

It is likely that diagnostic accuracy has increased due to technological developments and widespread availability of transthoracic and transesophageal echocardiography. This may lead to increase in the reported cases of cor triatriatum in the literature.

When the cor triatriatum is the only abnormality the clinical findings are very similar to mitral stenosis, because both conditions cause pulmonary hypertension and subsequently elevated pulmonary capillary pressures, pulmonary arterial hypertension and right heart chamber enlargements. The possibility of a left atrial membrane should be actively considered when signs and symptoms of mitral stenosis are present. The presence of membrane and absence of anterior leaflet doming excludes the possibility of rheumatic mitral stenosis. However, rarely two conditions may present together like in our patient. In the presence of both conditions, the correct method for calculation of mitral valve mean and peak gradients and measurement of mitral valve area with pressure half time method is conflicting.

### Conclusion

Cor triatriatum sinister remains an uncommon form of congenital heart disease, although it is being diagnosed with increasing frequency in adults due to improvements in diagnostic imaging. In this case, cor triatriatum was incidentally revealed in a patient with mitral stenosis.

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## Adult bi-ventricular noncompaction cardiomyopathy

### *Yetişkin biventriküler noncompaction kardiyomiyopati*

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### Introduction

Noncompaction cardiomyopathy (NCM) is an idiopathic disorder characterized by an altered structure of the myocardial wall as a result of intrauterine arrest of compaction of the myocardial fibers in the absence of any coexisting congenital lesion (1).

The diagnosis is based on 2D echocardiography (2), or magnetic resonance imaging (MRI) or computed tomography (CT) scans (3).

In this case report, the typical echocardiographic images, angiographic, MRI and histological results of a case with noncompaction cardiomyopathy and both ventricles involvement (BVNCM).

### Case Report

A 29-year-old male was referred to our institution because of palpitations. The electrocardiogram showed an atrial flutter and left bundle branch block (LBBB).

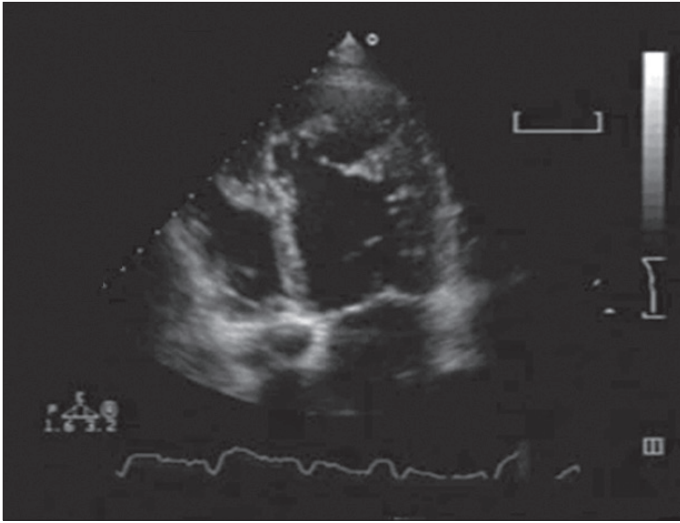
Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) showed features of BVNCM (Fig. 1, 2): dilated and globally hypokinetic left ventricle (LV) (ejection fraction-38%), prominent trabecularization of the apex and the mid-lateral wall, deep intertrabecular recesses with flow coming out from ventricular cavity; the end-systolic thickness ratio between noncompacted and compacted myocardium was 2.5; the apex of the right ventricle was hypokinetic and heavily trabeculated, with evident flow in intertrabecular recesses. During these examinations, the patient was in atrial flutter with a well-controlled ventricular rate response. The left atrium (LA) was mildly dilated, with spontaneous echo-contrast of 2°-3° degrees. A mild-moderate mitral regurgitation and tricuspid regurgitation were present and systolic pulmonary arterial pressure (sPAP) was 48mmHg.

The patient underwent endo-esophageal biphasic DC shock and sinus rhythm was restored.

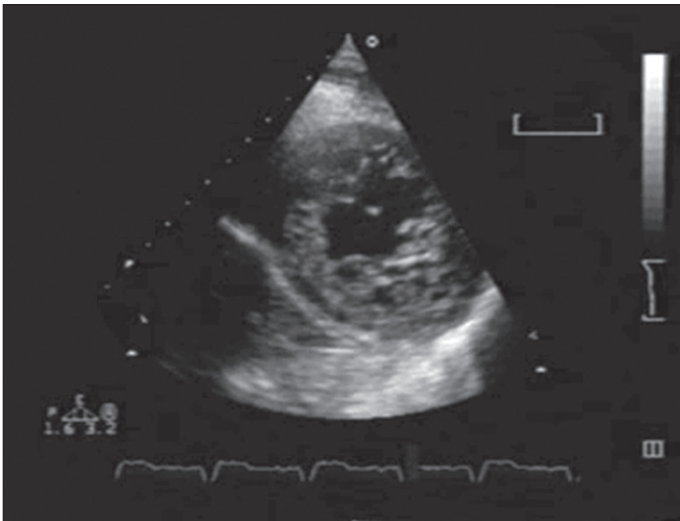
Angiographic study showed enlarged and globally hypokinetic LV with trabeculated apex and elevated right-sided pressure. Endomyocardial biopsy showed mild myocardial hypertrophy and thickened capillary

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**Figure 1.** Transthoracic echocardiography apical 4- chamber view of mildly dilated left ventricle (LV) with apical and mid-ventricular wall trabeculation. Right ventricle (RV) is hypokinetic and more heavily trabeculated than normal



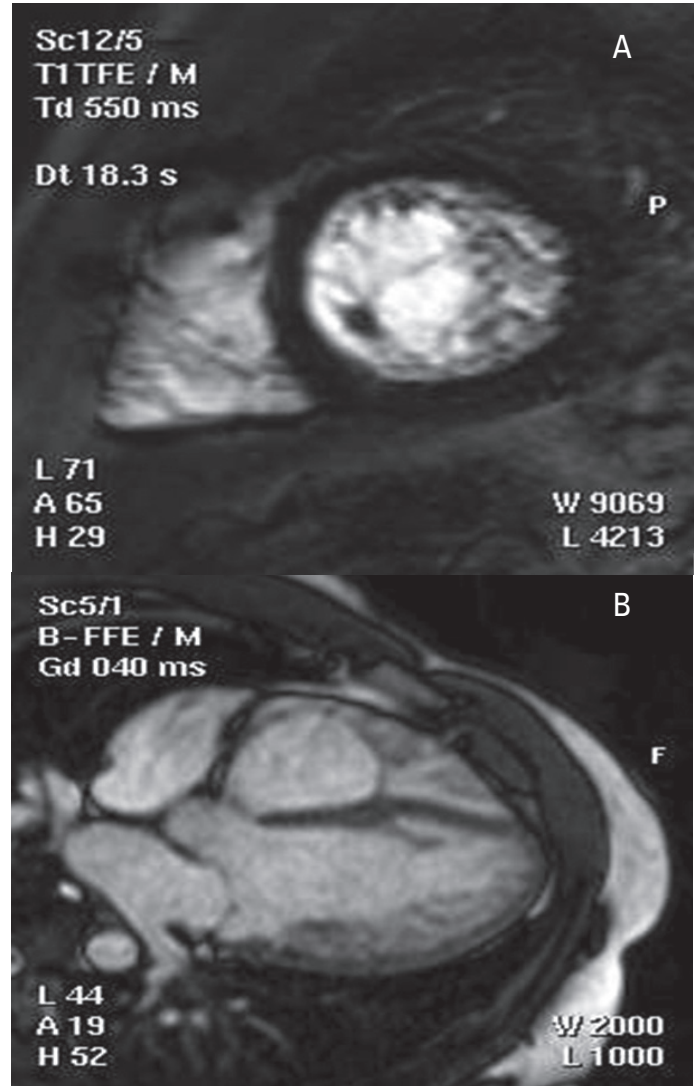
**Figure 2.** Transthoracic echocardiography short-axis apical level view of trabeculation of antero-lateral and infero-posterior walls

vessels, with nonspecific disarray. MRI study confirmed BVNCM (Fig. 3), showing: the mild dilatation and global hypokinesia of ventricles, the existence of abnormal inner zones of noncompaction, mainly located in LV (apical-mid-lateral wall and interventricular septum juxta-apex) and right ventricle (anterior wall and apex).

The patient was discharged on therapy with warfarin, amiodarone, carvedilol, and angiotensin-converting enzyme inhibitor. One year later, he was referred to our clinic because of atrial fibrillation. Transthoracic echocardiography confirmed the BVNCM, showing normal LV with ejection fraction of 55% and normal sPAP. The patient was discharged in sinus rhythm and currently he is in good condition. Patient's relatives underwent TTE and were found to be normal.

## Discussion

Non-compaction cardiomyopathy is a rare idiopathic cardiomyopathy reported by the WHO with an incidence of 0.05% (2). The NCM typically



**Figure 3.** Cine magnetic resonance imaging: a) Short axis: left ventricular apex with trabeculation b) Horizontal 4-chamber view: LV with prominent trabeculation of the mid- and apical-lateral wall and interventricular septum; RV with trabeculation of the anterior wall and the apex  
LV - left ventricle, RV - right ventricle

involves one or more segments of the LV (4), the right ventricular apex may be rarely involved (5).

The reported clinical features are: heart failure, conduction defects, syncopal episode, systemic embolic events and arrhythmias, although noncompaction alone does not seem to be a risk factor for supraventricular and malignant ventricular arrhythmias (6, 7). Long-term follow-up shows high incidence of heart transplantation and death (4). The diagnosis is based on the results of echocardiography (4), MRI, CT scan (3) or ventriculography. Magnetic resonance study, CT scan and TEE may play a role in uncertain cases (8), although no diagnostic criteria have yet been proposed, ventricular angiography might give an additional hint in some cases. Because of its wide availability, low cost and high diagnostic accuracy, TTE is the method of choice, at least when performed by experienced investigators. Few reports on use of TEE are available. More recently 4 clear-cut echocardiographic diagnostic criteria have been established: 1) Absence of coexisting cardiac abnormalities; 2) A

2-layered structure of the left ventricular wall with the end-systolic ratio of noncompacted to compacted layer  $>2$ ; 3) Finding this structure predominantly in the apical and mid-ventricular areas; 4) Blood flow directly from the ventricular cavity into the deep intertrabecular recesses as assessed by color Doppler echocardiography (4).

To our knowledge, our case is the 6<sup>th</sup> reported case of BVNCM in adults. In our case, echocardiography showed features of BVNCM. The diagnosis was further confirmed by angiography and by MRI. Clinical manifestations of BVNCM in our patient include cardiac arrhythmias, LBBB and reversible LV dysfunction. Atrial flutter is uncommon in patients with NCM (4, 7), while the incidence of LBBB is higher (up to 44%) (4). In adults with NCM the LV function is frequently impaired. However, some cases reported changes in LV morphology, dimension and function on carvedilol therapy (9). On presentation, our patient had atrial flutter, mildly dilated and globally hypokinetic LV, and persistent LV dysfunction in sinus rhythm.

Left ventricular noncompaction, by itself, does not seem to be a risk factor for stroke or embolic results, so there is no indication for oral anticoagulant therapy, except in patient at high risk for embolism (those presented with atrial flutter/fibrillation) (10). One year later the echocardiogram showed a normal LV function with ejection fraction of 55%; this finding could be an expression of LV morphology's changes over time upon carvedilol therapy.

### Conclusion

Echocardiography performed by skilled echocardiographers is the method of choice to diagnose BVNCM.

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