

Atrial Fibrillation and Cor Triatriatum Sinister: A Case Report

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ABSTRACT

A 29-year-old man presented with palpitations, shortness of breath, and orthopnea. After being admitted, he progressed to cardiogenic shock and respiratory failure, which required ventilator support and cardioversion. Subsequent evaluation revealed a fibromuscular membrane across the left atrium, requiring urgent corrective surgery. This case report highlights the importance of identifying and treating the relatively rare cor triatriatum.

CASE REPORT

A 29-year-old man presented to the ambulatory clinic after 4 days of palpitations, shortness of breath, and orthopnea. Electrocardiography revealed atrial fibrillation with a rate of 181 beats per minute and a chest x-ray consistent with pulmonary edema. The patient was transferred to the Emergency Department and became progressively dyspneic. The initial attempt at controlling his heart rate by using a standard regimen of intravenous beta-blockers and calcium-channel blockers was ineffective. His condition subsequently deteriorated and progressed to cardiogenic shock with acute hypoxemic respiratory failure, which required ventilator support and emergent synchronized cardioversion at 200 Joules to restore normal sinus rhythm. A transthoracic echocardiogram was performed to assess whether left ventricular dysfunction or valvular abnormalities could be implicated in the patient's clinical deterioration. The echocardiogram revealed the presence of a linear density across the left atrium, which was consistent

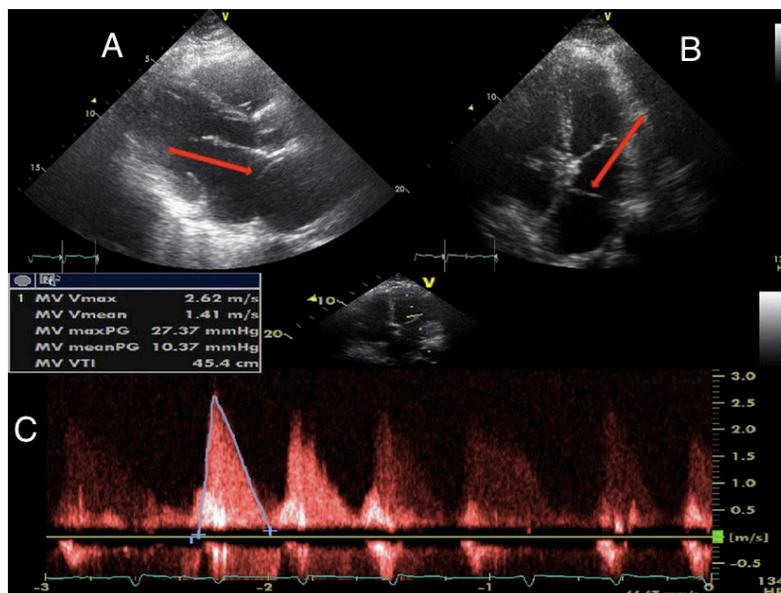


Figure 1A, 1B, and 1C. Transthoracic echocardiogram images of the left atrium. 1A) Parasternal long-axis view of the left atrium with the red arrow pointing to the cor triatriatum. 1B) Apical 4-chamber view of the left atrium with the red arrow pointing to the cor triatriatum. 1C) Inflow Doppler imaging at the mitral valve level highlighting an elevated peak gradient of 27 mm Hg and a mean gradient of 10 mm Hg (see outlined peak).

with cor triatriatum sinister (CTS) (Figures 1A and 1B). Echocardiographic Doppler measurements revealed an elevated peak and mean gradient of 27 mm Hg and 10 mm Hg, respectively, across the fibromuscular membrane (Figure 1C) with associated moderate-to-severe eccentric mitral regurgitation and an intact interatrial septum. The left ventricular ejection fraction was mildly reduced at 50% (normal for this patient would be > 55%).

The patient underwent urgent corrective cardiac surgery with resection of the fibromuscular septum to address the CTS, placement of a Number 30

Carpentier-Edwards Physio annuloplasty ring (Carpentier-Edwards, Irvine, CA) to address the dilatation of the mitral annulus, and a CryoMaze procedure to address the atrial fibrillation (Figures 2 and 3). The patient was postoperatively prescribed oral warfarin. A follow-up transthoracic echocardiogram 1 year after surgery demonstrated normal left ventricular systolic function and appropriate functioning of the mitral valve. Eighteen months after surgery, the patient remained asymptomatic and in normal sinus rhythm and was able to stop taking oral anticoagulant therapy.

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DISCUSSION

Cor triatriatum is a rare congenital cardiac anomaly represented by the development of a thick fibromuscular membrane that divides the left atrium (sinister) or the right atrium (dexter) into 2 chambers.^{1,2} CTS is much more common and occurs in approximately 0.1% to 0.4% of patients with congenital heart disease, whereas cor triatriatum dexter occurs in less than 8% of all cor triatriatum patients. CTS has a higher prevalence in males with a 1.5:1 male-to-female ratio. CTS is usually diagnosed in childhood; however, some cases, such the one reported here, may present in adulthood.³⁻⁵

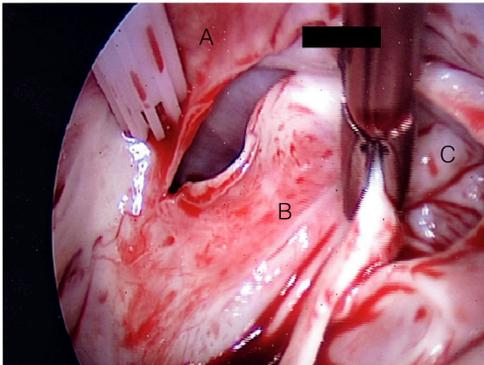


Figure 2. An intraoperative image of the left atrium of the heart: (A) left atrial wall; (B) fibromuscular membrane of cor triatriatum sinister; (C) view of the mitral valve through the foramen of the retracted fibromuscular membrane. The foramen measures 1 cm in length and 1.5 cm in width.



Figure 3. The specimen of the fibromuscular membrane after complete resection, measuring 5 cm in length and 7 cm in width.

According to a simple classification by Loeffler⁶ in 1949, there are three groupings of CTS. These are based on the size and number of fenestrations in the fibromuscular membrane. Group 1 has no fenestration, Group 2 has one or a few tiny fenestrations, and Group 3 has a single wide opening.⁶ Adults usually present as Group 3 because the other two groups become symptomatic early in life and are associated with a high early mortality.⁷

When CTS is diagnosed in adulthood, the patient may be asymptomatic with diagnosis made incidentally, or the patient may present with dyspnea, orthopnea, or hemoptysis.^{1,3,8} It is thought that patients become symptomatic owing to gradual fibrosis of the fenestrations in the fibromuscular membrane as well as development of valvular insufficiency and atrial arrhythmias.⁴ Classically, CTS is found in isolation; however, atypical CTS is associated with other anomalies, including secundum-type atrial septal defect, anomalous partial pulmonary venous connection, or mitral regurgitation, as in our patient's case.^{1,9}

CONCLUSION

The clinical features of CTS mimic mitral stenosis because of flow obstruction across the membrane, leading to an increase in pulmonary pressures and subsequently causing arrhythmias such as the atrial fibrillation found in our patient.¹⁰ On auscultation of the heart, one might appreciate a diastolic murmur similar to the diastolic murmur of mitral stenosis. However, the lack of an opening snap or a loud first heart sound distinguishes CTS from mitral stenosis. Surgical intervention involving removal of the fibromuscular membrane is usually reserved for cases with severe obstruction and has been shown to provide significant symptomatic relief as well as mortality benefit.^{9,11} ❖

Disclosure Statement

The author(s) have no conflicts of interest to disclose.

References

- Chen Q, Guhathakurta S, Vadalapali G, Nalladar Z, Easthope RN, Sharma AK. Cor triatriatum in adults: three new cases and a brief review. *Tex Heart Inst J* 1999;26(3):206-10.
- Sen T, Guray Y, Demirkan BM, Alioglu H, Korkmaz S. Cor triatriatum sinister in a 67-year-old man with atrial fibrillation. *Tex Heart Inst J* 2010;37(2):246-7.
- Slight RD, Nzewi OC, Buell R, Mankad PS. Cor-triatriatum sinister presenting in the adult as mitral stenosis: an analysis of factors which may be relevant in late presentation. *Heart Lung Circ* 2005 Mar;14(1):8-12. DOI: <http://dx.doi.org/10.1016/j.hlc.2004.10.003>.
- Orellana A, Orellana CE. Cor triatriatum. In: White CS, Haramati LB, Chen JJS, Levsky JM, editors. *Rotations in radiology: cardiac imaging*. New York, NY: Oxford University Press; 2014. p 120-2.
- Yarrabolu TR, Simpson L, Virani SS, Arora H, Navarajo J, Stainback RF. Cor triatriatum dexter. *Tex Heart Inst J* 2007;34(3):383-5.
- Loeffler E. Unusual malformation of the left atrium; pulmonary sinus. *Arch Pathol (Chic)* 1949 Nov;48(5):371-6.
- Nassar PN, Hamdan RH. Cor triatriatum sinisterum: classification and imaging modalities. *Eur J Cardiovasc Med* 2011 Jan;1(3):84-7. DOI: <http://dx.doi.org/10.5083/ejcm.20424884.21>.
- Troxclair D, Ross KF, Newman WP 3rd. Cor triatriatum sinisterum: a rare congenital cardiac anomaly presenting in an adult with chronic atrial fibrillation. *Am J Forensic Med Pathol* 2005 Sep;26(3):282-4. DOI: <http://dx.doi.org/10.1097/01.paf.0000176278.76218.93>.
- Krasemann Z, Scheld HH, Tjan TD, Krasemann T. Cor triatriatum: short review of the literature upon ten new cases. *Herz* 2007 Sep;32(6):506-10. DOI: <http://dx.doi.org/10.1007/s00059-007-2882-6>.
- Rorie M, Xie GY, Miles H, Smith MD. Diagnosis and surgical correction of cor triatriatum in an adult: combined use of transesophageal echocardiography and catheterization. *Catheter Cardiovasc Interv* 2000 Sep;51(1):83-6. DOI: [http://dx.doi.org/10.1002/1522-726X\(200009\)51:1%3C83::AID-CCD19%3E3.0.CO;2-7](http://dx.doi.org/10.1002/1522-726X(200009)51:1%3C83::AID-CCD19%3E3.0.CO;2-7).
- Rodefeld MD, Brown JW, Heimansohn DA, et al. Cor triatriatum: clinical presentation and surgical results in 12 patients. *Ann Thorac Surg* 1990 Oct;50(4):562-8. DOI: [http://dx.doi.org/10.1016/0003-4975\(90\)90190-H](http://dx.doi.org/10.1016/0003-4975(90)90190-H).