



Cor Triatriatum: Preoperative Diagnosis and Successful Surgical Correction in a Four-Year-Old Girl

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Abstract

Cor triatriatum is defined as a membrane within the left atrium, which might lead to restricted pulmonary venous return. Diagnosis is usually achieved by echocardiography in early infancy. Therapy of choice is the excision of the membrane. Herein, successful correction of cor triatriatum in a 4-year-old girl is presented, and the clinical features, echocardiographic findings, and the surgical treatment are discussed.

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Introduction

Cor triatriatum is one of the most sparse congenital cardiac anomalies, accounting for approximately 0.1% of all congenital heart diseases.^{1,2} A classic form of cor triatriatum may be defined as the left atrium subdivided into dorsal and ventral chambers by a fibromuscular diaphragm; the dorsal chamber received the pulmonary veins and the ventral chamber was attached to the left atrial appendage and led to the mitral valve (Figure 1). The communication between the dorsal and ventral chambers is variable or absent and depends on the size of the opening in the fibromuscular "membrane".^{3,4}

Approximately in 75% of individuals, there exists an atrial septal defect (ASD) between the right atrium and the proximal venous chamber (subtype A1 according to the Lam classification) or the true distal atrium, as was the case in our patient.^{5,6}

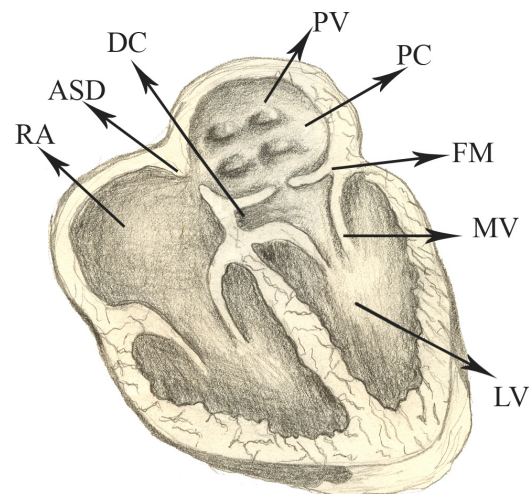


Figure 1. Schematic diagram of intraoperative finding of nearly atretic cor triatriatum and atrial septal defect

RA, Right atrium; ASD, Atrial septal defect; DC, Distal chamber; PV, Pulmonary veins entrance; PC, Proximal chamber; FM, Fibromuscular membrane; MV, Mitral valve; LV, Left ventricle; RV, Right ventricle

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The clinical symptoms depend on the size of the orifice (the classic case mimicking mitral stenosis) and the morphology of the defect, including the presence and location of the ASD and associated congenital heart disease. The diagnosis of cor triatriatum should be considered in patients with pulmonary congestion, pulmonary arterial hypertension, and pulmonary venous obstruction. Symptoms are usually not related to a specific anatomic subtype and comprise failure to thrive, tachypnea, dyspnea (as was the case in our patient), and occasionally, cyanosis. The latter can be caused by either the restricted pulmonary venous return (then associated with severe pulmonary symptoms like tachydyspnea) or the presence of severe stenosis and ASD; a right-to-left shunt might be present at atrial level.^{7, 8}

Previous studies have reported that cor triatriatum is usually diagnosed and surgically treated before 24 months old. Herein, we present a case of cor triatriatum diagnosed in a 4-year-old girl.

Case report

The patient was a 4-year-old girl, who was the fifth child of her family. Pregnancy and delivery were normal, birth weight was eleven pounds, the immediate postnatal period was event-free, and she was breastfed for two years. The patient was first hospitalized for one week at 2 months old due to severe respiratory tract infection. A diagnosis of congenital heart disease was made after a thorough examination, and treatment was offered for later.

At 4 years of age, the patient was admitted to our hospital due to tachypnea, early fatigue on exertion, and recurrent respiratory infections. Physical examination revealed a heart rate of 70-110 per minute, loud P₂, wide fixed splitting of S₂, systolic flow murmur along the left sternal border, and diastolic flow rumble across the tricuspid valve. Electrocardiography detected sinus rhythm, right-axis deviation, and incomplete right bundle branch block. Roentgenograms of the chest showed increased pulmonary vascular markings, prominence of the right ventricle, and main pulmonary artery comparable with atrial septal defect (ASD) findings. Transthoracic echocardiography was thereafter performed and demonstrated a membrane in the left atrium, dividing it into two chambers: the proximal part, which received the pulmonary veins, and the distal part, which was attached to the mitral valve and left atrial appendage (Figures 2 & 3). Indeed, there was a case of cor triatriatum with one reservoir in the posterior of the left atrium, receiving the pulmonary veins but without gradient in the left atrium. The size of the ASD/inter atrial septum was 23/34 mm, and left ventricular ejection fraction was 60%. In addition, there were secundum type ASD (Figure 4), dilated coronary sinus, elevated pulmonary artery pressure,

and severe right atrial and ventricular dilation.

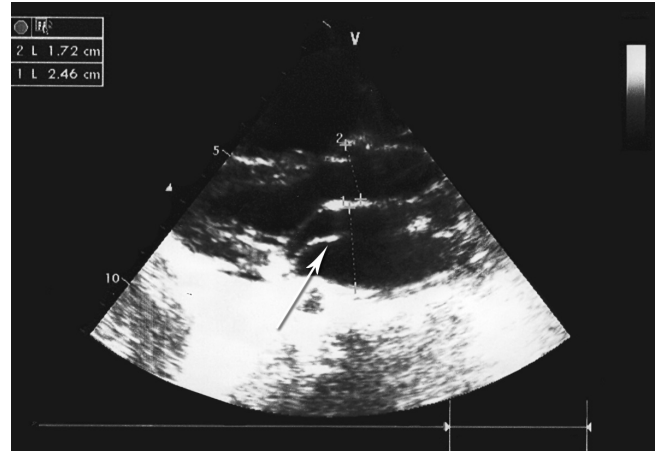


Figure 2. Parasternal long-axis view showing a membrane (arrow) in the left atrium

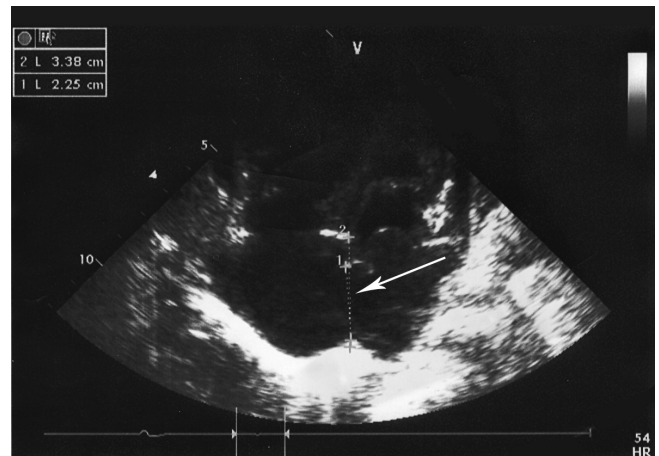


Figure 3. Atrial septal defect ostium secundum (arrow)

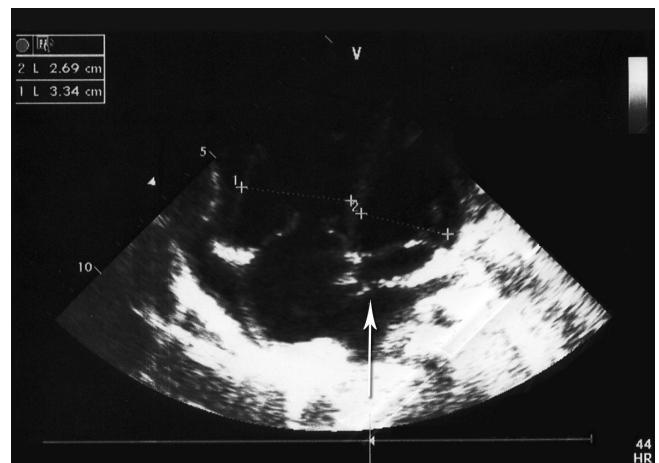


Figure 4. Membrane (arrow) separating the posterior chamber from the anterior one; the posterior space receiving the pulmonary veins and the anterior one being open to the mitral valve

A diagnosis of A1 type of cor triatriatum according to the Lam classification was established. The patient was, therefore,



transferred to the operating room. The heart was approached through a median sternotomy incision. Cardiopulmonary bypass (CPB) was initiated, and the aortic cross-clamp was performed under mild systemic hypothermia and cold blood cardioplegia. The cor triatrium membrane was thick with one orifice measuring about 2 mm; it was resected via a transeptal approach. The ASD (8 mm in diameter) was closed using a pericardial patch. The postoperative cardiac echocardiogram showed the status/post (S/P) ASD closure, the repair/removal of the cor triatrium, normal left ventricle size and function, severe right ventricle enlargement with mild right ventricular hypertrophy and good function, mild tricuspid regurgitation, mild pulmonary insufficiency, and trivial pericardial effusion. Additionally, there was no residual ASD or pulmonary stenosis or left atrium gradient. The postoperative course was event-free, and the patient was discharged on the 4th postoperative day.

Discussion

The youngest cor triatrium case was an 18-day-old patient who also had infracardiac total anomalous pulmonary venous drainage in the Baek MJ et al. study¹ and the oldest case was a 93-year-old patient in the Arciniegas E et al. study.⁹

In the Alphonso study 61% of the patients had ASD (Lam type A1/A2).⁵ Arciniegas and Richardson et al. reported 83% and 95% of ASD with cor triatrium.^{9,10} In the Krasemann et al. study, and there were two adult patients with an additional ASD, which was thermodynamically insignificant. In one of them, the closure of the defect was favored after a stroke to prevent possible right-to-left embolic events, and the membrane was resected during ASD repair. The other one had non-specific symptoms (decreased exercise tolerance), leading to diagnostic and, subsequently, surgical procedures.⁷

The treatment of cor triatrium is primarily surgical. There are only two reports of successful balloon catheter dilation of the communication between the proximal and distal chambers, and the follow-up periods for balloon dilatation are generally 3 and 12 months. The long-term outcome of balloon dilatation remains to be determined.⁵

According to the Alphonso study, the most current complications after surgery were supraventricular tachycardia and peritoneal dialysis.⁵ Other complications were seizure, pleural effusion requiring drainage, urinary tract infection, pyrexia of unknown origin, ventricular tachycardia, cerebrovascular accident, pneumothorax requiring drainage, acute renal failure, superficial wound infection requiring debridement, and pericardial effusion requiring drainage, which were seen in 10 (40%) patients. Seventeen (60%) patients made an uncomplicated recovery.⁵

Conclusion

In brief, cor triatrium is a rare congenital cardiac anomaly. Surgical correction offers good early and long-term results for both classic and atypical cases of cor triatrium.⁵

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