Cardiac magnetic resonance markers of left ventricular non-compaction in patients with suspicious echocardiographic findings

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Abstract

Left ventricular non-compaction (LVNC) is considered as a distinct form of cardiomyopathy with a poorly understood background. We decided to analyze markers differentiating patients with echocardiographic suspicion of LVNC in whom the diagnosis was confirmed or excluded by cardiac magnetic resonance (CMR). The CMR diagnosis of LVNC (non-compacted/compacted layer ratio≥ 2.3) was confirmed in 15 of the 28 patients (54%). Patients with LVNC had lower right ventricular (RV) ejection fraction and more often RV trabeculation comprising the whole cavity at the level of RV apical segments. In the LVNC group there was a significant correlation between LV and RV end-systolic volume and LV and RV ejection fraction and a trend towards a correlation between LV and RV end-diastolic volume and mass. Our findings suggest that analysis of the right ventricle may be used in differential diagnosis of LVNC and other diseases of the left ventricle related to excessive trabeculation of this cavity.

Key words:

right ventricle, hypertrabeculation, non-compaction, diagnostic test

Left ventricular non-compaction (LVNC) is considered as a distinct and rare form of cardiomyopathy with a poorly understood background[1,2]. It is characterized by two-layered structure of the left ventricle (LV) comprising a thicker inner non-compacted layer and a thinner outer compacted layer. Patients with LVNC are at higher risk of thromboembolic events, ventricular arrhythmias, sudden cardiac death or progression towards end-stage heart failure. The disease can be isolated, but sometimes it coexists with other cardiovascular diseases such as coronary artery disease, hypertension or dilated cardiomyopathy, which makes the differentiation from other more complex. The diagnosis can be based on echocardiographic or cardiac magnetic resonance (CMR) criteria described elsewhere[1,2]. Often the initial suspicion is based on echocardiography, but patients are further referred for CMR, which allows better visibility of the endocardium, to confirm the diagnosis. We decided to analyze markers differentiating patients with echocardiographic suspicion of LVNC in whom the diagnosis was confirmed or excluded by CMR.

The study included 28 patients (median age 36 years, minimum 15, maximum 74 years, male sex 57%) referred for CMR with suspicion of LVNC in a single tertiary center. The studies were performed using a 1.5T scanner (Magnetom Avanto, Siemens, Germany). The diagnosis of LVNC was made if the non-compacted to compacted layers ratio on any of the end-diastolic long axis views (2-chamber, 4-chamber or 3-chamber) in at least one segment, excluding the apex, was ≥2.3 as defined previously[3]. A panel of clinical and CMR markers was analyzed and compared using common statistical tests.

The initial suspicion of LVNC was confirmed in 15 patients (54%). Baseline characteristics and CMR parameters of the two groups are presented in Table 1. There was no difference between the groups in terms of baseline characteristics, comorbidities, LV end-diastolic volume, ejection fraction or the presence of late gadolinium enhancement (LGE) of non-ischemic origin. Per definition, patients with confirmed LVNC had a higher non-compacted/compacted layer ratio, which was

Table 1. Baseline characteristics and CMR parameters in both studied groups

	LVNC (+) n=15	LVNC (-) n=13	p
Median age – years (min-max)	31 (17-71)	42 (15-74)	0.39
Male sex – n (%)	9 (60)	7 (54)	0.74
Hypertension – n (%)	2 (13)	2 (15)	1.00
Dyslipidemia – n (%)	2 (13)	2 (15)	1.00
Ventricular arrhythmia – n (%)	4 (27)	2 (15)	0.65
Coronary artery disease – n (%)	2 (13)	2 (15)	1.00
Congestive heart failure (NYHA II-III) – n (%)	5 (33)	5 (38)	1.00
Congenital heart disease – n (%)	3* (20)	0	0.23
NC/C ratio (min-max)	3.0 (2.2-4.3)	1.66 (1.0-2.0)	<0.001
NC maximal thickness – mm (min-max)	16 (7.5-26)	12 (9-16)	0.04
C minimal thickness – mm (min-max)	5 (3-7)	7 (5-9)	<0.001
Number of LV segments with NC – n (min-max)	8 (3-14)	5 (0-12)	0.02
LVEDVI – ml/m2 (min-max)	103 (81-313)	114 (83-222)	0.58
LVESVI – ml/m2 (min-max)	53 (23-260)	63 (25-185)	0.98
LVMI – g/m2 (min-max)	63 (42-133)	71 (52-101)	0.56
LVEF – % (min-max)	50 (15-75)	46 (17-72)	0.63
RVEDVI – ml/m2 (min-max)	95 (61-128)	86 (63-165)	0.45
RVESVI – ml/m2 (min-max)	44 (20-94)	31 (17-73)	0.14
RVMI – g/m2 (min-max)	23 (15-64)	22 (19-34)	0.61
RVEF – % (min-max)	53 (19-76)	60 (37-73)	0.02
RV hypertrabeculation – n (%)	11 (73)	1 (8)	<0.001
Non-ischemic LGE – n (%)	3 (20)	2 (15)	1.00

* restrictive ventricular septal defect (n=1), status post atrial septal defect closure (n=2)

LGE – late gadolinium enhancement, **LV** – left ventricular, **LVEDVI** – left ventricular end-diastolic volume index, **LVEF** – left ventricular ejection fraction, **LVESVI** – left ventricular end-systolic volume index, **LVMI** – left ventricular mass index, **NC** – non-compacted layer, **C** – compacted layer, **NYHA** – New York Heart Association, **RV** – right ventricular, **RVEDVI** – right ventricular end-diastolic volume index, **RVEF** – right ventricular ejection fraction, **RVESVI** – right ventricular end-systolic volume index, **RVMI** – right ventricular mass index

caused by both lower minimal thickness of the compacted layer and higher maximal thickness of the non-compacted layer in comparison to the LVNC (-) group. Patients with LVNC had more non-compacted LV segments and, what was most intriguing, a lower right ventricular (RV) ejection fraction and more often pronounced RV trabeculation (comprising the whole cavity at the level of RV apical segments). For those reasons we decided to analyze the LV/RV correlations of CMR parameters, which are presented in Table 2. We found a significant correlation between LV and RV end-systolic volume and LV and RV ejection fraction and a trend towards a correlation between LV and RV end-diastolic volume and mass, but only in the LVNC group.

The main finding of our study is RV involvement in LVNC, demonstrating that the disease is more likely a biventricular entity and not just an isolated LV disease. There are only a limited number of reports on RV involvement in LVNC in the literature [4-6]. Leung et al. reported that RV dysfunction was present in half of patients with LVNC. Significant RV dysfunction seemed to be a marker of advanced LVNC[4]. In

Table 2. Correlation between left and right ventricular parameters in both studied groups

Parameter	LVNC (+) n=15		LVNC (-) n=13	
	Pearson's R statistic	p	Pearson's R statistic	p
LVEDVI vs. RVEDVI	0.46	0.08	0.1	0.74
LVESVI vs. RVESVI	0.53	0.04	0.32	0.28
LVEF vs. RVEF	0.59	0.02	0.44	0.13
LVMI vs. RVMI	0.51	0.05	0.45	0.12

For abbreviations see Table 1.

a study by Nucifora et al. LV systolic function was the only variable independently related to RV systolic function, with a correlation coefficient similar to ours[5]. Finally, in another study, RV dysfunction in a morphologic LVNC population was strongly associated with increased trabeculations of the RV apex, as also observed in our study[6]. However, we demonstrated that not only RV systolic function but also its volume correlates well with LV parameters in LVNC patients. Other results of our study are in line with previous research confirming that the disease usually comprises more than five LV segments and that despite similar LV mass in both studied groups the thickness of the compacted layer is lower in patients with LVNC[3,7].

The main limitation of this analysis is its small character, but most other studies on this subject have included similar groups of patients[1-7]. Furthermore, for practical purposes we decided to use only one CMR definition of LVNC (Petersen's definition) and not the LV trabeculation mass[1,2]. Finally, we were not able to perform genetic testing.

In conclusion, our findings suggest that RV analysis may be used in differential diagnosis of LVNC and other diseases of the left ventricle related to excessive trabeculation of this cavity. Observation of hypertrabeculation of the RV apex and signs of RV enlargement and/or dysfunction may strengthen the suspicion of this pro-arrhythmogenic disease. Further studies in a larger sample of patients or systematic reviews of published reports are needed to confirm these observations.

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