

Evaluation of Mechanical Dyssynchrony in Idiopathic Dilated Cardiomyopathy versus Non-compaction of Left Ventricle

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Abstract

Objective- Left ventricular non-compaction (LVNC) is a reportedly uncommon genetic disorder of endocardial morphogenesis and is being increasingly recognized. The purpose of this study was to evaluate the echocardiographic features, including mechanical dyssynchrony indices of patients with LVNC versus idiopathic dilated cardiomyopathy (IDC).

Methods- Between December 2004 and February 2006, we evaluated 116 patients with dilated cardiomyopathy candidates for cardiac resynchronization therapy (CRT) at our institution. The patients were divided into LVNC and IDC without LVNC groups, according to the diagnostic criteria for LVNC. Transthoracic echocardiography was done for all the patients, and pre-ejection periods as well as inter- and intra-ventricular delays were measured and the asynchrony index was calculated.

Results- Seventy-seven patients were male. LVNC was diagnosed in 23% of the patients. There was no significant difference in the patients' age and mean age of the patients (46 ± 16.5 years in LVNC vs. 51.13 ± 16.43 years in IDC). Mean left ventricular ejection fraction in the LVNC group was $16.65\% \pm 6.6\%$ and in the IDC group it was $18.91\% \pm 7.2\%$; mean age in the LVNC group was 46 ± 16.5 years and 51.13 ± 16.43 years in the IDC group, with no significant difference between the two groups.

Conclusion- LVNC is increasingly being reported and has become an important differential diagnosis in heart failure patients. Our study showed that there was no significant difference in the mechanical dyssynchrony indices between the two groups (*Iranian Heart Journal 2009; 10 (2):15-19*).

Key words: ventricular non-compaction ■ cardiomyopathy ■ ventricular dyssynchrony

Left ventricular non-compaction (LVNC) is supposed to be the result of an arrest or failure of the myocardial trabeculae during endomyocardial embryogenesis;¹ thus, myocardial non-compaction best characterizes the basic nature of this disorder and respects its embryogenesis. The definition of non-compaction includes thickened myocardium with a two-layered structure consisting of

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the compaction of the epicardial and non-compaction of the endocardial myocardium (maximum end-systolic ratio of non-compacted to compacted layer > 2), with a meshwork of perfused intertrabecular recesses and regional hypokinesia.²⁻⁶ It may occur without any other cardiac abnormality (isolated ventricular non-compaction) or may be associated with other congenital cardiac malformations.

Diagnosis can be made via echocardiography, and the current echocardiographic criteria for diagnosis typically include the following:

1) Presence of multiple echocardiographic trabeculations, particularly in the apex and free wall of the left ventricle. Stollberg and Finsterer suggested the anatomically confirmed definition of 3 trabeculations within 1 imaging plane, apically from the insertion of the papillary muscles, as a practically useful diagnostic criterion.

2) Multiple deep intertrabecular recesses communicating with the ventricular cavity, as demonstrated by color Doppler imaging.

3) A 2-layered structure of the endomyocardium with an increased non-compacted to compacted ratio (suggested as >2.0 in adults, >1.4 in children).⁶

The proportion of the ventricular wall affected (>50%) is also used in the definition by some authors.⁷ Various imaging techniques have been employed in the diagnosis of LVNC, including ventriculography,⁸ ultra-fast computed tomography,⁹ and magnetic resonance.¹⁰ Nevertheless, echocardiography is the imaging modality of choice for LVNC.^{10, 11} Magnetic resonance studies also have a role in diagnosis and can provide convincing evidence in cases where the echocardiographic findings are uncertain.¹²

Methods

Patients

Between December 2004 and February 2006, we evaluated 116 consecutive patients with dilated cardiomyopathy candidate for cardiac resynchronization therapy (CRT) for an assessment of ventricular dyssynchrony.

Up to three-fourths of the patients were in New York Heart Association function class (NYHA FC) II and the remainder was in NYHA FC III and IV. The inclusion criteria were all the patients with isolated LVNC. We excluded obese patients or patients with lung disease who had poor echo windows inadequate or difficult for interpretation.

2D and M-mode echocardiography

Echocardiography was performed in the standard manner using Vivid 7 digital ultrasound scanner equipment with an ergonomically-designed multi-frequency M3S transthoracic sector transducer and tissue velocity imaging facility. Standard views (apical, parasternal, and subcostal) were used for the evaluation of the chamber size and function. Left ventricular volume and ejection fraction were measured by Simpson's rule and eye-ball estimation, considering all standard views. LV volumes and ejection fractions were obtained in the apical 4- and 2-chamber views (biplane method).

The distribution of prominent trabeculations in the ventricle was assessed using parasternal, apical, and subcostal views. In order to improve the visualization of the trabeculations within the left ventricular endocardium, color Doppler was used in the parasternal short-axis and apical 4-chamber views. The echocardiography images were reviewed and interpreted by two authors to confirm the diagnosis.

Tissue Doppler imaging

To determine intraventricular dyssynchrony, spectral displays of 6 basal and 6 middle LV segments with pulsed wave tissue Doppler imaging (TDI) were obtained in the apical 4-, 3-, and 2-chamber views and stored digitally. In brief, pulsed wave TDI was obtained by placing the sample volume in the middle of each myocardial segment. Gain and filter settings were adjusted as needed to eliminate background noise and to allow for a clear spectral display. The measurements were performed with a sweep of 100 mm/s. Offline analysis of 3 end-expiratory beats were performed,

and the results were averaged. The time interval from the onset of the QRS complex to aortic valve opening (Q-AV), time interval from the onset of the QRS complex to pulmonic valve opening (Q-PV) and time interval from the onset of the QRS complex to the peak systolic velocity (TTP) were measured.

The parameters evaluated for intraventricular dyssynchrony were as follows:

1) Septum-to-posterior wall mechanical delay (SPWMD), 2) Septum-to-lateral wall mechanical delay, 3) Maximum difference in time-to-peak systolic velocity of two segments between twelve segments (TS-diff), and 4) Total asynchrony index (TS-SD) defined as the standard deviation of the TTP of the 12 LV segments.

Results

The most common presenting symptom was dyspnea, secondary to low cardiac output state. A high prevalence of ECG abnormalities was noted in both groups; the most prominent feature was left bundle branch block (LBBB). Characteristic biventricular non-compaction was found only in one patient and isolated LVNC was present in the other patients. None of the patients had isolated right ventricular non-compaction without the involvement of the LV. Use of color Doppler in the parasternal short-axis and apical 4-chamber views improved the visualization of the trabeculations within the LV endocardium. The median non-compacted to compacted myocardium ratio was 2.08 (range, 1.4 to 2.3). There was no significant difference in terms of age and LV ejection fraction (46 ± 16.5 versus 51.13 ± 16.43 years, and $16.65 \pm 6.6\%$ versus $18.91 \pm 7.2\%$, between the LVNC and IDC groups, respectively). One patient had LV apical thrombus. The echocardiographic features of the two groups are shown in Table I.

Discussion

During early fetal life, the myocardium consists of a loosely interwoven meshwork of myocardial fibers. The compaction of these fibers occurs with the reduction of the intertrabecular recesses in normal development. The failure of this process causes altered structure and non-compaction of the ventricle.¹³

Table I. Echocardiographic features of LVNC group versus IDC

	LVNC	IDC	PV
LV Pre ejection period (msec)	126.68 \pm 33.92	109 \pm 31.69	0.03
RV Pre ejection period (msec)	105.73 \pm 35.02	92.98 \pm 28.49	NS
Interventricular delay (msec)	34.87 \pm 21.36	31.43 \pm 21.43	NS
Septum-to-lateral delay (msec)	42.36 \pm 24.78	42.16 \pm 32.37	NS
SPWMD (msec)	87.36 \pm 53.11	83.37 \pm 45.26	NS
Basal asynchrony index (msec)	29.2 \pm 11.96	27.58 \pm 13.14	NS
Total asynchrony index (msec)	27.8 \pm 10.6	27.74 \pm 13.04	NS
dp/dt (mmHg/sec)	608.13 \pm 166.18	709.78 \pm 196.18	NS
LVOT VTI(cm)	9.1 \pm 3.6	12.66 \pm 3.9	NS

Echocardiography is the diagnostic modality of choice for LVNC. Recently, echocardiographic criteria for the diagnosis of LVNC have been proposed. Our patients fulfilled these criteria with the absence of congenital heart disease and

differences in the non-compaction-to-compaction ratio. However, when non-compaction is subtle or incomplete, disorders such as prominent normal myocardial trabeculations, hypertrophic cardiomyopathy, dilated cardiomyopathy, and LV apical thrombus should be included in the differential diagnosis. The clinical manifestations of LVNC come mainly from heart failure, arrhythmias, and embolism. The clinical findings and age at symptom onset are highly variable.¹⁴⁻²⁰ One long-term follow-up study showed that most patients developed ventricular dysfunction independent of their initial status.¹⁵ Atrial and ventricular tachyarrhythmias and conduction defects are frequently detected in these patients. The increased risk of embolism results from the ventricular dysfunction, atrial arrhythmias, and local thrombi within the deep intertrabecular recesses. LV thrombus was detected in one patient, but systemic embolization was not reported by any of the patients. Medical treatment depends on associated comorbidities, including systemic embolism and arrhythmia. The treatment of LVNC does not differ from the treatment of other cardiomyopathies, and ultimate treatment is heart transplantation. To date, there have been three reported cases treated with heart transplantation.^{14,16} Over the last decade, CRT has become one of the standard treatments of heart failure and on the other hand, LVNC has been recognized as a distinct form of cardiomyopathy with a distinct underlying cause and prognosis.²² Therefore, it was important for us to evaluate the mechanical dyssynchrony in patients with LVNC and compare these patients with those considered as IDC and identify the differences between these two groups. Our study showed that there was no significant difference between the mechanical dyssynchrony indices in patients with LVNC compared to those with IDC; as a result, the selection of patients for CRT implantation in both groups was based mainly on the clinical criteria and also the presence of ventricular dyssynchrony.

Conclusion

LVNC is increasingly being reported and has become an important differential diagnosis in heart failure patients. Our study showed that there was no significant difference in the mechanical dyssynchrony indices between the two groups except for the pre-ejection period of the aorta.

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