Three-dimensional transthoracic echocardiographic evaluation of cor triatriatum

Eldad Einav, Gila Perk*, and Itzhak Kronzon

Non Invasive Cardiology Laboratory, The Leon H. Charney Division of Cardiology, NYU School of Medicine, NYU Medical Center, New York, NY 10016, USA

Received 30 November 2006; accepted after revision 4 February 2007; online publish-ahead-of-print 20 March 2007

We present an adult patient with cor triatriatum (CTT) due to a left atrial (LA) membrane. Two-dimensional and real-time three-dimensional transthoracic echocardiography (3DE) were performed as well as echocardiographic examination after exercise. These non-invasive modalities provided a comprehensive anatomic and hemodynamic evaluation of the anomaly.

**KEYWORDS**
Cor triatriatum; Echocardiography; Three-dimensional echocardiography

**Case presentation**

A 27-year-old man was referred to our echocardiography laboratory after complaining of shortness of breath and near fainting during a basketball game. The patient was an active man with no previous history of similar episodes, orthopnea or paroxysmal nocturnal dyspnea. Physical examination was unremarkable with normal heart sounds and no audible heart murmurs.

Echocardiography showed a normally appearing mitral valve and a membrane dividing the LA into an antero-inferior chamber which connects to the LA appendage and a postero-superior chamber to which the pulmonary veins drain.

Colour Doppler demonstrated a flow through a fenestration in the membrane (Figure 1).

Continuous wave Doppler tracing of the flow through the membrane showed a systolic wave that reached 1 m/s and diastolic wave that reached 2.3 m/s (Figure 2). The mean pressure gradient across the membrane was 8 mmHg. In order to further evaluate the hemodynamics of the lesion, the patient performed 10 sit-ups. The maximal diastolic velocity increased to 3 m/s with a mean gradient of 12 mmHg. Pulse Doppler with the sample volume at the tips of the mitral leaflets showed normal flow velocity across the mitral valve. Real-time 3DE demonstrated the entire intra-atrial membrane (Figure 3A) with two fenestrations. Three-dimensional colour Doppler confirmed the flow through these fenestrations (Figure 3B).

**Discussion**

CTT is a rare congenital anomaly, usually discovered at infancy. Typically in CTT, the LA appendage is part of the distal atrial chamber, differentiating it from a supravalvular mitral ring in which the LA appendage is connected to the proximal chamber. In contrast to mitral stenosis where there is only a diastolic flow across the valve, in CTT there is typically a trans-membrane gradient throughout the cardiac cycle resulting in flow both during systole and diastole. The natural history and prognosis of CTT correlates with the degree of flow obstruction and the magnitude of the gradient across the membrane fenestrations. In adulthood, patients typically present with dyspnea, hemoptysis and orthopnea. These patients can be often misdiagnosed as having mitral valve disease or primary pulmonary hypertension.

Symptomatic patients with significant gradient across the membrane benefit from a surgical repair. An emerging therapeutic procedure is a percutaneous balloon dilation of the CTT fenestrations. Transthoracic echocardiography (TTE) alone can correctly diagnose most cases of CTT. Transesophageal echocardiography (TEE) has the advantage of better visualizing the LA, LA appendage, the membrane and the pulmonary veins, hence provides higher sensitivity. More recently 3DE was suggested as an alternative to TEE in the diagnosis of CTT.

In our patient, we were able to diagnose and evaluate CTT by a completely non-invasive technique. Using 3DE allowed the demonstration of the membrane, the size as well as the location of the fenestrations, not as clearly...
demonstrated on the 2D images. These findings may help to guide the less invasive percutaneous approach for the treatment of CTT.

By continuous wave Doppler, we were also able to fully evaluate the hemodynamics of the anomaly both during rest and exercise, which correlated with the patient’s symptomatology.

This case suggests that 3DE and echocardiographic hemodynamic evaluation after exercise are helpful in the diagnosis and evaluation of CTT.
References