

Left Ventricular Non-compaction with Associated Anomalies: A Rare Congenital Cardiomyopathy

AR Moaref, Y Mahmoody*

Department of Cardiology, Cardiovascular Research Center, Faghihi Hospital, Shiraz University of Medical Sciences, Shiraz, Iran

Abstract

A 22-year-old man was admitted with chief complaint of dyspnea on exertion. The patient had a history of operation 10 years ago for coarctation of the aorta. Transthoracic echocardiography showed bicuspid aortic valve, aortic regurgitation, left ventricular (LV) enlargement with ejection fraction of 45%, and a deeply trabeculated LV with deep intertrabecular recesses communicating with left ventricular cavity as demonstrated by color Doppler flow, the characteristic of the left ventricular non-compaction. Medical therapy and aortic valve replacement was performed. In the follow up, he was free of symptoms 12 months later.

Keywords: Left Ventricular; Non-compaction; Congenital Cardiomyopathy

Introduction

Non-compaction cardiomyopathy (NCC), also called spongiform cardiomyopathy, is a rare congenital cardiomyopathy.¹ Because it is particularly evident in the left ventricle, the condition is also called left ventricular non-compaction (LVNC). Diagnosis can be made by echocardiography. The current echocardiographic criteria for diagnosis are as follows: i) Presence of multiple echocardiographic trabeculations, particularly in the apex and free wall of the left ventricle; ii) Multiple deep intertrabecular recesses communicating with the ventricular cavity, as demonstrated by color Doppler imaging and iii) A 2-layered structure of the endomyocardium with an increased noncompacted to compacted ratio (suggested as >2.0 in adult and >1.4 in children).^{2,3}

Researchers have repeatedly suggested that LVNC is considerably under-diagnosed or mis-diagnosed as hypertrophic or dilated cardiomyopathy.⁴ Clinical presentations are similar to other cardiomyopathies in that they include depressed systolic and diastolic function, systemic embolization, and tachy-

arrhythmias. Medical treatment depends on the functional abnormalities (e.g. heart failure) and associated comorbidities, including systemic embolism and arrhythmia.⁴

Case Report

A 22-year-old man was referred to Cardiovascular Research Center of Shiraz University of Medical Sciences in Shiraz, southern Iran with chief complaint of severe dyspnea on exertion for 2 months. He has had surgery for coarctation of the aorta 10 years ago. Electrocardiogram showed biventricular hypertrophy with extreme QRS voltage. He underwent a transthoracic echocardiogram (TTE) that revealed bicuspid aortic valve, severe aortic regurgitation, left ventricular (LV) enlargement with ejection fraction of 45%, and a deeply trabeculated LV with deep intertrabecular recesses (Figure 1 and 2). Low-scale color flow Doppler confirmed communicating of intertrabecular recesses with the left ventricular cavity, characteristic of the left ventricular non-compaction (Figure 3). The patient had no family history of cardiomyopathy. Also, the patient did not have a history of arrhythmias and systemic embolization. Medical treatment started and aortic valve replacement was performed. During 12 months of the follow up, he was free of symptoms.

*Correspondence: Yadallah mahmoody, MD, Department of Cardiology, Cardiovascular Research Center, Faghihi Hospital, Shiraz University of Medical Sciences, Shiraz, Iran. Tel: +98-711-2343529, Fax: +98-711-2343529, e-mail: mahmoody_6@yahoo.com
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Fig. 1: Transthoracic echocardiography in four chamber view shows deeply trabeculated left ventricle with deep intertrabecular recesses

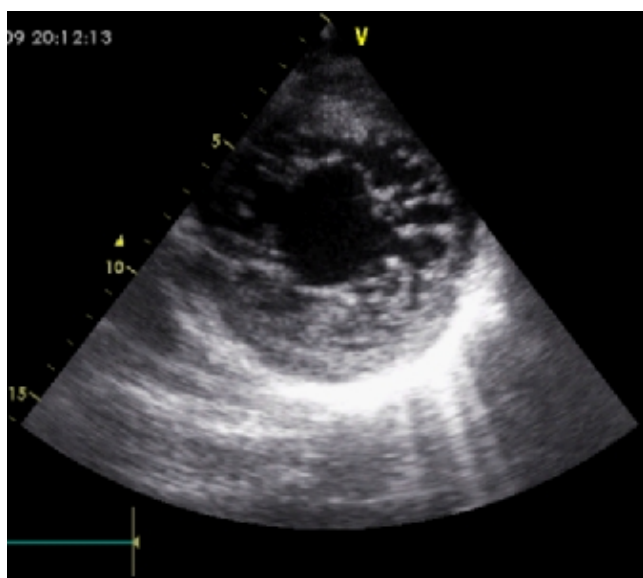


Fig. 2: Transthoracic echocardiography in parasternal short axis view shows deeply trabeculated left ventricle with deep intertrabecular recesses

Discussion

Non-compaction of the left ventricle (LVNC) is a rare disorder, classified as an unclassified cardiomyopathy by The European society of Cardiology working group on myocardial and pericardial diseases.⁵ Several case series and review of the literature have shown a slight excess of male, with a male proportion ranging from 56% to 82%.^{1,6}

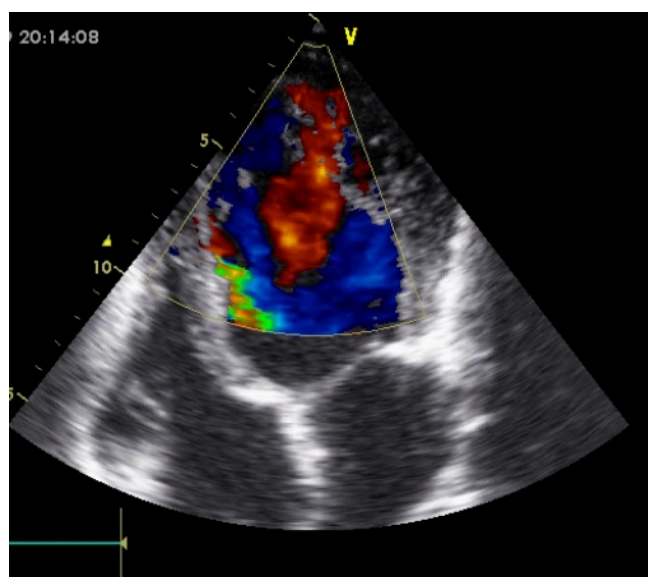


Fig. 3: Transthoracic echocardiography in four chamber view with Low-scale color flow Doppler shows communicating intertrabecular recesses with left ventricular cavity, characteristic of left ventricular noncompaction

Non-compacted myocardium is occasionally seen accompanying other congenital cardiac disorders such as Ebstein's anomaly, bicuspid aortic valve, ventricular septal defect, aorta-to-left ventricular tunnel, L-transposition of the great arteries, and isomerism of the left atrial appendage.⁷

The most common presentation reported in the literature has been tachypnea due to the low cardiac output.¹ Based on the limited data, the frequency and type of arrhythmias appear to vary by age. Among children, the more common arrhythmia includes WPW with or without supraventricular tachycardia, as well as ventricular tachycardia.⁵ Among adults, ventricular tachycardia and various forms of bundle branch block through complete atrioventricular block have been described.⁸ In this case, the patient had dyspnea on exertion and transthoracic echocardiography revealed characteristics of LVNC with associated anomalies (bicuspid aortic valve and coarctation of the aorta). The patient did not have a history of arrhythmia, systemic embolization, and family history of cardiomyopathy.

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Conflict of interest: None declared.

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