ARCHIVES OF THE TURKISH SOCIETY OF CARDIOLOGY

Official journal of the TURKISH SOCIETY OF CARDIOLOGY

Evaluation of Pulmonary Hypertension with Exercise Right Heart Catheterization in an Adult Patient with Cor Triatriatum Sinister

Kor Triatriatum Sinisterli Erişkin Bir Hastada Egzersiz Sağ Kalp Kateterizasyonu ile Pulmoner Hipertansiyonun Değerlendirilmesi

ABSTRACT

Cor triatriatum sinister (CTS) is a rare adult congenital heart disease. The usual presentation may vary according to the size of the hole in the membrane in the left atrium and the pressure gradient. In addition to acute clinical presentations including acute pulmonary edema and sudden cardiac death, patients may present with chronic findings such as right heart failure due to pulmonary hypertension. The development of pulmonary hypertension is an important indicator of mortality. In cases where non-invasive methods are not sufficient for the diagnosis of pulmonary hypertension, exercise right heart catheterization may also be used. We present a patient with CTS, in whom the final decision was made with the help of an exercise right heart catheterization.

Keywords: Congenital heart malformation, cor triatriatum, echocardiography, exercise right heart catheterization, pulmonary hypertension

ÖZET

Cor triatriatum sinister erişkinlerde nadir görülen doğuştan kalp hastalığıdır. Hastaların genel klinikleri sol atriyumdaki zardaki deliğin boyutuna ve basınç gradiyentine göre değişiklik göstermektedir. Hastalar akut pulmoner ödem ve ani kardiyak ölüm gibi akut klinik tabloların yanı sıra pulmoner hipertansiyona bağlı sağ kalp yetmezliği gibi kronik bulgularla başvurabilirler. Pulmoner hipertansiyon gelişimi mortalite için önemli bir göstergedir. Pulmoner hipertansiyon tanısında girişimsel olmayan yöntemlerin yeterli olmadığı durumlarda egzersiz sağ kalp kateterizasyonu da kullanılabilir. Egzersiz sağ kalp kateterizasyonu ile pulmoner hipertansiyon tanısı konulan kor triatriatum sinister bir hastayı sunuyoruz.

Anahtar Kelimeler: Konjenital kalp malformasyonu, kor triatriatum, ekokardiyografi, egzersiz sağ kalp kateterizasyonu, pulmoner hipertansiyon

CASE REPORT
OLGU SUNUMU

Emre Aslanger[1]
Dursun Akaslan[1]
Hatice Kaya[2]
Ahmet Cem Nizam[1]
Redwan Seid Busery[1]
Bülent Mutlu[1]

[1]*Department of Cardiology, Marmara University, Pendik Training and Research Hospital, Istanbul, Türkiye*
[2]*Department of Radiology, Marmara University, Pendik Training and Research Hospital, Istanbul, Türkiye*

Corresponding author:
Dursun Akaslan
✉ dursun_akaslan@yahoo.com

Received: March 23, 2023
Accepted: August 31, 2023

Cite this article as: Aslanger E, Akaslan D, Kaya H, Nizam AC, Busery RS, Mutlu B. Evaluation of pulmonary hypertension with exercise right heart catheterization in an adult patient with cor triatriatum sinister. *Turk Kardiyol Dern Ars.* 2023;51(7):502-506.

DOI:10.5543/tkda.2023.36177

Cor triatriatum sinister (CTS) is a rare congenital anomaly, which is characterized by the division of the left atrium (LA) into two separate chambers by a membrane, due to the failure of the main pulmonary vein resorption during embryogenesis.[1] It has a wide variety of clinical manifestations, depending on the degree of obstruction and the presence of associated cardiac defects. The most common associated cardiac anomaly is mitral regurgitation, which is followed by atrial septal defect or patent foramen ovale, persistent left vena cava superior, and partial abnormal pulmonary venous return.[2]

The diagnosis is usually made in infancy or childhood. In this age group, CTS results in death in 75% of patients if left untreated.[3] Although as much as 17.5% of adult patients can be diagnosed incidentally, symptoms may range from mild exertional dyspnea, orthopnea, and palpitations to atrial fibrillation, thromboembolic events, and frank right heart failure due to the development of pulmonary hypertension.[4] As the risk of mortality sharply increases when cor triatriatum is complicated by pulmonary hypertension, early diagnosis of pulmonary hypertension is essential. However, at the earlier stages, pulmonary hypertension may only become manifest during exercise.[5]

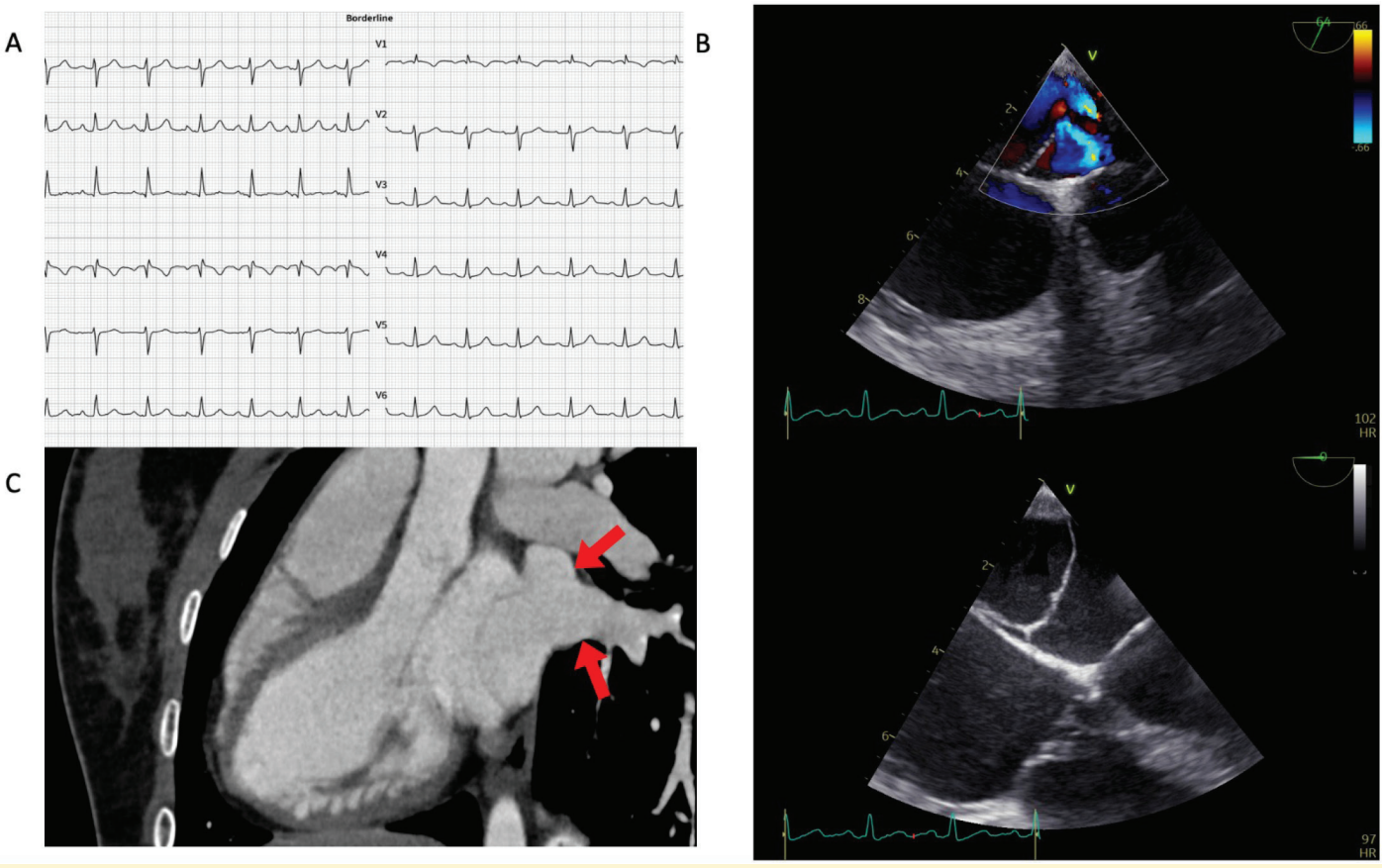

Figure 1. Electrocardiogram (ECG), transesophageal echocardiography (TEE), and cardiac computerized tomography (CT) images. ECG demonstrates sinus rhythm and right ventricular hypertrophy (A), and TEE confirms the presence of a multi-fenestrated membrane in the left atrium (LA) separating dilated four pulmonary veins and proper left atrium (B). Cardiac CT confirmed all pulmonary veins (the openings of two left-sided pulmonary veins are indicated by red arrows) open into the posterior superior chamber (C).

In this case report, we present a case with CTS and mild symptoms, in whom we used exercise right heart catheterization (RHC) to investigate the presence of pulmonary hypertension.

Case Report

A 19-year-old female patient without any prior cardiac disease presented with mild exertional dyspnea, which she had since she was 10 years old, but progressed in the past 6 months. Her family history and physical examination were unrevealing. Her electrocardiogram demonstrated sinus rhythm and right ventricular hypertrophy (Figure 1A), ambulatory rhythm Holter monitoring was normal except rare atrial and ventricular premature beats. Transthoracic echocardiography revealed a septation in the LA, mild mitral and tricuspid regurgitation (with a peak regurgitation velocity of 2.6 ms^{-1}), borderline dilated right atrium and right ventricle. Transesophageal echocardiography confirmed the presence of a multi-fenestrated membrane in the LA separating dilated four pulmonary veins and proper LA, and demonstrated an intact interatrial septum (Figure 1A). Cardiac computed tomography confirmed that all pulmonary veins open into the posterior superior chamber (Figure 1C). Cardiac magnetic resonance imaging showed the mobile membrane in the LA, hypertrophy, and dilatation in the right ventricle (Supplemental Video 1).

To elucidate the hemodynamic effect of CTS, we performed a RHC. After resting measurements, which revealed a 9 mmHg pulmonary capillary wedge (PCWP) to the left ventricular end-diastolic gradient, the patient was taken to the bicycle ergometer device. A staged protocol with 20 Watt increments in every 3 min was used. Confirming PCWP and diastolic pulmonary pressures were similar, the pulmonary arterial catheter was pulled into the main pulmonary artery and monitored as a surrogate for PCWP. Resting and exercise hemodynamic measurements were presented in Table 1. Pulmonary artery pressure traces during exercise are given in Figure 2.

ABBREVIATIONS

CO	Cardiac output
CTS	Cor triatriatum sinister
LA	Left atrium
mPAP	Mean pulmonary artery pressure
PCWP	Pulmonary capillary wedge pressure
RHC	Right heart catheterization

Table 1. Exercise Right Heart Catheterization Measurements

	Rest	1.Step (20 Watt)	2.Step (40 Watt)	Recovery
SBP, mmHg	120/75	155/93	152/90	114/70
SpO2, %	96	96	96	96
Pulse, beat/min	105	134	137	108
dPAP, mmHg	16	22	25	16
mPAP,mmHg	18	28	32	18
PVR, Woods	2	3	2	2
CO, $L.min^{-1}$	7.42	9.28	11.75	9.49
CI, $L.min^{-1}.m^2$	4.75	5.94	7.51	6.07
SV, mL	70.7	69.3	79.9	87.8
SVI, $mL.m^{-2}$	45.2	44.3	51.1	56.1

SBP, systemic blood pressure; dPAP, diastolic pulmonary arterial pressure; mPAP, mean pulmonary arterial pressure; PVR, pulmonary vascular resistance; CO, cardiac output; CI, cardiac index; SV, stroke volume; SVI, stroke volume index.

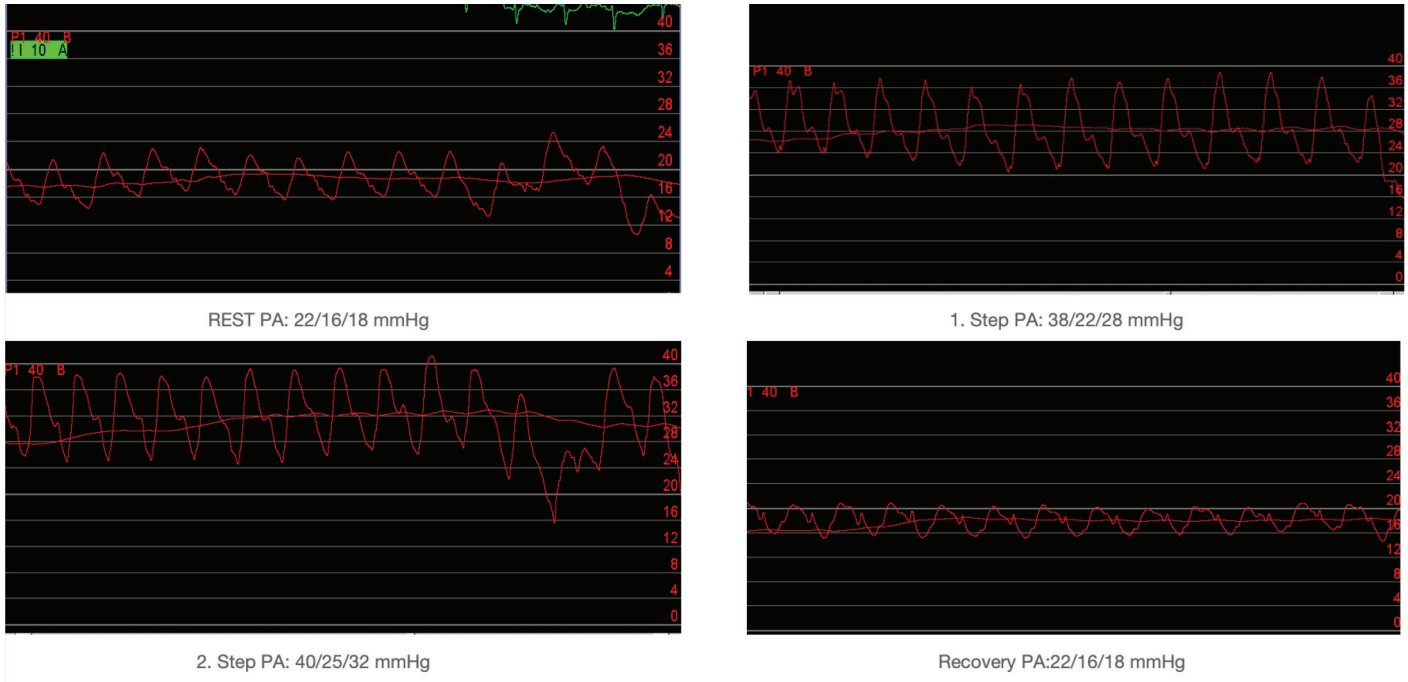

Figure 2. Pulmonary artery pressure tracings during exercise. Pulmonary artery pressure tracings were recorded at rest, exercise, and recovery periods. Since diastolic pulmonary arterial pressure (dPAP) and pulmonary capillary wedge pressure (PCWP) were found to be equal during resting recordings, the catheter was kept in the main pulmonary artery during exercise and dPAP was taken as a surrogate for PCWP.

With these data in hand, a comprehensive discussion was done with the patient and her family. The patient was referred to surgery for membrane resection. At the operation, the membranous structure, which is located 1 cm above the mitral valve, divides the LA into distal and proximal parts, and has an opening approximately 1 cm^2, was explored and removed by resection (Figure 3). An informed consent form was obtained for the use of the patient's clinical information in scientific settings.

Discussion

CTS is a rare congenital disease that may cause a wide range of presentations, from asymptomatic to death. Many of adult patients may be asymptomatic, especially those who have a >1 cm connection between two chambers, but sudden decompensation can occur even in these patients when a hyperdynamic physiology develops, such as pregnancy.[6] Symptoms occur earlier if the total connection area is less or CTS is associated with other malformations. In our case, the membrane defect was multi-

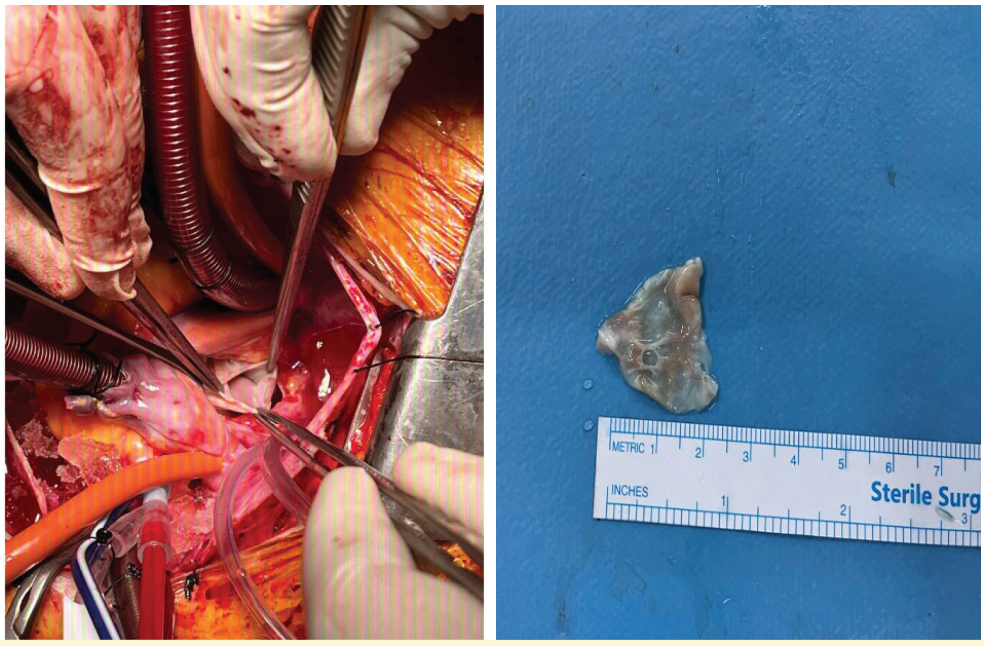

Figure 3. Multifenestrated appearance of the resected membranous structure after exploration of the left atrium. The membranous structure dividing the left atrium into distal and proximal parts was located 1 cm above the mitral valve, and its opening was approximately 1 cm².

fenestrated; therefore, it was hard to determine the total defect size, but due to the borderline increase in the right ventricle size, the presence of dilated pulmonary veins, and the significant resting gradient between PCWP and left ventricular diastolic pressure indicated an obstructive pathophysiology.

A comprehensive evaluation is needed in cases when the hemodynamic effect of the membrane is not clear. Transthoracic echocardiography is the method of choice and transesophageal echocardiography is recommended to evaluate associated congenital abnormalities, to elucidate the nature of connection through membrane, and to demonstrate pulmonary veins connections. Magnetic resonance imaging is the gold standard method in the evaluation of congenital heart diseases and can also be used for hemodynamic evaluation.[7] Invasive methods, such as RHC, are generally recommended for patients in whom a surgical intervention is planned, or who are considered to be at high risk for the development of pulmonary hypertension.[8] In cases in whom resting measurements are not conclusive, an exercise RHC is recommended. According to the guidelines, exercise pulmonary hypertension can be diagnosed if PCWP increases >25 mmHg or if the mean pulmonary artery pressure (mPAP) to cardiac output (CO) slope is >3 $mmHg.L^{-1}.min^{-1}$. In our patient, increased PCWP up to 25 mmHg and steep mPAP to CO slope approaching to 3.44 $mmHg.L^{-1}.min^{-1}$ were suggestive of exercise pulmonary hypertension.

The pathophysiology of CTS is similar to mitral stenosis. Therefore, close follow-up of the development of the right heart failure and diuretic treatments are at the forefront in its medical treatment. It is also known that the risk of thromboembolic events increases and routine antiaggregant use is recommended by some authors. In addition, there are a few case reports on the use of sildenafil in patients with increased PVR values. The definitive treatment of CTS is surgical resection. Since it was first performed in 1955, it has been performed with a high success and low complication rate.[9] Surgery is generally recommended in symptomatic patients with a gradient above 8 mmHg on the membrane.[10] Although anecdotal cases treated with balloon dilatation or radiofrequency ablation were reported, these procedures cannot be routinely recommended.[11] It should be bear in mind that even apparently non-obstructive cases can deteriorate with a hyperdynamic circulation and thromboembolic events are always a risk.

Conclusion

Although CTS is a rare adult congenital heart disease, it has a broad-spectrum of manifestations that can be acute, including acute pulmonary edema and sudden cardiac death, and chronic including right heart failure and severe pulmonary hypertension. Early diagnosis of obstructive CTS is important to prevent long-term complications due to pulmonary vascular disease and to differentiate it from other causes of left atrial inflow obstruction. Exercise RHC can be helpful in cases when the hemodynamic effect of CTS is not clear.

Informed Consent: Informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - E.K.A, D.A.; Design - E.K.A, D.A.; Supervision - B.M.; Materials - D.A., A.C.N., R.S.B., H.K.; Data Collections and/or Processing - A.C.N., H.K., R.S.B.; Analysis and/or Interpretation - E.K.A.; Literature Review - D.A.; Writing - E.K.A., D.A.; Critical Review - B.M.

Conflict of Interest: No conflict of interest disclosure has been received from the authors.

Funding: The authors declared that this study received no financial support.

Video Supplement 1-2: Cardiac magnetic resonance imaging showed the mobile membrane in the left atrium, and hypertrophy and dilatation of the right ventricle.

References

1. Church WS. Congenital malformation of heart: Abnormal septum in the left auricle. *Trans Pathol Soc Lond.* 1868;19:188–190.
2. Al Kindi HN, Shehata M, Ibrahim AM, et al. Cor triatriatum sinister (divided left atrium): Histopathologic features and clinical management. *Ann Thorac Surg.* 2020;110(4):1380-1386. [CrossRef]
3. Moss AJ, Adams FH. Stenosis of the common pulmonary vein: cor triatriatum. In: Moss AJ, Adam FH (eds). *Heart Disease in Infants, Children, and Adolescents*. Baltimore: Williams and Wilkins; 1989:863–868.
4. Rudienė V, Hjortshøj CMS, Glaveckaitė S, et al. Cor triatriatum sinistrum diagnosed in the adulthood: a systematic review. *Heart.* 2019;105(15):1197-1202. [CrossRef]
5. Kovacs G, Herve P, Barbera JA, et al. An official European Respiratory Society statement: pulmonary haemodynamics during exercise. *Eur Respir J.* 2017;50(5):1700578. Erratum in: *Eur Respir J.* 2018;51(1). [CrossRef]
6. Karas BJ, Ramirez E, Humbarger C, Yaacoub A. Malignant pulmonary edema in a pregnant woman caused by cor triatriatum. *Obstet Gynecol.* 2013;122(2 Pt 2):505-507. [CrossRef]
7. Sakamoto I, Matsunaga N, Hayashi K, Ogawa Y, Fukui J. Cine-magnetic resonance imaging of cor triatriatum. *Chest.* 1994;106(5):1586-1589. [CrossRef]
8. Humbert M, Kovacs G, Hoeper MM, et al.; ESC/ERS Scientific Document Group. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J.* 2022;43(38):3618-3731. Erratum in: *Eur Heart J*. 2023;44(15):1312. [CrossRef]
9. 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(6):713–720.
10. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: executive summary: A report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines. *J Am Coll Cardiol.* 2019;73(12):1494-1563. Erratum in: *J Am Coll Cardiol.* 2019;73(18):2361.
11. Tuan SH, Chen GB, Chen CH, *et al*. Serial cardiopulmonary exercise testing in an asymptomatic young female receiving percutaneous balloon dilatation for cor triatriatum sinistrum at an early age. *J Form Med Assoc.* 2021;120(4):1143–1147. [CrossRef]