

Cor triatriatum in an 86-year-old woman: initial presentation with pulmonary hypertension discovered during preoperative evaluation

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ABSTRACT

Cor triatriatum is a congenital heart malformation that is characterised by the division of the left or right atrium into two separate chambers by a membrane or diaphragm. Reports among adults are scarce, as most cases are diagnosed during childhood. The risk of mortality is increased when cor triatriatum is complicated by pulmonary hypertension. This is a report of an 86-year-old woman with World Health Organization Group 2 pulmonary hypertension secondary to cor triatriatum, discovered during preoperative workup. Echocardiography showed a membrane dividing the left atrium into two. Doppler studies revealed a reversal of normal flow, similar to mitral stenosis. The right ventricle was dilated, with reduced long axis function.

Keywords: congenital heart malformation, cor triatriatum, echocardiography, pulmonary hypertension

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INTRODUCTION

Cor triatriatum was first reported in 1868.⁽¹⁾ It is a rare congenital anomaly of the heart that is characterised by the division of the left (cor triatriatum sinistrum) or right atrium (cor triatriatum dextrum) into two chambers by a fibromuscular band, membrane or diaphragm.⁽²⁾ This anomaly can be associated with other complex congenital anomalies, such as tetralogy of Fallot, persistent left superior vena cava with unroofed coronary sinus, ventricular septal defect, atrioventricular septal (endocardial cushion) defect and common atrioventricular canal.⁽³⁾

Cor triatriatum is extremely rare, with an incidence of 0.1%–0.4%.⁽⁴⁾ The morbidity and mortality rates are high in symptomatic infants due to the association with restrictive physiology at the membrane opening as well as the association with cyanotic and acyanotic congenital heart diseases.⁽⁵⁾ Adult cases are very rare;

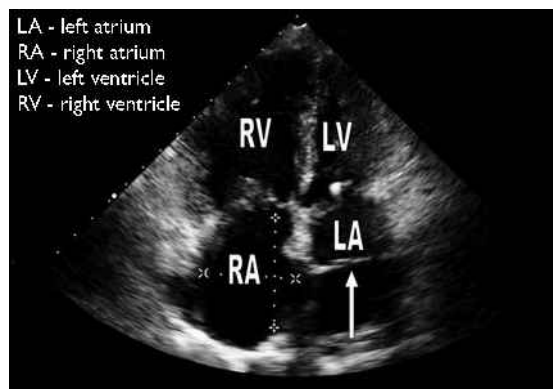


Fig. 1 4-chamber apical echocardiography image shows the left atrium divided by a membranous structure into an upper and lower division associated with floppy interatrial septum. Note the membrane and the opening of the membrane dividing the left atrium into two chambers (arrow).

most of the adults reported were in their fifth or sixth decade of life.⁽⁶⁻⁸⁾ In this report, an 86-year-old woman was coincidentally discovered to have cor triatriatum during routine preoperative evaluation.

CASE REPORT

An 86-year-old woman was referred to our institution as part of her preoperative review for echocardiographic examination. She was being prepared for excisional biopsy of an epithelioma near the right eye. She denied any past history of cardiovascular disease but reported mild dyspnoea on exertion, which was not profound enough to affect her lifestyle. She had no significant past medical or surgical history and was not a known hypertensive or diabetic. There was no family history of cardiovascular disease or stroke. She denied any past history of orthopnoea, bilateral leg swelling or cough. Examination revealed an elderly woman who was in good general physical condition. She had no clinical signs of dehydration, finger clubbing or bilateral pitting pedal oedema.

Two-dimensional echocardiography revealed a membrane-like, echo-dense structure partitioning the left atrium into two, suggesting a diagnosis of cor triatriatum associated with a floppy interatrial septum (Fig. 1). No

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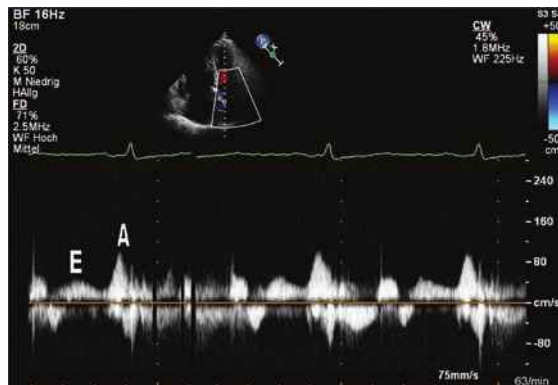


Fig. 2 Doppler echocardiography flow across the opening on the membrane shows reversed early and late atrial.

E: early atrial flow; A: late atrial flow

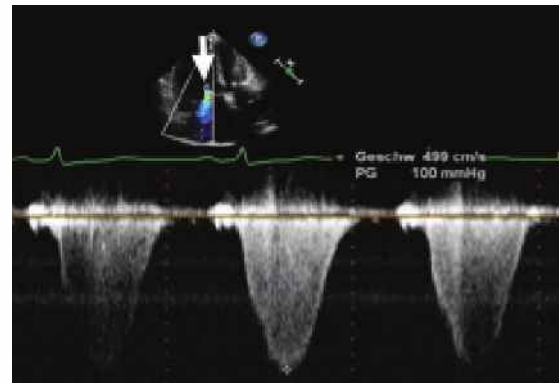


Fig. 3 Colour Doppler echocardiography image shows tricuspid regurgitation with elevated pulmonary systolic pressure. Note the tricuspid regurgitation flow with an estimated right ventricular systolic pressure of 100 mmHg (arrow).

atrial septal defect was demonstrated. The flow across the membrane-like structure was typically the reversed early (E)/late atrial (A) ratio (Fig. 2). There was no significant pressure gradient across the membrane. The right atrium and right ventricular chamber dimension were minimally dilated, with reduced right ventricular long axis function (tricuspid annular plane systolic evaluation [TAPSE] 16 mm) (Figs. 1 & 3). Right ventricular hypertrophy and moderate tricuspid regurgitation were present, and pulmonary arterial pressure was severely elevated (100 mmHg) (Fig. 3). Left ventricular ejection fraction was normal at 76%, while fractional shortening was 36%, and concentric left ventricular hypertrophy was noted.

DISCUSSION

Cor triatriatum is extremely rare in the adult population. Its pathophysiology is similar to that of mitral stenosis.⁽⁹⁾ Asymptomatic cases have been reported among adults, but reports among the very elderly are extremely rare.

Our patient was a 86-year-old woman with cor triatriatum, who presented for the first time during a routine preoperative assessment with primary pulmonary hypertension. The left atrium was divided into two chambers by a horizontal membrane, with reversal of normal flow across the membrane, suggesting relaxation abnormalities similar to those found in mitral stenosis or other causes of diastolic dysfunction. There was associated floppy interatrial septum but no atrial septal defect or other congenital heart disease. No other aetiology was found for the elevated pulmonary arterial systolic pressure in this patient. The mitral valve was morphologically normal, with unrestricted opening and no evidence of stenosis. There was neither a past history suggestive of significant smoking nor the presence of a barrel-shaped chest in the patient. The right ventricle was

dilated and hypertrophied with reduced pump function (TAPSE 16 mm).

Cor triatriatum presenting with primary pulmonary hypertension in the very elderly has scarcely been reported in the literature, with most being reported in children.^(10,11) Pulmonary hypertension may have resulted from backward transmission of the elevated left atrial pressure. In the initial phase, pulmonary artery vasoconstriction may contribute to the elevated pulmonary artery pressure.⁽¹²⁾ However, the pulmonary arteries were dilated in our patient, suggesting that the pressure changes had been ongoing for a long time and may have reversed with progressive dilatation of the pulmonary arteries, progressive right heart failure and associated changes in the pulmonary vascular bed. Severe pulmonary hypertension gradually leads to right heart failure. The associated changes in the pulmonary vascular bed (elevated precapillary resistance) tend to protect the left heart by making it less likely to develop pulmonary congestion at the expense of a reduced cardiac output.⁽¹³⁾

Thus, it is very likely that the haemodynamic changes in our patient, which led to progressive right heart failure, pulmonary hypertension and possibly, associated apparent reduced cardiac output, had occurred over a very long period of time due to the relatively large size of the opening. In rare cases, the onset of symptoms may be delayed if the opening is large enough.^(2,6)

The treatment of cor triatriatum depends on the level of obstruction between the left atria chambers. In symptomatic patients, resection of the membrane is the definitive treatment.⁽¹⁴⁾ Based on the associated symptoms and haemodynamic changes, our patient possibly requires definitive treatment. She is expected to have right and left heart catheterisation to study the haemodynamics and calculate the peripheral vascular resistance. This

will further determine the line of treatment. However, considering the fact that the patient's condition can deteriorate further after such definitive procedures, conservative management remains an option.

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