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Cor Triatriatum Sinistrum Presenting as Cyanotic Congenital Heart Disease: A Rare Case Report

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ABSTRACT

Cor triatriatum is an acyanotic congenital heart disease. We present a rare case of cor triatriatum sinistrum in a 6-month-old female infant who was presented with cyanosis and failure to thrive. The 2D transthoracic echocardiography and the Doppler color flow imaging showed a proximal venous chamber communicating to the distal left atrium through restrictive opening to the low-pressure, distal left atrial chamber.
The Saline Contrast Echocardiography confirmed a right-to-left atrial shunt due to a minor atrial septal defect. The defect was caused by a persistent pulmonary hypertension which had raised the right atrial pressure in the infant.
To the best of our knowledge, barely any such cases have been reported in the literature so far. Our report highlights the clinical utility of the Saline Contrast Echo in other cases of congenital heart diseases.

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Introduction

Cor triatriatum is a rare acyanotic congenital cardiac anomaly which comprises of 0.1 - 0.4% of all the patients with congenital heart disease (1). Cor triatriatum sinistrum is characterized with the division of the left atrial chamber into two parts by a fibromuscular membrane, the proximal posterosuperior venous chamber and the distal anteroinferior left atrium proper.

Classically, the proximal (upper or superior) chamber of the left atrium (LA) receives pulmonary venous connections whereas the distal (lower or inferior) chamber contains LA appendage as well as the true atrial septum containing fossa ovalis.

The distal chamber is in continuity with the atrioventricular (AV) valve while the two chambers communicate via a defect in the membrane. In this regard, various echocardiography modalities are known to be helpful in the diagnosis and detailed anatomic characterization of the different subtypes.

Case Report

A six-month-old female infant weighing 3.5 kg was presented with complaints of respiratory distress and failure to thrive. General physical examinations were indicative of a pulse rate of 120 beat/min, respiratory rate of 40 breath/min, blood pressure of 84/56 mm Hg and Spo2 of 75%.

During the cardiovascular auscultation, a diastolic murmur was audible in the mitral area. The chest X-ray was indicative of cardiomegaly with prominent bronchovascular markings in the bilateral lung fields. The electrocardiogram (ECG) revealed LA enlargement and right ventricular hypertrophy.

Furthermore, during the transthoracic echocardiography and the subcostal four-chamber view, a membrane was observed dividing the LA into two chambers with a linear, echogenic structure extending from the posterosuperior to the anterolateral wall of the LA (Figure 1).

On the other hand, Atrial Septal Aneurysm (ASA) was seen and thus, the coronary sinus

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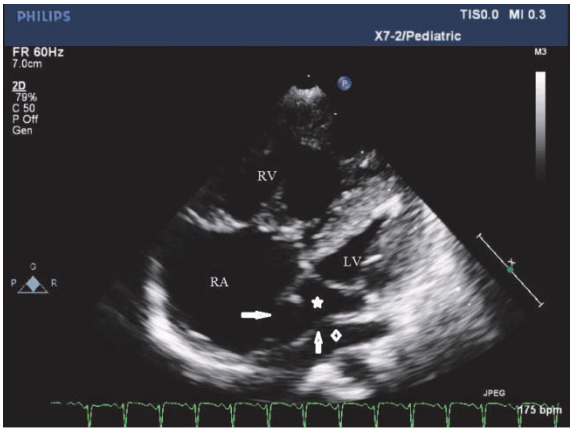

Figure 1. 2 D echo subcostal 4 chamber view showing RA, RV, LV; upward pointing arrow denotes the membrane dividing left atrium into two chambers visualised as a linear echogenic structure with a stenotic opening. Horizontal arrow denotes ASD with atrial septal aneurysm. (*) distal LA compartment- LA proper, (◊) proximal LA compartment- venous chamber, RA- right atrium, RV- right ventricle, LV- left ventricle

dilated. Afterwards, the right atrium and the right ventricle were enlarged. The LA and the cardiac valves were normal. Mild tricuspid regurgitation with severe pulmonary artery hypertension (RVSP = 94 mm Hg) was present as well.

On the Doppler color flow imaging, a turbulent, continuous flow from the proximal LA chamber to the distal left atrial chamber was detected (Figure 2). The main cause of the respiratory distress was still unknown to us. Reviewing the background of the cyanotic infant, we suspected that there might be a case of intracardiac right-to-left shunt.

To confirm the right-to-left shunt, Saline Contrast Echocardiography was performed through the left antecubital vein revealing the appearance of bubbles: first, in the right atrium followed by the LA. Then, there was an almost simultaneous appearance of the bubbles in both of the ventricular chambers suggesting a communication from the right atrium to the distal left atrial chamber while

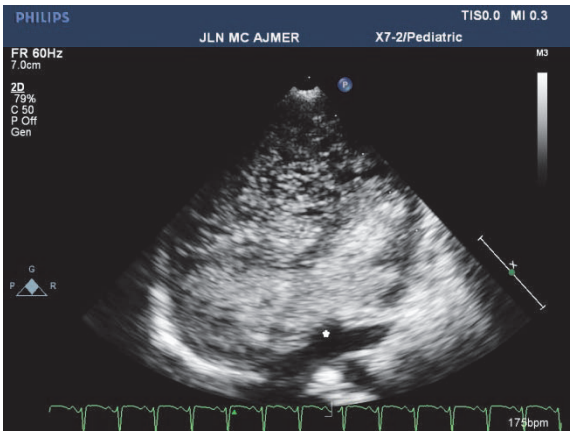

Figure 3. Saline contrast echo demonstrating opacified RA, RV, distal LA compartment and LV and non opacified proximal LA compartment with negative contrast effect into distal LA chamber (*)

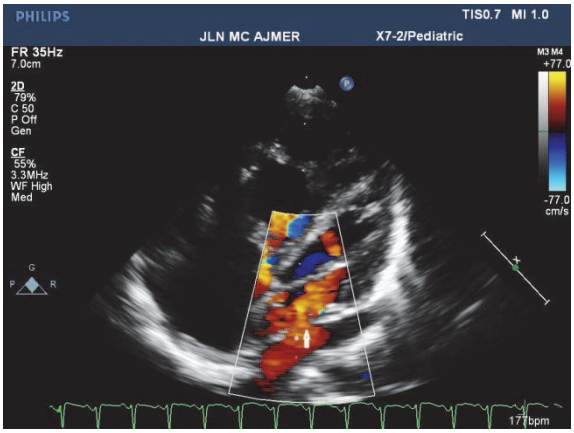

Figure 2. Colour flow imaging - subcostal 4 chamber view showing proximal to distal LA compartment colour flow via stenotic opening (arrow)

the proximal compartment of the LA was not opacified (Figure 3).

The failure of contrast opacification of coronary sinus (Figure 4) during this maneuver ruled out the possibility of persistent left superior vena cava. Furthermore, the Saline Contrast Echo appeared as negative in the distal left atrial compartment which was indicative of an inflow from the proximal left atrial chamber.

Discussion

Cor triatriatum sinistrum is a rare congenital anomaly in which a fibromuscular membrane divides the left atrium into two chambers. In the process, the accessory (proximal) chamber communicates with the distal chamber, the true left atrium which includes the left atrial appendage, through one or more openings (2). These openings could be classified into the three following types: diaphragmatic, hourglass or tubular.

In the diaphragmatic type, the membrane is oriented horizontally and it does not alter the anatomy of the chambers. This is the most prevalent of all types and it is the one detected in the present case.

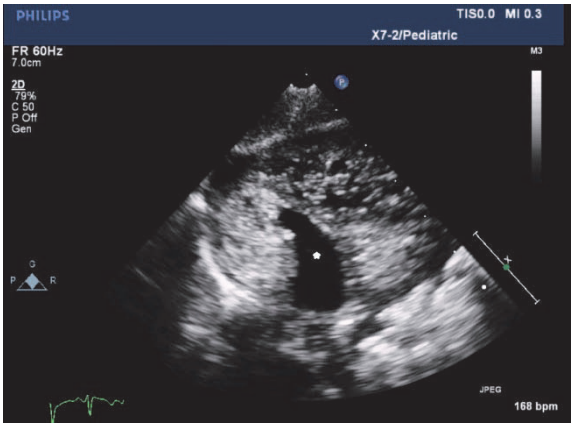

Figure 4. Saline contrast echo demonstrating dilated coronary sinus with absent contrast bubbles (*)

With reference to the hourglass type, the junction between the accessory chamber and the true left atrium is constricted externally while in the tubular type, the confluence of the pulmonary veins communicates with the left atrium through a channel.

Generally, around 70-80% of cor triatriatum cases are associated with other cardiac anomalies. The most frequent association (50-60% of all cases) is either patent foramen ovale or Ostium Secundum Atrial Septal Defect, which is usually between the right atrium and the distal chamber (3). Other associated defects are not proven to be as prevalent (4, 5).

In the classic cor triatriatum where there is no alternative pathway for the pulmonary venous blood, the stenotic opening in the membrane results in the supravalvular mitral stenosis with all the features of the elevated pulmonary venous pressure which are transmitted to the lungs causing pulmonary edema; the features that determine the condition of the patient. If the opening is 3 mm in diameter or less, the symptoms emerge in infancy and are similar to those of the total anomalous pulmonary venous returning with obstruction. If the opening is larger, the symptoms manifest later in infancy, childhood or occasionally later in life (6).

Most patients with this anomaly succumb within the first few months of life. Nonetheless, there is the exception of those patients with an adequate size of orifice in the membrane who might survive until their teens and even into adulthood. Death is likely to occur with pulmonary oedema, pneumonia or right-sided failure.

Predominantly, cor triatriatum sinistrum represents as an acyanotic congenital heart disease. Our patient had a rare cyanotic presentation of the disease. She had persistent pulmonary arterial hypertension of newborn leading to the right-to-left atrial shunt through an atrial septal defect, hence the systemic desaturation and cyanosis. On the other hand, the pulmonary venous hypertension caused by the stenotic left atrial inflow was regarded as a possible mechanism for the failure in the natural regression of the high pulmonary vascular resistance of the newborn.

The differential diagnosis of this disease include the total anomalous pulmonary venous connection to the coronary sinus or the right atrium, pulmonary vein stenosis and partial anomalous pulmonary venous connection which are all effectively discriminated on Routine 2D and Contrast Echocardiography obviating the need for more cumbersome and sophisticated diagnostic modalities.

Conclusion

To the best of our knowledge, cor triatriatum presenting as a cyanotic heart disease is quite a rarity. This case report underscores the diagnostic value of echocardiography. The diagnosis of cor triatriatum sinistrum is commonly made on Transthoracic Echocardiography and Saline Contrast 2D Echocardiography is also known to be useful in delineating various chambers, flow and shunts which may avoid the need for more sophisticated imaging modalities in unstable patients.

Conflict of interests

The author has no conflict of interests.

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