

A rare case of biventricular non-compaction cardiomyopathy associated with ventricular septal defect and atrial septal aneurysm

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A 35 year-old woman with a restrictive ventricular septal defect and an atrial septal aneurysm was admitted to our clinic. The patient reported mildly limited physical activity and palpitations. Transtho-

racic echocardiography showed an enlarged left ventricle and left atrium. The size of the right heart cavities was normal. The structure of both ventricle walls revealed a compact epicardial layer and an

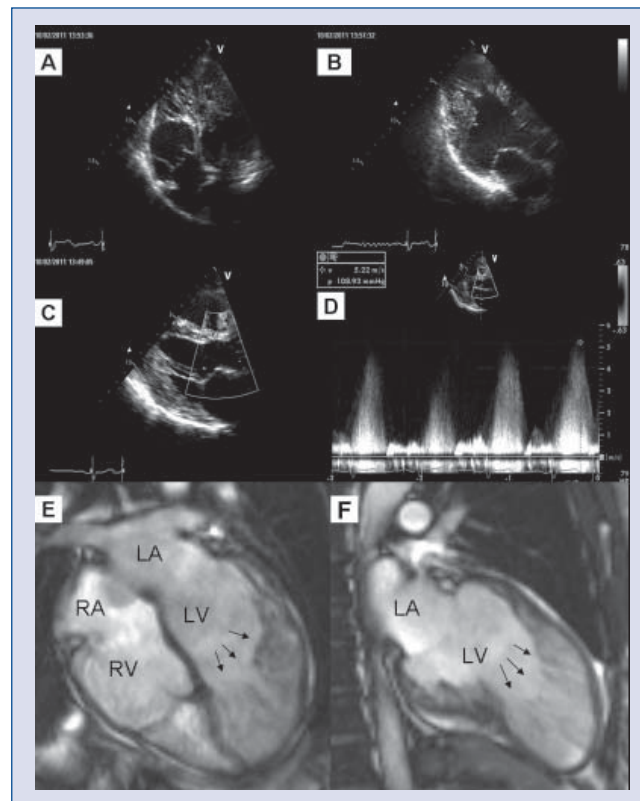


Figure 1. Echocardiography (A–D). Large trabecular meshwork and deep intertrabecular spaces of non-compacted myocardium in four- and three-chamber views (A, B). Small defect of interventricular septum (C) with left-to-right shunt and gradient of 110 mm Hg (D). Cardiac magnetic resonance imaging (E, F). Cine balanced steady-state free precession images in end-diastole in four- and two-chamber views. The area of non-compacted myocardium is shown by black arrows; LA — left atrium; LV — left ventricle; RA — right atrium; RV — right ventricle.

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endocardial layer consisting of a trabecular meshwork and deep intertrabecular spaces filled with Doppler color flow. The ejection fraction (EF) and peak systolic tissue velocity of basal septal segment were decreased (EF 30%, s' 8 cm/s, respectively). There was a small defect of the membranous interventricular septum with left-to-right shunt and maximum left ventricle to right ventricle gradient of 110 mm Hg. In the atrial septum, there was an aneurysm without signs of shunt. Magnetic resonance imaging was performed to confirm the diagnosis. The examination proved the non-compaction cardiomyopathy (NCCM) with the ratio of non-compact/compact layer > 2.3:1 [1]. The left-to-right shunt was estimated for 10 mL, which corresponded with insignificant pulmonary to systemic flow ratio (Qp:Qs 1.2:1). The 12-lead ECG Holter revealed more than 24,000 single ventricular extrasystoles. The patient was sent to the Electrophysiological Lab for ablation.

Ventricular non-compaction is a rare, unclassified cardiomyopathy. It results from lack of compaction of the loose myocardial meshwork during

morphogenesis [2]. Heart failure, thromboembolism and ventricular arrhythmias are the major clinical manifestations [3]. NCCM can be seen as an isolated malformation or associated with an obstructive lesion of the outflow tracts [4]. Hence, it is rare when NCCM is linked with ventricular septal defect and an atrial septal aneurysm.

Conflict of interest: none declared

References

1. Petersen SE, Selvanayagam JB, Wiesmann F et al. Left ventricular non-compaction: Insights from cardiovascular magnetic resonance imaging. *J Am Coll Cardiol*, 2005; 46: 101–105.
2. Tatu-Chitoiu A, Bradisteanu S. A rare case of biventricular non-compaction associated with ventricular septal defect and descending aortic stenosis in a young man. *Eur J Echocardiogr*, 2008; 9: 306–308.
3. Ozkutlu S, Ayabakan C, Celiker A, Elshershari H. Noncompaction of ventricular myocardium: A study of twelve patients. *J Am Soc Echocardiogr*, 2002; 15: 1523–1528.
4. Alehan D, Dogan OF. Right ventricular noncompaction in neonate with complex congenital heart disease. *Cardiol Young*, 2005; 15: 434–436.