A Very Rare Case of Co-Existence of Cor Triatriatum Sinister and Left Pulmonary Vein Atresia

Mustafa Aparci, Murat Yalcin, Zafer Isilak, Mehmet Dogan and Ejder Kardesoglu

Cor triatriatum sinister (CTS) is a rare congenital abnormality. Clinical presentation of patients with CTS mainly depends on the anatomic features of membrane and may vary from mild or moderate symptoms mimicking mitral stenosis to more severe and complicated cardioembolic stroke or a new onset heart failure. We herein have reported on a young male who presented with the signs and symptoms of mitral stenosis and was diagnosed as CTS with gradient on the orifice of the membrane after transthoracic echocardiography. Computerized tomographic angiography revealed that the patient had unilateral, left sided pulmonary arterial hypoplasia and pulmonary vein atresia, with only the right pulmonary veins draining into the left atrial chamber. Further cardiac imaging by either computed tomography or magnetic resonance imaging is necessary in order to seek accompanying cardiac and vascular abnormalities. Patients with CTS have improved short and long term survival rates if CTS and accompanying abnormalities are surgically treated before the disease is complicated with heart failure, pulmonary hypertension, stroke and etc.

Key Words: Cor triatriatum sinister • Mitral stenosis • Pulmonary artery hypoplasia • Pulmonary vein atresia

INTRODUCTION

Cor triatriatum sinister (CTS) is generally recognized to be a rare congenital abnormality. Clinical presentation of patients with CTS may vary from mild or moderate symptoms mimicking mitral stenosis to more severe and complicated cardioembolic stroke or a new onset heart failure.1,2

A membrane separates the left atrium into two chambers: the proximal or superior chambers with pulmonary venous blood flow, and the distal inferior chamber-true atrium is surrounded by the mitral valve, atrial appendage, and interatrial septum. An orifice on the membrane connects those two chambers and transfers the pulmonary venous blood to the systemic circulation through anatomical left atrium and to the mitral valves. Either CTS or its coexistence with pulmonary vein atresia and also pulmonary artery hypoplasia are very rare congenital abnormalities. Clinical signs and symptoms of CTS depend on the obstruction of the hole on the membrane and accompanying congenital abnormalities of the central and pulmonary vasculatures. Treatment of CTS is most effectively achieved through surgical excision of this membrane; however, in a limited number of cases, percutaneous balloon dilatation of the orifice (as in mitral stenosis) has been reported.2

CASE REPORT

A 19-year-old male presented with recent onset dyspnea on exercise. Upon physical examination, the patient’s blood pressure was 110/65 mm Hg, heart rate was 85 beats/min, and a 1/4 grade diastolic rumble is auscultated at the cardiac apex. Following transthoracic...
echocardiography examination, apical four chamber view exactly demonstrated the presence of membrane separating the left atrium into proximal and distal chambers. Interpretation of the color flow Doppler ascertained the location of the orifice on the membrane, communicating the chambers and continuous wave with Doppler echocardiography, and measured a 20 mmHg maximum peak gradient on the orifice (Figure 1). Since the orifices of pulmonary veins draining into the left atrium could not be observed on transthoracic echocardiography, computed tomography (CT) angiography was planned to evaluate the pulmonary arterial tree and pulmonary venous return to the left atrium.

CT angiography documented that the left pulmonary artery was hypoplastic, and the left pulmonary veins were atresic, with no pulmonary venous return to the left atrium. There were only right pulmonary veins draining to the left atrium, and increased vasculature and enlargement of right sided pulmonary vessels. Additionally, cardiac shifting to the left pulmonary space with reduced vasculature was remarkable (Figure 2).

The patient decided to be treated with surgical resection of the membrane lying within the left atrium.

DISCUSSION

Clinical manifestations of patients with CTS vary from dyspnea, recurrent syncope to stroke, heart failure, pulmonary edema, severe pulmonary arterial hypertension, and atrial fibrillation. The signs and symptoms of CTS are mainly associated with the pulmonary

![Figure 1](image1.png)

**Figure 1.** (A) Chest x-ray shows left-sided cardiac displacement and the engorgement of right sided pulmonary hilus. (B) Transthoracic echocardiography indicates the membrane which divides the left atrium. (C) Continuous wave Doppler image shows 20 mmHg maximum peak gradient on the orifice. LA, left atrium; PG, peak gradient; RA, right atrium.

![Figure 2](image2.png)

**Figure 2.** CT angiography revealed that the left pulmonary artery was hypoplastic (A), atresic left pulmonary veins with no pulmonary venous return to the left atrium (B) and increased vasculature and enlargement of right sided pulmonary veins (C). LA, left atrium; LPV, left pulmonary vein; RPV, right pulmonary vein.
congestion due to obstruction of pulmonary venous return
from the proximal atrial chamber to true left atrium and
late onset pulmonary arterial hypertension, which mimic
the clinical spectrum of mitral stenosis. Slight et al. re-
viewed all of the cases presented in the English-language
medical literature, and concluded that almost all the pa-
tients had manifested with the signs and symptoms of mi-
tral stenosis, increased pulmonary capillary wedge pres-
sure and mean pressure gradient, and also an increased
prevalence with aging. As in our case, continuous wave
Doppler echocardiography can demonstrate the increased
gradient on either diastolic or systolic phase of flow
through the orifice of the membrane opening to the true
left atrium (Figure 1C). We found that the max and mean
peak gradients were measured on the orifice of membrane
as 20 mmHg and 14 mmHg, respectively.

The patient’s chest x-ray revealed left-sided cardiac
displacement and the engorgement of right sided pul-
monary hilus (Figure 1A). Although echocardiography is
the best way to initially diagnose CTS, it cannot defini-
tively demonstrate all the congenital abnormalities ac-
companying CTS. A high quality image of membrane in
the left atrium on parasternal long axis view should be
evaluated from the apical views and by means of color
flow Doppler echocardiography. Orifices of pulmonary
veins should be sought in the proximal and distal-true
left atrium. Nevertheless, further cardiac imaging tech-
niques should be performed in order to identify and de-
scribe the exact anatomy. Furthermore, 2- or 3-dimen-
sional transesophageal echocardiography may also re-
veal the anatomy and structure of the membrane and
pulmonary venous return. CT angiography or magnetic
resonance imaging can detect all the components of
cardiac abnormalities related to the left atrium, pulmo-
nary venous return and pulmonary arterial tree, and
also pulmonary vasculature. We observed that only the
right pulmonary veins return to the left atrium, and left
pulmonary veins were absent. Since the left pulmonary
artery was hypoplastic, drainage of left-sided pulmonary
venous return had likely been achieved by mediastinal
or bronchial venous collaterals.

Cor triatriatum sinistra may be observed alone or in
association with other anomalies. In adults, the most
common accompanying anomalies are mitral regurgita-
tion, secundum-type atrial septal defect, persistent left
superior vena cava, and anomalies of PV return. Less
frequent concomitant anomalies are reported to be
hypoplastic left heart syndrome, patent ductus arteri-
osus, cor triatriatum dexter, aortic regurgitation, bicus-
pid aortic valve and tetralogy of Fallot. Several theories
have been suggested to better understand the formation
mechanism of CTS. Although there is no consensus on
such proposed mechanisms, the most accepted patho-
physiologic mechanism is failure of resorption of the
common PV in the intrauterin life. Proposed mecha-

CONCLUSIONS

Cor triatriatum sinistra is a rare congenital abnor-
mality, with varying signs and symptoms related to the
anatomic features membrane and the components of
the accompanying cardiac abnormalities. Further car-
diac imaging by computed tomography or magnetic
resonance imaging is necessitated to identify the co-
exiting cardiac and vascular abnormalities and to more
efficaciously guide the surgical treatment.

CONFLICT OF INTEREST

The authors declared no conflicts of interest.

REFERENCES


