

Live 3-D Echocardiography in Diagnosis of Cor Triatriatum

S Kunapuli, A Rahman, M Dimaano, M Ahmad

Citation

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Abstract

Cor triatriatum is a rare congenital cardiac anomaly in which a common pulmonary venous chamber (proximal chamber) is separated from the left atrium (distal chamber) by a fibromuscular septum (membrane). Majority of the patients have one or more openings in the membrane. Cor triatriatum is usually an isolated congenital anomaly, but may be associated with cyanotic and acyanotic congenital heart diseases. The clinical manifestations depend on the size of the opening in the membrane. In most patients, the opening is severely restrictive and approximately 75 % of patients with Cor triatriatum die in infancy. In this case, we report for the first time, the use of Live 3 D echocardiography in the diagnosis of Cor triatriatum in a 27 year old Hispanic male.

CASE REPORT

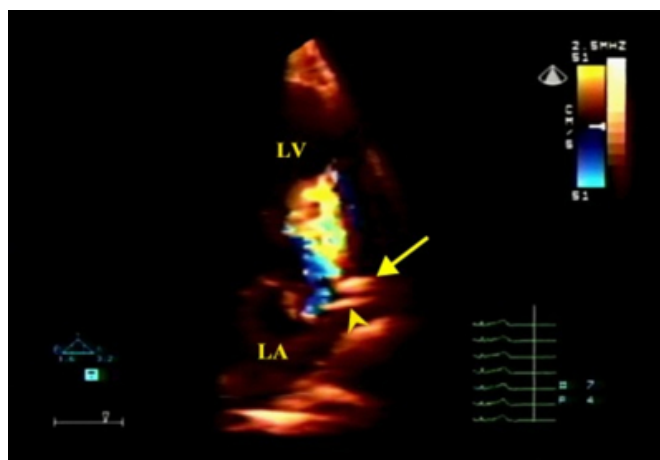
A 27 year old Latin American male was admitted for gross hematuria for 3 days. He had a past medical history of syncopal episodes, paroxysmal atrial fibrillation/flutter, and hypertension. His medications included Warfarin, Enalapril, hydrochlorothiazide, and Metoprolol.

Cardiovascular exam revealed normal first heart sound, and a physiologically split second heart sound. No murmurs, rubs or gallops were heard. Significant labs on admission: Pro Time 79.1 sec, Activated Partial Thromboplastin Time 63 sec, INR 8.7, Urinalysis positive for nitrites, leukocyte esterase, and red blood cells.

The patient underwent a trans-thoracic 2-D Echocardiogram (2-D) which demonstrated a normal left ventricle with an ejection fraction of 55-60%, normal right ventricle, normal left atrial and left ventricular size, and possible presence of a membranous band in the left atrium without obvious obstruction by Doppler flow measurements. To further confirm the 2-D findings, a live 3-D Echocardiogram (3D-Sonos 7500, Philips) was performed. A non-obstructing shelf like partition above the level of the pulmonary veins was visualized in multiple views (Figure 1, video clip). Based on the findings on 2-D and 3-D, the diagnosis of Cor triatriatum was confirmed. The patient was asymptomatic during his hospital stay and was discharged after correction of INR and treatment of urinary tract infection.

Figure 1

Figure 1: 3-D – cropped apical 4 chamber view with color flow showing left atrium (LA), left ventricle (LV), mitral valve (arrow) and the left atrial membrane (arrowhead).



Video Clip: Starts with a labeled still frame similar to figure 1, followed by a 3-D movie of the apical 4 chamber view demonstrating the partition in the left atrium. Cross sectional color flow views at the LA membrane and the mitral valve levels are shown in the last part of the movie clip.

On a follow up visit 1 month later, a Cardiac MRI was obtained for comparison with 2-D and 3-D images. MRI further confirmed the presence of a horizontal band across the left atrium causing very mild turbulence in flow.

DISCUSSION

Cor triatriatum is a rare congenital cardiac anomaly. The

incidence of Cor triatriatum is reported as 0.1 % to 0.4% (1,2). There are three main theories that explain the embryological basis of Cor triatriatum. In 1881, Fowler (3) suggested the malseptation hypothesis. He proposed that the septum subdividing the left atrium was an abnormal growth of the septum primum. The second theory of malincorporation hypothesis, suggests that Cor triatriatum is the result of incomplete incorporation of the embryonic common pulmonary vein into the left atrium (4). Van Praagh and Corsini proposed the entrapment hypothesis that suggests that the left horn of the sinus venosus entraps the common pulmonary vein and thereby prevents its incorporation into the left atrium (5). The most recent theory by Gharagozloo et.al. suggests that Cor triatriatum forms from the impingement of a prominent or persistent left superior vena cava on the left atrium, which in turn induces the formation of an abnormal left atrial membrane (6). In 1949, Loeffler classified Cor triatriatum into 3 groups based on the number and the size of the openings in the anomalous membrane into the left atrium: Group I = no opening; Group II = 1 or more small openings; and Group III = a wide opening (7).

Cor triatriatum is most commonly diagnosed in infancy or childhood, but there have been reports of Cor triatriatum in patients presenting in their eighth or ninth decade of life (8). Infants usually present with evidence of low cardiac output, including pallor, diminished peripheral pulses, and tachypnea. Children are typically small, suffering from poor weight gain. In adults, the most common presenting symptoms are dyspnea, hemoptysis, and orthopnea, or patients may be completely asymptomatic and the diagnosis of Cor triatriatum is incidental (9). On physical examination, cardiac murmurs are the most significant findings. The murmur is usually nonspecific and is variable in time, quality, and location (10).

A number of imaging techniques have been utilized in the diagnosis of Cor triatriatum, including 2D Echocardiography, Trans esophageal Echocardiography, CT scan and MRI. Recently introduced, Live 3-D Echocardiography (11) has advantages in being able to quickly obtain a comprehensive view of the left atrial anatomy from multiple vantage points. A wide angled full volume 3-D image can be obtained from apical or

parasternal acquisition. Left atrial chamber can be sliced from any plane to visualize the inside of the chamber and precisely localize the membrane, its attachment, size of the opening and relationships to the pulmonary veins. Compared to MRI, 3-D Echocardiography can be obtained quickly and easily at the bedside and in comparison to TEE, transthoracic 3D echo offers a completely noninvasive approach in diagnosis.

Based on this case report, we feel that 3-D technique has the potential to become the technique of choice in diagnosis of Cor triatriatum.

CORRESPONDENCE TO

Masood Ahmad Division of Cardiology University of Texas Medical Branch 301 University Blvd, Route 0766 Galveston, TX 77555 E mail: mahmad@utmb.edu

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Author Information

Sanjay Kunapuli, M.D.

Department of Internal Medicine, University of Texas Medical Branch at Galveston

Atiar Rahman, M.D.

Division of Cardiology, University of Texas Medical Branch at Galveston

Meneleo Dimaano, M.D.

Division of Cardiology, University of Texas Medical Branch at Galveston

Masood Ahmad, M.D.

Division of Cardiology, University of Texas Medical Branch at Galveston