Annals of Case Reports and Clinical Studies
Review Article (ISSN: 2834-5673)

Cor Triatriatum: A Rare Congenital Cardiac Anomaly—Current Concepts in Diagnosis and Management

Jyothsna Goranti[1*], Vishak Prakash[1], Sachin Sapkota[1], Vaishnavi Sabesan[1], Sindhu Goddanakoppal RAJENDRA[2], Mehran Abolbashari[3]

[1]Department of Internal Medicine, Texas Tech University Health Sciences Center El Paso, Paul L. Foster School of Medicine, El Paso, Texas, USA

[2]Assistant Professor, Department of Internal Medicine, Texas Tech University Health Sciences Center El Paso, Paul L. Foster School of Medicine, El Paso, Texas, USA

[3]Department of Cardiology, Center of the Heart, 1575 N Resler Dr, El Paso, Texas, USA

Citation: *Jyothsna Goranti, Vishak Prakash, Sachin Sapkota, Vaishnavi Sabesan, Sindhu Goddanakoppal RAJENDRA, Mehran Abolbashari. Cor Triatriatum: A Rare Congenital Cardiac Anomaly—Current Concepts in Diagnosis and Management. Ann Case Rep Clin Stud. 2026;5(2):1-8.*

Received Date: 13 February 2026; **Accepted Date:** 20 February 2026; **Published Date:** 21 February 2026

***Corresponding author:** Jyothsna Goranti, Department of Internal Medicine, Texas Tech University Health Sciences Center El Paso, Paul L. Foster School of Medicine, El Paso, Texas, USA

ABSTRACT

Cor triatriatum is a rare congenital cardiac malformation characterized by the presence of a fibromuscular membrane that divides the atrium into two chambers, interfering with normal cardiac blood flow. This anomaly is classified into cor triatriatum sinister, affecting the left atrium, and cor triatriatum dexter, affecting the right atrium. The clinical presentation varies widely, ranging from asymptomatic cases to severe manifestations such as pulmonary edema, heart failure, and death. Transthoracic and transesophageal echocardiography are the primary diagnostic modalities, while cardiac computed tomography and magnetic resonance imaging provide additional anatomical detail in complex cases. Surgical excision of the membrane remains the definitive treatment, although minimally invasive and catheter-based interventions are emerging as alternative options in selected patients, particularly adults. This review summarizes the etiology, anatomical variations, pathophysiology, clinical presentation, diagnostic approaches, prognosis, and management strategies of cor triatriatum, with emphasis on recent advances in imaging and treatment.

Keywords: Cor triatriatum; congenital heart disease; Echocardiography; Cardiac magnetic resonance imaging; Surgical management

INTRODUCTION

Cor triatriatum is a rare congenital cardiac anomaly characterized by the presence of a fibromuscular membrane that divides the atrium into two distinct chambers, potentially resulting in varying degrees of obstruction to blood flow. Depending on the degree of obstruction and associated cardiac anomalies, clinical presentation may range from an incidental finding to severe pulmonary venous hypertension, heart failure, or life-threatening complications, particularly in infancy [1].

Cor triatriatum accounts for approximately 0.1% to 0.4% of congenital cardiac anomalies and may be associated with other structural heart defects in up to 50% of cases, including tetralogy of Fallot, double outlet right ventricle, coarctation of the aorta, anomalous pulmonary venous return, atrial septal defect, ventricular septal defect, and atrioventricular septal defect [2]. In adults, however, cor triatriatum is more frequently an isolated finding.

Cor triatriatum sinister is more common than cor triatriatum dexter. In this condition, a proximal chamber receives pulmonary venous return, while a distal chamber communicates with the mitral valve. The morphology of the dividing membrane may vary, appearing diaphragm-like, band-like, or funnel-shaped, and may contain one or more fenestrations of variable size, which determine the degree of obstruction and timing of symptom onset [1,3].

Cor triatriatum dexter is significantly less common and results from persistence of the right sinus venosus valve, which divides the right atrium into two chambers. The proximal chamber receives venous blood from the vena cavae, while the distal chamber communicates with the tricuspid valve and right atrial appendage [4].

A theoretical variant, cor triatriatum intermedium, has been described in the literature as resulting from a persistent interseptovalvular space, although no confirmed clinical cases have been reported [5].

Although cor triatriatum accounts for only a small proportion of congenital heart defects, its clinical significance lies in its variable presentation, potential for delayed diagnosis, and the need to differentiate it from other conditions that mimic mitral stenosis or pulmonary venous obstruction [2,3]. Advances in echocardiography and multimodality cardiac imaging have significantly improved the recognition and characterization of this condition, facilitating earlier diagnosis and better treatment planning.

This review aims to provide a comprehensive overview of the embryology, anatomical variants, pathophysiology, clinical manifestations, diagnostic strategies, differential diagnosis, and current management approaches of cor triatriatum, with particular emphasis on emerging minimally invasive and catheter-based therapies.

Methods of Literature Search

A narrative review of the literature was conducted to summarize current knowledge on cor triatriatum. A systematic search was performed using PubMed, Google Scholar, and MEDLINE databases for articles published in English up to January 2025. The search terms included *"cor triatriatum," "cor triatriatum sinister," "cor triatriatum dexter," "congenital heart disease," "atrial membrane," "echocardiography,"* and *"surgical management."* Relevant original research articles, case reports, review articles, and clinical guidelines were screened. Reference lists of selected articles were also reviewed to identify additional relevant studies. Articles focusing on epidemiology, pathophysiology, clinical presentation, imaging, and management were included in this review.

Pathophysiology

Several embryologic theories have been proposed to explain the development of cor triatriatum. The malincorporation theory suggests incomplete incorporation of the common pulmonary vein into the left atrium during fetal development, resulting in a persistent membrane that divides the atrial chamber [6]. However, this theory does not fully explain the presence of atrial muscle fibers and the fossa ovalis in some cases [7].

The mal-septation theory proposes abnormal development of the septum primum, leading to formation of a fibromuscular membrane. Another hypothesis, the entrapment theory, suggests that the common pulmonary vein becomes entrapped in the embryonic sinus venosus, preventing its normal incorporation into the left atrium [1].

The hemodynamic consequences depend largely on the size and number of fenestrations within the membrane. Severe obstruction results in pulmonary venous hypertension, pulmonary edema, and right-sided heart failure, whereas mild obstruction may remain asymptomatic for years and present later in life [1].

Clinical Presentation

In neonates and infants, cor triatriatum sinister may present with severe symptoms when pulmonary venous outflow is significantly restricted. Clinical manifestations may include tachypnea, respiratory distress, poor feeding, failure to thrive, cyanosis, and pulmonary edema. Pulmonary hypertension and recurrent respiratory infections are also commonly reported [8-10].

In older children and adults, cor triatriatum is often diagnosed incidentally or during evaluation for unexplained dyspnea. Patients may present with exertional dyspnea, orthopnea, paroxysmal nocturnal dyspnea, and palpitations. Atrial fibrillation may develop due to chronic atrial dilation and pressure overload [10,16,17].

Complications

Complications of cor triatriatum may include pulmonary hypertension, right-sided heart failure, arrhythmias, thromboembolism, bacterial endocarditis, hepatic congestion, ascites, and death in untreated cases [1].

DIAGNOSIS

Transthoracic echocardiography is the primary diagnostic tool and allows visualization of the atrial membrane, measurement of gradients, and assessment of associated anomalies. Transesophageal and three-dimensional echocardiography provide improved visualization of membrane morphology and pulmonary venous drainage patterns [13,14].

Cardiac CT and MRI provide detailed anatomical evaluation and are particularly useful in complex congenital heart disease or when surgical planning is required [11,12].

Cardiac catheterization may be used to measure pulmonary artery pressures, assess shunt physiology, and evaluate the severity of obstruction in selected cases [11,12].

Differential Diagnosis

Conditions that should be considered in the differential diagnosis include supravalvular mitral stenosis, mitral stenosis, atrial septal defect, pulmonary venous hypertension, anomalous pulmonary venous return, atrial myxoma, and pulmonary arterial hypertension [1].

Management

Medical Management

Asymptomatic patients with minimal obstruction may be managed conservatively with regular follow-up. Medical therapy is primarily supportive and may include diuretics, rate or rhythm control, and anticoagulation in patients with atrial fibrillation [16-18].

Surgical Management

Surgical excision of the membrane is the definitive treatment for symptomatic patients. The procedure involves removal of the membrane and correction of associated defects when present. Long-term outcomes after surgery are generally favorable, particularly in isolated cases [21,22].

Emerging and Minimally Invasive Approaches

Recent advances in minimally invasive and catheter-based techniques have provided alternative treatment options in selected patients, particularly adults with favorable anatomy. These approaches aim to reduce procedural morbidity while maintaining effective relief of obstruction [23-25].

Prognosis

The prognosis of untreated severe cor triatriatum in infancy is poor, whereas outcomes following timely surgical intervention are excellent, especially in patients without associated congenital anomalies [1].

DISCUSSION

Surgical membrane excision remains the gold standard treatment for symptomatic cor triatriatum, with excellent long-term outcomes and low recurrence rates, particularly in isolated disease [21,22]. Surgical intervention allows complete removal of the obstructing membrane and correction of associated congenital anomalies when present.

Catheter-based balloon dilation and minimally invasive surgical techniques have emerged as alternative approaches in selected patients, especially adults with favorable anatomy and non-calcified membranes [23-25]. These less invasive procedures may reduce perioperative morbidity and shorten recovery time; however, long-term comparative data remain limited. Careful patient selection and individualized decision-making are therefore essential when considering catheter-based therapies.

Despite its rarity, cor triatriatum remains an important diagnostic consideration in patients presenting with unexplained pulmonary hypertension or symptoms resembling mitral stenosis.

A schematic overview of the anatomical variants, clinical presentation, and management strategies of cor triatriatum is shown in **(Figure 1)**.

A summary of the classification, clinical presentation, and management of cor triatriatum is presented in **(Table 1)**.

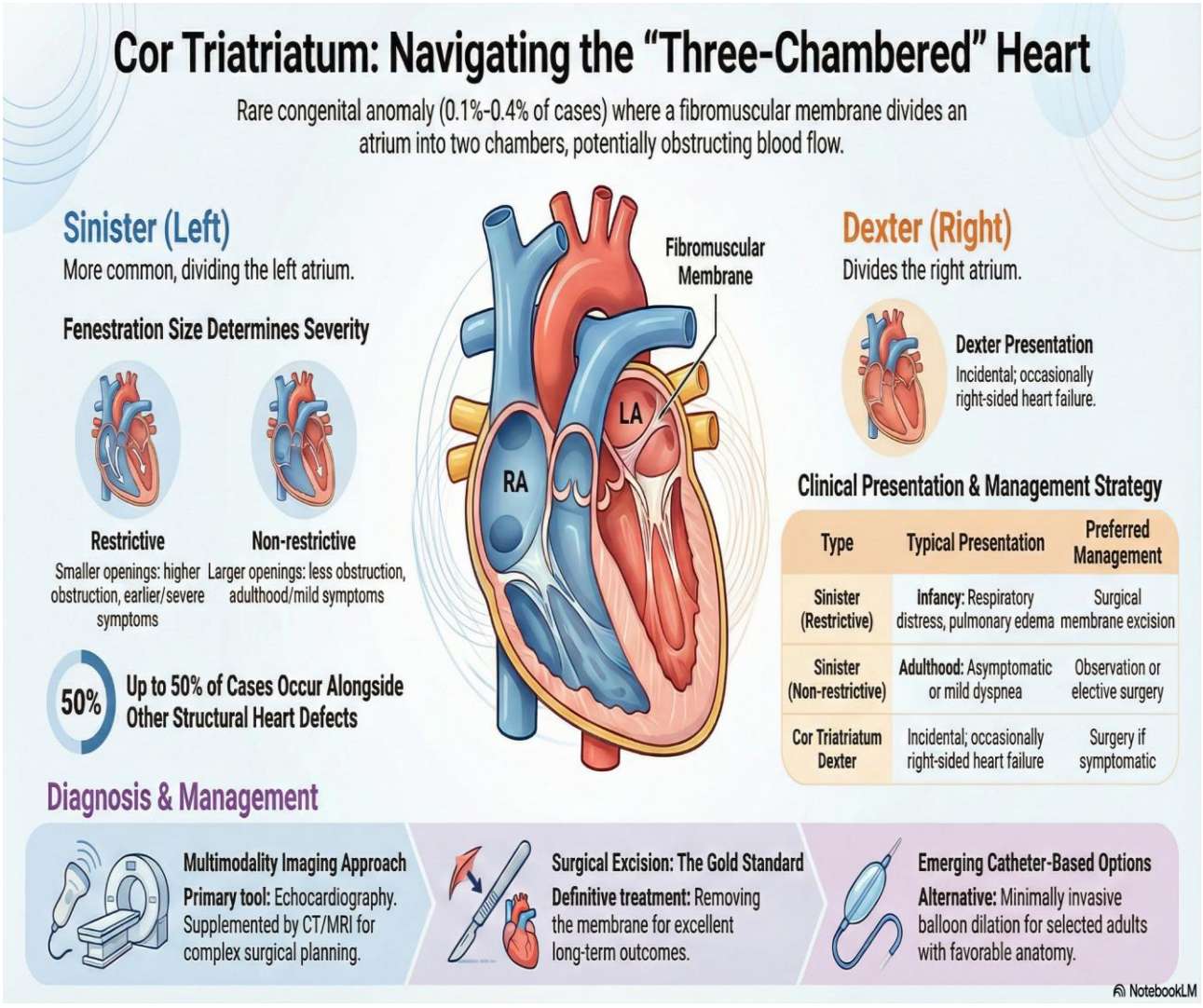

Figure 1: Schematic Representation of Cor Triatriatum Showing Anatomical Variants, Clinical Features, and Management Approaches

Table1. Classification, Clinical Features, and Management of Cor Triatriatum

Type	Anatomical Features	Typical Clinical Presentation	Preferred Management
Cor triatriatum sinister (restrictive)	Small or absent fenestrations causing significant obstruction to pulmonary venous flow	Infancy: respiratory distress, pulmonary edema, failure to thrive, pulmonary hypertension	Surgical membrane excision
Cor triatriatum sinister (non-restrictive)	Large fenestrations with minimal obstruction	Often asymptomatic or mild dyspnea in adolescence or adulthood	Observation or elective surgery if symptomatic
Cor triatriatum dexter	Persistence of right sinus venosus valve dividing right atrium	Often incidental; occasionally right-sided heart failure or arrhythmias	Surgery if symptomatic
Adult incidental cor triatriatum	Mild obstruction or calcified membrane	Dyspnea, atrial fibrillation, or incidental imaging finding	Individualized management; surgery or catheter intervention in selected cases

CONCLUSION

Cor triatriatum is an uncommon but clinically important congenital cardiac anomaly that may present across a wide age spectrum, from severe disease in infancy to incidental findings in adulthood. The degree of obstruction and the presence of associated congenital anomalies largely determine clinical presentation and prognosis. Early recognition through echocardiography and complementary imaging modalities is essential to guide timely intervention and prevent complications.

Surgical excision of the membrane remains the definitive treatment for symptomatic patients and is associated with excellent long-term outcomes. Minimally invasive and catheter-based interventions are emerging as promising alternatives in selected cases, although further studies are needed to define long-term outcomes and optimal patient selection.

Greater awareness of this rare entity among clinicians is important to ensure accurate diagnosis, appropriate management, and improved patient outcomes.

REFERENCES

1. Ather B, Meredith A, Siddiqui WJ. Cor Triatriatum. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024.
2. Ullah W, Sattar Y, Rauf H, Roomi S, Shah M. A systematic review of a long-forgotten cause of atrial fibrillation and stroke: cor triatriatum. Cureus. 2019;11(12):e6371.
3. Mashadi AH, Narasimhan SL, Said SM. Cor triatriatum sinister: Long-term surgical outcomes in children and a proposal for a new classification. J Card Surg. 2022;37(12):4526-4533.

4. Elagizi A, Marvin R, O'Bryan G, Vyas V, Arcement L. Three's a crowd—an extremely rare case of cor triatriatum dexter. J La State Med Soc. 2017;169(2):50-51.
5. Thilenius OG, Bharati S, Lev M. Subdivided left atrium: an expanded concept of cor triatriatum sinistrum. Am J Cardiol. 1976;37(5):743-752.
6. Parsons CG. Cor triatriatum: concerning the nature of an anomalous septum in the left auricle. Br Heart J. 1950;12(4):327-338.
7. Narayanapillai J. Cor triatriatum sinister with severe obstruction: a rare presentation in an adult. BMJ Case Rep. 2016;2016.
8. Goel A, Viswamitra S, Reddy BN, Gaduputi J. Computed tomography features of cor triatriatum: an institutional review. Br J Radiol. 2021;94(1119):20201252.
9. Kilkenny K, Frishman W. Cor triatriatum: a review. Cardiol Rev. 2023.
10. Ostman-Smith I, Silverman NH, Oldershaw P, Lincoln C, Shinebourne EA. Cor triatriatum sinistrum: diagnostic features on cross sectional echocardiography. Br Heart J. 1984;51(2):211-219.
11. Thakrar A, Shapiro MD, Jassal DS, Neilan TG, King ME, Abbara S. Cor triatriatum: the utility of cardiovascular imaging. Can J Cardiol. 2007;23(2):143-145.
12. Hamdan R, Mirochnik N, Celermajer D, Nassar P, Iserin L. Cor triatriatum sinister diagnosed in adult life with three-dimensional transesophageal echocardiography. BMC Cardiovasc Disord. 2010;10:54.
13. Slight RD, Nzewi OC, Mankad PS. Echocardiographic diagnosis of cor triatriatum sinister in the adult. Heart. 2004;90(1):63.
14. Fuchs MM, Connolly HM, Said SM, Egbe AC. Outcomes in patients with cor triatriatum sinister. Congenit Heart Dis. 2018;13(4):628-632.
15. Sentilhes L, Verspyck E, Bauer F, Marpeau L. Management of maternal cor triatriatum during pregnancy. Obstet Gynecol. 2004;104(5):1212-1215.
16. Hayes C, Liu S, Tam JW, Kass M. Cor triatriatum sinister: an unusual cause of atrial fibrillation in adults. Case Rep Cardiol. 2018;2018:9242519.
17. Park KJ, Park IK, Sir JJ, Kim HT, Park YI, Chung JM, et al. Adult cor triatriatum presenting as cardioembolic stroke. Intern Med. 2009;48(13):1149-1152.
18. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease. Circulation. 2019;139(14):e698-e800.
19. Shirani J. Cor Triatriatum. Medscape. Published December 18, 2020.
20. Al Kindi HN, Shehata M, Ibrahim AM, Roshdy M, Simry W, Aguib Y, Yacoub MH. Cor triatriatum sinister (divided left atrium): histopathologic features and clinical management. Ann Thorac Surg. 2020;110(4):1380-1386.

21. Ozyuksel A, Yildirim O, Avsar M, Hayirlioglu M, Demiroluk S, Kucukosmanoglu O, Bilal MS. Surgical correction of cor triatriatum sinister in the paediatric population: mid-term results in 15 cases. Eur J Cardiothorac Surg. 2015;47(1):e25-e28.
22. Silva López DE, Martinez GB, Ortiz Argüelles G, Campos Sánchez VY, Barrios Muñoz AM, González Cárdenas SA. Comprehensive insights into cor triatriatum: an overview of pathophysiology, clinical presentation, diagnostic strategies, and management options. Int J Med Sci Clin Res Stud. 2024;4(11):2173-2179.
23. Prousi G, Guthier J, Sumner A, et al. Cor triatriatum: the workup of a rare finding. J Am Coll Cardiol. 2018;71(11):A2529.
24. von Bartheld B, Rijnierse M, van Loon R, Meijboom L. A rare case of partial cor triatriatum diagnosed at an adult age: a case report. Eur Heart J Case Rep. 2023;7(3):ytad082.
25. Butt M, Halim H, Qureshi A, Koene R, Aung S, Karamlou T, Saini A. A tale of three chambers: cor triatriatum sinistrum. CASE (Phila). 2024;8(3):221-225.