sinister.^[8] The surgical approach consists of left or right atriotomy, excision of the obstructing membrane, and repair of the associated intracardiac anomalies. Among the patients in the current study, both right and left atrial incisions were used. ASD was an associated cardiac defect and repaired concomitantly. There were no instances of recurrent constriction after surgical treatment of the cor triatriatum membrane. Throughout the follow-up period, the patients remained symptom free.

As surgical repair is an easy and definitive treatment choice, cor triatriatum sinister should be considered in patients showing left heart chamber obstruction symptoms. Currently, diagnosis can be achieved per echocardiography. Surgical intervention resolves the obstructed membrane-related pathologies and other associated intracardiac defects immediately. In the surgical management of this easily and fully treatable congenital cardiac anomaly, it is difficult to de-

termine which atriotomy approach is comparatively more advantageous. However, in the management of this anomaly, priority should be given to confirmation of the diagnosis and full resection of the membrane. Thus, the surgeon should not hesitate to perform additional incisions if deemed necessary.

Conflict-of-interest issues regarding the authorship or article: None declared

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Keywords: Cardiovascular diseases; cor triatriatum; echocardiography.

Anahtar sözcükler: Kardiyovasküler hastalıklar; kor triatriatum; eko-kardiyografi.