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Myocardial deformation pattern in left ventricular non-compaction: Comparison with dilated cardiomyopathy

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ABSTRACT

Introduction: Left ventricular (LV) systolic dysfunction is the most frequent initial presentation of patient with LV noncompaction (NC). Our objectives were to evaluate myocardial contraction properties in patients with LVNC and the relationship of non-compacted segments with the degree of global and regional systolic deformation.
Methods: We included 50 LVNC with an echocardiography and speckle imaging calculation of peak longitudinal strain (PLS). Each of the 16 LV myocardial segments was defined as NC (ratio NC/compacted layer > 2), borderline (NC/C 0–2) and compacted (NC/C = 0). Basal, median and apical strain values were calculated as the average of segmental strain values. For comparison a group of 50 patients with dilated cardiomyopathy (DCM) underwent the same measurements.
Results: There was no statistical difference between the 2 groups for any conventional LV systolic parameters. A characteristic deformation pattern was observed in LVNC with higher strain values in the LV apical segments (-12.8 ± 5.9 vs -10.7 ± 5.7) and an apical–basal ratio (1.52 ± 0.73 vs 1.12 ± 0.42; $p < 0.001$). There was no correlation between LV function and the degree of NC. Among 726 segments, compacta thickness was thinner in NC vs C segments (6.4 ± 1.4 vs 7.7 ± 1.8 mm; $p < 0.05$). There was no difference in WMS but regional strain values were significantly higher in NC compared to C segments (-13.1 ± 6.1 vs -10.2 ± 6.3; $p < 0.05$).
Conclusions: Compared to DCM, LVNC presented with relatively preserved apical deformation as compared to basal segments. Lower regional deformation values in compacted segments confirm the concept that LVNC is a phenotypic marker of an underlying diffuse cardiomyopathy involving both C and NC myocardium.

1. Introduction

According to the European Society of Cardiology Working Group on Myocardial and Pericardial Disease, left ventricular noncompaction (LVNC) is still an unclassified cardiomyopathy [1]. This cardiomyopathy is characterized by trabeculations and recesses within the ventricular myocardium. LV systolic dysfunction associated with heart failure is the main predictor of outcome and the most frequent initial presentation of patient with LVNC [2–4]. But its mechanism and relation to non-compaction is not clearly established.

In the context of dilated and hypokinetic left ventricles, it is unclear whether LVNC is a morphologic trait rather than a distinct cardiomyopathy. Pronounced trabeculations are present in both LVNC and dilated cardiomyopathy (DCM), which sometimes makes the differentiation difficult.

Although cardiac magnetic resonance imaging (CMR) has emerged as the gold standard modality in the diagnosis and evaluation of this disease [5], echocardiography is critical to perform and is still used as the first-line imaging. The echocardiographic criteria of Chin, Jenni and Stöllberger [6–8] are used to confirm the diagnosis. Furthermore, Paterick and Tajik proposed to introduce the concept of myocardium contraction of the NC segments as an additional diagnosis criterion [9], so that abnormal ventricular function and myocardial deformation patterns, on top of criteria of pathologic hypertrabeculation may lead to diagnose LVNC.

Deformation imaging with peak systolic longitudinal strain (PLS) calculation is able to evaluate the degree and the extent of LV dysfunction and differentiate mechanisms of segmental abnormalities. Dilated cardiomyopathy is associated with reduction of strains in all directions

Abbreviations: LV, left ventricle; LVNC, left ventricular noncompaction; DCM, dilated cardiomyopathy; NC, noncompacted; C, compacted; 2D, two dimensional; 2DSI, two dimensional speckle imaging; PLS, peak longitudinal strain; GLS, global longitudinal strain; CMR, cardiovascular magnetic resonance.

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with attenuation of LV twist [10]. This character is a marker of global systolic dysfunction and helps to understand the physiological nonuniformity of regional LV performance [11–12]. Furthermore, global longitudinal strain value is a prognostic marker of heart failure in cardiomyopathy [13–14].

The purpose of this study was to illustrate the role of deformation imaging to evaluate the relationship between the degree and the extent of NC with regional and global systolic dysfunction. The second objective was to define a new tool using functional approach to help in the classification of patients with similar dilated phenotypes and discriminate primitive from non-compacted dilated cardiomyopathy.

2. Material and methods

2.1. Populations

Fifty patients newly referred for LVNC were screened between January 2001 and December 2013. The diagnosis of isolated LVNC was based on the presence of the following criteria: (1) visual appearance of two distinct compacted epicardial layer and a non-compacted endocardial layer; (2) marked trabeculation and deep intertrabecular recesses within the non-compacted layer; (3) non-compacted to compacted end-diastolic myocardial ratio >2, and (4) absence of other associated congenital or acquired heart disease. [6–9]. Atrial fibrillation was an exclusion criterion, because it made it impossible to perform 2D speckle analysis.

In addition, 50 age- and LVEF-matched nonischemic dilated cardiomyopathy (DCM) patients were enrolled in our study (mean age 51.9 ± 15.2 years).

Oral informed consent was obtained from each participant.

2.2. Echocardiographic imaging and analysis

A complete 2-dimensional and Doppler echocardiography (Vivid 7 and Vivid 9, General Electric Medical Systems, Horten, Norway) with 2D speckle tracking analysis was performed in all patients.

2.2.1. 2D standard echocardiography

Echocardiographic images were obtained from the parasternal short axis views at the basal, median and apical levels and from the 3 standard LV apical views (4-, 2- and 3-chambers). All images were acquired at a frame rate of 50 to 70 frames/s for 2D views. For each patient we measured 2D LVEF (Simpson biplane method). Left ventricular mass was calculated by the Teichholz method. Diastolic function was assessed by the E/A and E/E′ ratio and the LA area according to guidelines [15].

LV wall motion was assessed according to a 17 segment model (ACC/AHA) [16]. Each segment was analyzed individually and scored on the basis of its motion and systolic thickening ranging from 1 to 4: 1 = normokinetic or hyperkinetic, 2 = hypokinetic, 3 = akinetic, and 4 = dyskinetic. The wall motion score index (WMSI) was calculated as the sum of all scores divided by the number of segments visualized. Apical segment (apical cap-segment 17) was excluded from the segmental analysis.

2.2.2. Extent and degree of non-compaction

The severity of NC in each of the 16 LV segments was assessed quantitatively by measuring the non-compacted and compacted myocardial layer thickness from the 3 parasternal short axis view in end-diastole (millimeter and percentage of extent). We calculated the ratio between NC and C layer to obtain the value of NC/C ratio. The segmental degree of non-compacted myocardium was categorized into 3 grades: NC if the ratio was above 2; borderline if the NC/C ratio was between 0 and 2 and compacted if there was no trabeculation (NC/C ratio = 0) (Fig. 1).

2.2.3. 2D longitudinal peak systolic strain

Strain measurements were performed offline with a dedicated automated software (Automated Function Imaging, EchoPAC PC, version 110.1.0, GE Healthcare). Only good quality images were used. From each apical view, 3 sample points were placed manually along the endocardium to define the LV base and the apex at the end-systolic frame. Each LV wall was divided into 3 segments (basal, median and apical) and bull's eye according to the 17 segment classification was displayed. The values of longitudinal systolic strain of all the segments were averaged to obtain a 2D global longitudinal strain (GLS) value. Peak longitudinal strain (PLS) was defined as the lowest strain value obtained for the longitudinal direction during systole (before the reference time point of the end of systole). Basal, median and apical strain values were respectively calculated as the average of the strain values of the 6 basal, 6 median, and 4 apical segments.

2.3. Statistical analysis

The statistical analysis was performed using SPSS for Windows (SPSS version 17, Chicago, Illinois). Quantitative values are expressed as the mean value ± SD. Intergroup comparisons were made by the independent samples Student's paired sample *t*-test or Mann–Whitney *U* test when appropriate. We assessed the association of LVEF with the degree of NC by the use of Pearson correlation analysis. A p value < 0.05 was considered to indicate statistical significance. Agreements were assessed for PLS measurement using the method proposed by Bland and Altman for the inter/intra-observer variability repeated by two independent observers in 30 patients.

3. Results

3.1. Baseline TTE characteristics

The mean age was 51.8 ± 15.1 years (39 male/76%). There was no statistical difference in 2D conventional systolic function parameters between LVNC and DCM patients. The mean LVEF was 33.7 ± 11.2% in LVNC and 34.0 ± 10.9% in DCM group. LV tends to be more dilated in LVNC group (LVEDV 190.9 ± 73.0 vs 173.7 ± 69.6 ml, $p = 0.067$). The degree of diastolic dysfunction was similar in the two groups. Only the left atrial area was significantly greater in LVNC patients (26.5 ± 8.2 vs 23.5 ± 8.7 cm^2, $p < 0.005$). Right ventricular function was preserved in both groups.

Conventional echocardiographic characteristics are shown in Table 1.

3.2. Global and regional deformation parameters (Table 2)

The comparison between LVNC and DCM patients showed no significant differences for the global PLS (−10.6 ± 4.9% vs −10.1 ± 4.9%).

The analysis of PLS values at basal and mid-level showed no significant differences between the 2 groups. We observed a significant correlation between LVEF and GPLS ($r = -0.886$; $p < 0.001$) but there was no correlation between the degree and the extent of the non-compaction with the level of systolic dysfunction (LVEF or GPLS).

The analysis of basal, median and apical levels of the LV showed a significant gradient from the base toward the apex among patients with LVNC. In patient with similar LVEF, LVNC group demonstrated a relatively preserved and significantly higher apical LV PLS values compared to DCM patients (−12.8 ± 5.9 vs −10.7 ± 5.7 $p = 0.025$). This difference was confirmed with a ratio of apex/basal strain values of 1.5 ± 0.7 in LVNC group compare to 1.1 ± 0.4 in DCM group ($p < 0.001$).

3.2.1. Segmental analysis (Table 3)

A total of 726 segments were analyzed on 3 parasternal short axis views (90.1% of all segments) in LVNC group. The remaining segments