Original Article

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A Novel Method for Quantification of Left Ventricular Noncompaction Using Two-Dimensional Echocardiography in Children

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Abstract

Objective: Although there are several echocardiographic criteria, there is not yet a general consensus about the diagnosis of left ventricular noncompaction. The current criteria are mostly based on the areas with maximal noncompaction in the heart. The echocardiographer may miss this maximal point leading to a misdiagnosis. Accordingly, we suggested a new method to measure the percentage of myocardial noncompaction using two-dimensional echocardiography.

Methods: In this study, the new method was examined on 4 noncompaction and 26 dilated cardiomyopathies, and 25 normal subjects. The percentage of noncompaction was measured at 3 levels (apical, papillary muscle and mitral valve) and averaged.

Findings: The mean percentages of myocardial noncompaction were 3.59±2.27, 8.86±5.52 and 34.7±26.1 in the control, dilated cardiomyopathy and noncompaction groups, respectively. A value of 17% or greater could distinguish left ventricular noncompaction from dilated cardiomyopathy with 92% specificity and 100% sensitivity and from normal subjects with 100% specificity and sensitivity. This percentage had a statistically significant association with noncompacted to compacted myocardial thickness ratio (P<0.001).

Conclusion: This method showed good correlations with the existing echocardiographic and magnetic resonance criteria. However, it is not dependent on finding the area of maximal involvement. Being comparable to magnetic resonance imaging in accuracy, it is easier to perform and more available.

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Key Words: Left Ventricular Noncompaction; Cardiomyopathy; Echocardiography; Children

Introduction

Despite the large number of studies about left ventricular noncompaction (LVNC), many uncertainties exist. The diagnosis is particularly important[1]. Echocardiography is accepted as the method of choice for LVNC diagnosis[2]. Magnetic resonance imaging (MRI) was used extensively for this purpose as well; however, artifacts from arrhythmias or breathing and higher cost are its disadvantages[2].

Large areas of hypertrabeculation (HT) can be seen in many diseases including dilated cardiomyopathy (DCMP) and even in normal subjects[3,4]. It is not known whether this hypertrabeculation is a milder form of non-compaction (NC) or a different pathology[1]. Some experts use the term hypertrabeculation/

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noncompaction (HT/NC) instead[5]. We preferred to use this term throughout this paper as well.

Three sets of echocardiographic criteria proposed by Chin, Jenni, and Stöllberger developed to differentiate LVNC from other diseases[3,5,6]. In spite of the existing criteria, there may be many cases of delayed or overlooked diagnosis[7,8]. This problem may result from the nature of these criteria which depends on the finding of an area with maximal NC[3,6], or the number of HT/NC areas in LV[5]. Failure to find the area with maximal NC or all HT/NC areas may result in a misdiagnosis. Measuring the percentage of hypertrabeculated or noncompacted myocardium (HT/NC%) may decrease misdiagnosis. We introduce a method for this measurement by echocardiography and examined it on a cohort of patients with LVNC cardiomyopathy, DCMP, and normal subjects. In order to create a 3-dimensional concept, we measured areas from 3 different levels in the LV and averaged them.

Subjects and Methods

From September 2009 to June 2011 targeted subjects for this study were enrolled. All children with a diagnosis of dilated or LVNC cardiomyopathy admitted at our hospital or referred to our outpatient clinics were included and a control group of healthy subjects were also selected in a convenient method. For LVNC cardiomyopathy patients, the study period extended to June 2012 due to the small number of patients. A noncompacted to compacted myocardial (NC/C) ratio of at least 1.4 was confirmatory for LVNC[9]. DCMP was diagnosed when a patient had left ventricular ejection fraction (LVEF) less than 50% (present or past) and disease duration of at least 3 months. The control group consisted of age-matched children referred to our outpatient clinics, with a final diagnosis of innocent murmur or non-cardiac chest pain and normal echocardiogram. Two DCM patients with poor echocardiographic windows were excluded from the study.

After obtaining informed consent from the parents, all study subjects were echocardiographically investigated by a single echocardiographer using a single echocardiographic scanner (MicroMaxx Ultrasound System, Sonosite Inc., USA). The main reason for using this scanner was its portability which permitted us to study subjects in both the intensive care unit and outpatient clinics. Echocardiographic parameters including left ventricular ejection fraction (LVEF), left ventricular fractional shortening (LVFS), and left ventricular myocardial performance index (LVMPI) were obtained. The NC/C ratio during systole was measured as described by Jenni et al[3].

The HT/NC% was defined as the mean percentages of HT/NC myocardium at three levels: apical, papillary muscles, and mitral valve. At each level, short axis view at systole was obtained and three areas were measured: whole myocardium and LV cavity (a), area encompassing only HT/NC myocardium and LV cavity (b), and only LV cavity (c) (Fig 1). The HT/NC% at each level was calculated by using the following formula:

$$\%HT\text{-}NC = 100 \times (b-c)/(a-c)$$

The apical level was defined when a small amount of LV cavity can be seen to precisely measure all desired areas (Fig. 1). The papillary muscle level was defined when these muscles were first viewed when sweeping echocardiography probe from the apex toward the cardiac base. The mitral valve level was defined just inferior to the valve level. Maximal care was undertaken not to include papillary muscles, aberrant chordae tendineae and LV bands in the HT/NC areas. The study was accepted by the ethical committee board of Tehran University of Medical Sciences and in accordance with good clinical practice and the Declaration of Helsinki.

The statistical analysis was performed using PASW Statistics 18 software. Descriptive statistics such as mean, standard deviation (expressed after ±), and frequency were calculated for each variable. Independent-sample T, Chi-square, and Pearson Correlation tests were used for the statistical analyses. A *P*-value less than 0.05 was considered statistically significant.

Findings

Twenty six patients with DCMP, 4 with LVNC cardiomyopathy and 25 normal subjects were

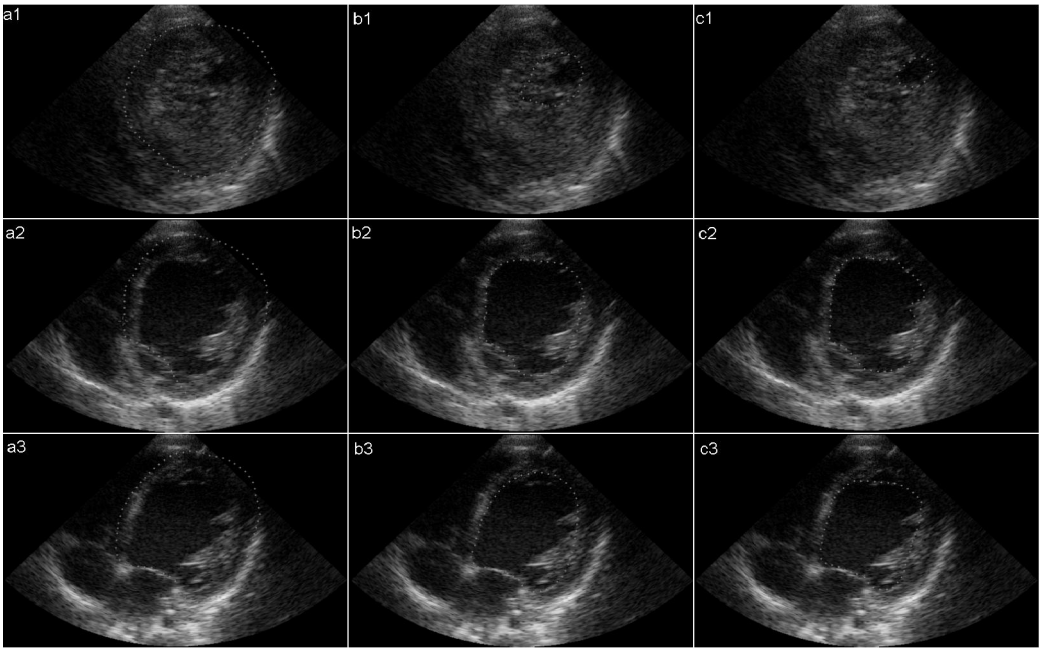

Fig. 1: The parameters required to calculate the percentage of hypertrabeculated/noncompacted myocardium. The areas a, b, and c are defined in the text. The numbers 1, 2, and 3 refer to the apical, papillary muscle and mitral valve levels, respectively

enrolled in the study. There was no statistically significant difference regarding the sex, age, weight, and body surface area between DCMP and control groups (Table 1).

Patients with DCMP had statistically lower EF, FS, MPI, and higher NC/C ratio in comparison to the normal subjects (Table 1). The %HT/NC% at all 3 levels and its average were significantly higher in the DCMP group than in the normal subjects (Table 2). The HT/NC% was significantly correlated with the NC/C ratio (correlation coefficient=0.543, P<0.001).

Discussion

LVNC was accepted as a distinct type of cardiomyopathy by the World Health Organization[10]. However, there are many controversies regarding its etiology, pathogenesis, diagnosis, and management[1]. The diagnosis is especially important because the prognosis and management of LVNC differ from those of its differential diagnoses[3].

There are 3 echocardiographic criteria for the diagnosis of LVNC. The Chin criteria was first

Table 1: Demographic and basic echocardiographic data in three groups*

Parameter	Control n=25	DCMP n=26	LVNC n=4	*P* value**
Age (years)	5.4	6	6.1	0.6
Sex (female/ male)	10/ 15	13 / 13	2 / 2	0.5
Weight (kg)	18.8	19	17.4	0.9
Body surface area (m^2)	0.73	0.72	0.69	0.9
Left ventricular ejection fraction (%)	69.7	35.5	46.3	<0.001
Left ventricular fractional shortening (%)	38.6	16.7	22.68	<0.001
left ventricular myocardial performance index	0.35	0.77	0.73	<0.001
NC/C ratio	0.44	0.79	1.83	<0.001

NC/C, noncompacted to compacted myocardial thickness; DCMP, dilated cardiomyopathy, LVNC, left ventricular noncompaction;
* comparing the averages in DCMP and control groups

Table 2: Left ventricular hypertrabeculation/noncompaction percentage at three different levels in the left ventricle

Level	Control Mean (SD)	DCMP Mean (SD)	LVNC Mean (SD)	*P.* value*
Apical	1.75 (2.25)	9.76 (7.35)	31.3 (21.6)	<0.001
Papillary Muscle	4.22 (3.78)	7.91 (6.35)	32.5 (20.1)	0.02
Mitral Valve	4.80 (6.09)	8.89 (8.23)	40.7 (38.0)	0.049
Average	3.59 (2.27)	8.86 (5.52)	34.7 (26.1)	<0.001

SD: Standard Deviation; DCMP: dilated cardiomyopathy; LVNC: left ventricular noncompaction;
* comparing the averages in DCMP and control groups

described and based on X-to-Y ratios at 3 levels (mitral valve, papillary muscle, and apex)[6]. X corresponds to the distance between the epicardial surface and trough of a trabecular recess, while Y to the distance between the epicardial surface and peak of the trabeculation. The measurements should be performed at end diastole and a ratio up to 0.5 is required for the diagnosis of LVNC[1].

The Jenni criterion postulates a systolic ratio of NC myocardial thickness to the adjacent compacted myocardium of more than 2 at the thickest part of myocardium, confirmation of blood flow in the recesses, absence of any cardiac abnormality, and the presence of characteristic trabeculations and deep recesses between the trabeculations[3]. Pignatelli et al suggested that a ratio of at least 1.4 is sufficient for the diagnosis of LVNC in children[9]. Jenni criterion is the most accepted among these three sets and used for the diagnosis of LVNC in this study.

The Stöllberger criteria require the presence of at least 4 trabeculations distal to the papillary muscles at one plane and intertrabecular spaces filled from the ventricular cavity[5]. Finsterer and Stöllberger introduced definite, probable and possible LVNC diagnoses[11]. Definite diagnosis is present when the patients fulfill both Jenni and Stöllberger sets of criteria, while probable corresponds to the fulfillment of just one, and possible to the situation where the number of trabeculations is less than 4 or the NC/C ratio is lower than two[11].

Kohli et al studied a cohort with heart failure and investigated all these 3 sets of diagnostic criteria[4]. At least one set of criteria was fulfilled by 47 patients. Of those, 78.7% met the Chin criterion, 63.8% the Jenni criterion, and 53.2% the Stöllberger criteria. Only one set of criteria was fulfilled by 36.2% of the patients, while only 29.8% met all sets[4].

Bodiwala et al used live 3-dimensional echocardiography for measuring maximal NC/C ratio[12]. They reported 8 patients diagnosed by this method. Only 3 out of these 8 patients were diagnosed accurately by 2-dimensional echocardiography using Jenni criterion.

The idea of LVNC quantification by area measurement was first introduced by Belanger et al[13]. However, they used only absolute area values for this purpose. We introduced a new method for the quantification of HT/NC of LV and examined it on patients with DCMP, LVNC cardiomyopathy and normal subjects. Although more time-consuming, this method has the advantage of being straightforward. The echocardiographer should investigate at least 3 levels in the myocardium and the result is based on an average rather than the areas of maximal involvement. We showed that this HT/NC% is well correlated with a carefully measured maximal NC/C ratio.

MRI was used to diagnose LVNC as well. Petersen et al measured the NC/C ratio during diastole and showed that a value greater than 2.3 could be 86% sensitive and 99% specific for the diagnosis of LVNC[14]. Alhabshan et al measured the end-systolic NC/C ratio by both MRI and echocardiography[15]. They found that MRI can found a higher NC/C ratio than echocardiography in some subjects.

Korcyk et al first described MRI quantification of HT/NC%[16]. They found a diastolic value of 22% in their unique LVNC patient and a mean of 11.3% (range 1.5-19) in 10 DCM patients. They proposed a value of 20% as a threshold for distinguishing LVNC from DCM. Fernndez-Golfin et al studied the same method on a larger cohort of

ischemic heart disease, DCM, valvular heart disease, and left ventricular hypertrophy patients, and normal subjects[17]. They found relatively higher values in their patients (17.2±4.9 in normal subjects, 23.3±6 in DCM patients). Jaquier et al used MRI quantification of HT/NC% as well[18]. They studied patients with LVNC, DCM, hypertrophic cardiomyopathy (HCM), and normal subjects[18] They found that mean percentages were 32±10 in LVNC, 11±4 in DCM, 12±4 in HCM, and 12±5 in controls. They reported that a percentage of more than 20% can be 93.7% specific and similarly sensitive to predict the presence of LVNC[18].

In our cohort, only two DCMP subjects had a percentage higher than 17% while our 4 LVNC patients had values of 73, 24, 17, and 24%. This means that the threshold value of 17% can distinguish LVNC from DCMP with 92% specificity and 100% sensitivity and from normal subjects with 100% specificity and sensitivity. Color Doppler echocardiography confirmation of direct blood flow from the ventricular cavity into deep intertrabecular recesses seems to be necessary for the diagnosis of LVNC, as it is a unique feature of this disease[3]. Our suggested echocardiographic threshold value of 17% is very close to the MRI threshold of 20% which was suggested by Jaquier et al, and Korcyk et al[16,18]. In summary, our suggested method is the echocardiographic equivalent of MRI HT/NC% measurement while the Jenni criterion is the equivalent of MRI NC/C ratio measurement.

Study Limitations:

The greatest limitation was in our cohort of patients. First, we found only 4 patients with LVNC during the study period. Second, the study was performed in a pediatric setting; therefore, this method was not examined on adult patients.

In addition, it was impossible to blind the echocardiographer to the diagnosis of the studied subjects. Although the echocardiographer tried his best, there may be some bias in the echocardiographic measurements.

Conclusion

We introduced a new echocardiographic method to measure HT/NC% and tested it on patients with DCM or LVNC, and normal subjects. We showed that the percentage had a statistically significant correlation with the noncompacted to compacted LV ratio.

Acknowledgment

Conflict of Interest: None

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