Cor triatriatum sinister: a case series

Kor triatriatum sinister: Olgu serisi

Onur Işık, M.D., Muhammet Akyüz, M.D., Mehmet Fatih Ayık, M.D., Ertürk Levent, M.D.,[#] Yüksel Atay, M.D.

Department of Cardiovascular Surgery, Ege University Faculty of Medicine, İzmir, Turkey
[#]Department of Pediatric Cardiology, Ege University Faculty of Medicine, İzmir, Turkey

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.

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 konulalabilir. 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 tıkayıcı 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.

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

Abbreviations:

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

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

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

Correspondence: Dr. Muhammet Akyüz. Ege Üniversitesi Tıp Fakültesi, Kalp ve Damar Cerrahisi Anabilim Dalı, 35100 Bornova, İzmir, Turkey.
Tel: +90 232 - 444 13 43 e-mail: drmak100@gmail.com

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

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 appendage 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 concomitant 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.

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

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.

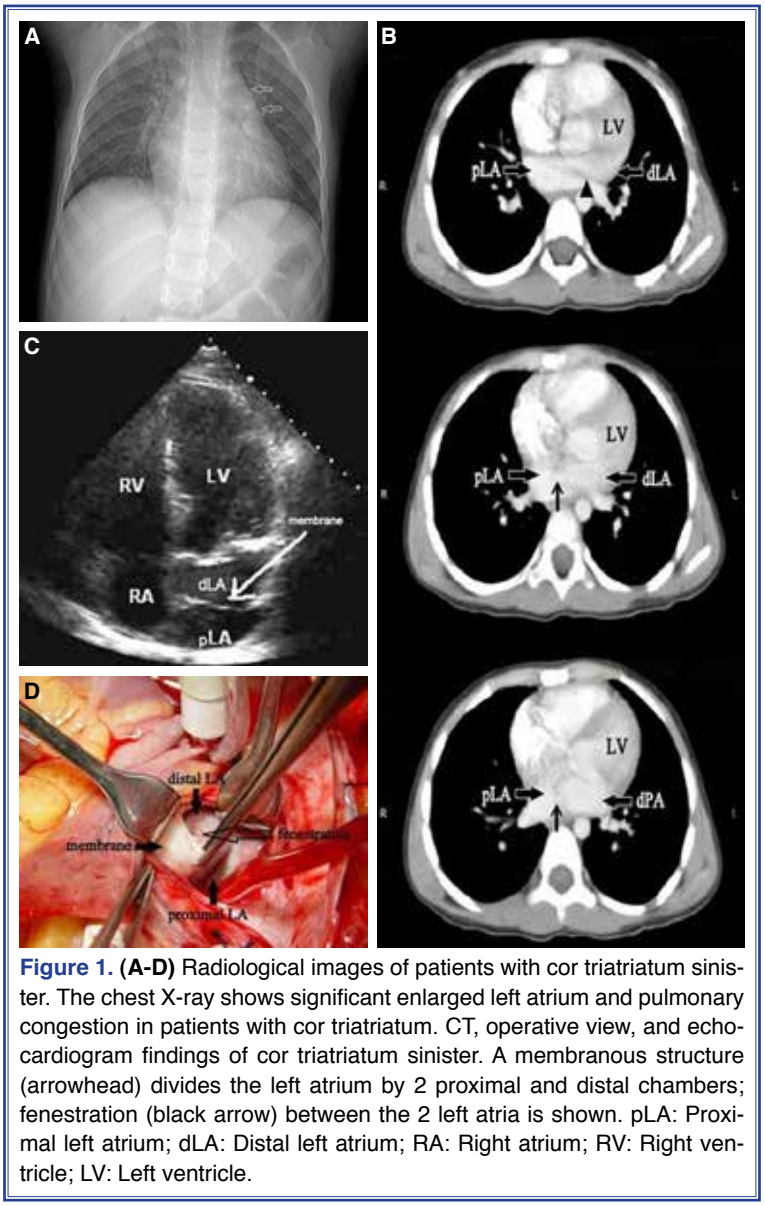

Figure 1. (A-D) Radiological images of patients with cor triatriatum sinister. The chest X-ray shows significant enlarged left atrium and pulmonary congestion in patients with cor triatriatum. CT, operative view, and echocardiogram findings of cor triatriatum sinister. A membranous structure (arrowhead) divides the left atrium by 2 proximal and distal chambers; fenestration (black arrow) between the 2 left atria is shown. pLA: Proximal left atrium; dLA: Distal left atrium; RA: Right atrium; RV: Right ventricle; LV: Left ventricle.

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.

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, supravalvular 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 determine 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

REFERENCES

1. Wolf WJ. Diagnostic features and pitfalls in the two-dimensional echocardiographic evaluation of a child with cor triatriatum. Pediatr Cardiol 1986;6:211–3. CrossRef
2. Salomone G, Tiraboschi R, Bianchi T, Ferri F, Crippa M, Parenzan L. Cor triatriatum. Clinical presentation and operative results. J Thorac Cardiovasc Surg 1991;101:1088–92.
3. Lam CR, Green E, Drake E. Diagnosis and surgical correction of 2 types of triatrial heart. Surgery 1962;51:127–37.
4. Su CS, Tsai IC, Lin WW, Lee T, Ting CT, Liang KW. Usefulness of multidetector-row computed tomography in evaluating adult cor triatriatum. Tex Heart Inst J 2008;35:349–51.
5. Krasemann Z, Scheld HH, Tjan TD, Krasemann T. Cor triatriatum: short review of the literature upon ten new cases. Herz 2007;32:506–10. CrossRef
6. Shuler CO, Fyfe DA, Sade R, Crawford FA. Transesophageal echocardiographic evaluation of cor triatriatum in children. Am Heart J 1995;129:507–10. CrossRef
7. Erden EC, Erden I, Kayapinar O. Cor triatriatum sinister with significant pressure gradient in an adult patient. Turkish J Thorac Cardiovasc Surg 2013;21:143–5. CrossRef
8. Lewis FJ, Varco RL, Taufic M, Niazi SA. Direct vision repair of triatrial heart and total anomalous pulmonary venous drainage. Surg Gynecol Obstet 1956;102:713–20.

Keywords: Cardiovascular diseases; cor triatriatum; echocardiography.

Anahtar sözcükler: Kardiyovasküler hastalıklar; kor triatriatum; ekokardiyografi.