

Cardiac Magnetic Resonance Imaging for Evaluation of Non-Compaction Cardiomyopathy in Patients with or without Left Ventricular Systolic Dysfunction

ALEJANDRO DEVIGGIANO^{MTSAC, 1}, PATRICIA CARRASCOSA^{MTSAR, 1, 1}, CARLOS CAPUÑAYMSAR, ^{†, 1}, HÉCTOR DESCHLE ^{MTSAC, 1}, JULIO M. LEWKOWICZ^{MTSAC, 1}, CARLOS D. TAJER^{MTSAC, 1}, ALEJANDRO STEWART-HARRIS^{2, 3}, JAVIER VALLEJOSMSAR, ¹

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Address for reprints:
Alejandro Deviggiano.
Av. Maipú 1668, (B1602ABQ)
Vicente López, Buenos Aires,
Argentina.
Tel-Fax: 54-11-48377596.
e-mail: adeviggiano@gmail.com

SUMMARY

Objective

Non-compaction cardiomyopathy (NCC) is a genetic disorder characterized by the presence of an extensive layer of trabecular myocardium with intertrabecular recesses communicated with the ventricular cavity. The aim of this work is to evaluate the clinical and morphological characteristics of patients with NCC with and without systolic dysfunction assessed by cardiac magnetic resonance imaging (Cardiac MRI).

Material and Methods

Twenty patients with NCC diagnosis were retrospectively included. End-diastolic volume (LVEDV) and end-systolic volume, end-diastolic diameter (LVEDD) and end-systolic diameter, ejection fraction (LVEF), cardiac mass and LV trabeculations were determined. The distribution of NC myocardium was carried out with the model of 17 myocardial segments.

Results

The average thickness of NC myocardium and normal myocardium was 13.1 ± 3.3 mm and 3.6 ± 0.6 mm respectively. LVEDD, LVEDV, global, compacted and trabecular left ventricular mass were significantly increased in patients with ventricular dysfunction. LVEF had a negative linear correlation with the trabecular myocardial mass (MM)/m² (R = 0.67, p = 0.001), LVEDV/m² (R = .77, p <0.001) and LVEDD/m² (R = 0.7, p <0.001). There was also a negative linear correlation between MM/m² and LVEDV/m² (R = 0.76, p <0.001).

Conclusions

There are two ways of presenting the disorder, a subtle or mild with preserved systolic function and another associated to a ventricular dysfunction that behaves as dilated cardiomyopathy.

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Key words > Non-compaction cardiomyopathy; cardiac magnetic resonance imaging; trabeculations; cardiomyopathies; echocardiogram.

Abbreviations >	
NCC:	Non-compaction cardiomyopathy
Cardiac	
MRI:	Cardiac magnetic resonance imaging
LV:	Left ventricle
NC:	Non-compacted
C:	Compacted
LVEF:	Left ventricular ejection fraction
Global LV	
mass:	Global left ventricular mass
Compacted	
LV mass:	Compacted left ventricular mass
LVTM:	Left ventricular trabecular mass

BACKGROUND

Non-compaction cardiomyopathy (NCC) is a genetic disorder characterized by the presence of an extensive layer of trabecular myocardium with multiples and deep intertrabecular recesses communicated

with the ventricular cavity (1, 2). Its main clinical manifestations are heart failure, thromboembolic events, arrhythmias, and sudden death (3). Within the same family, the penetrance and phenotype of NCC are very variable (4-9).

^(*) Diagnóstico Maipú. Vicente López, Buenos Aires, Argentina.

^(†) Hospital General de Agudos Dr. Cosme Argerich. Ciudad Autónoma de Buenos Aires, Argentina.

¹ To apply for Miembro Titular de la Sociedad Argentina de Cardiología.

^{MTSAC} Miembro Titular de la Sociedad Argentina de Cardiología

^{MTSAC} Full Member of the Argentine Society of Cardiology

¹To apply as full member of the Argentine Society of Cardiology

Although the prevalence of NCC is higher in symptomatic patients with ventricular dysfunction (10), the new diagnostic modalities increased its detection in asymptomatic patients with preserved systolic function. Despite of that, the pathology is still being subdiagnosed in the clinical practice due to the low index of suspicion added to the use of echocardiographic classifications with uneven diagnostic criteria (8, 11, 12). There are two criteria for cardiac MRI that recognize correctly this entity (13, 14). To date there are no studies in literature that compare carriers of NCC with and without ventricular dysfunction through cardiac MRI.

The discrepancy between the diagnostic criteria for cardiac MRI and echocardiography has established the debate within the cardiological community with the aim of achieving the right diagnosis of the pathology.

The purpose of this work is to evaluate the clinical and morphological characteristics of patients with NCC with and without systolic dysfunction assessed by cardiac MRI, and to discuss the scopes and limitations of echocardiography and cardiac MRI in the diagnosis of this pathology.

MATERIAL AND METHODS

Twenty patients with NCC diagnosis were retrospectively included for a period of time between January, 2008 and March, 2010. The diagnosis was based on the absence of associated congenital heart defect or other cardiomyopathies and the finding, through MRI, of two well-differentiated layers of myocardium, one of NC or trabecular myocardium of higher thickness with intertrabecular recesses and other of thinner C myocardium with a mean between both (NC/C myocardium) > 2.3 in a more trabecular part (13) (Figure 1). From the total number of studied patients, 14 received contrast.

Acquisition of MRI images

An equipment 1.5 Tesla (Achieva, Philips Medical Systems, Best, The Netherlands) was used. A 16-channel specific cardiac antenna was also used and the images were acquired with cardiac synchronization through vectorcardiogram. The anatomic-functional study was done with balanced cine MRI images in stationary state, parallel image acceleration acquired at the end of exhalation using a TR 3.5 ms, a TE 1.8 ms and an angle of 60° on the axis, 2 cameras, 4 cameras, a short axis covering all the extension of the left ventricle and the outflow tract. Turbo spin-echo sequences with black blood in proton density with a TR 1935 ms and a TE 40 ms were also done.

Delay-enhancement sequences together with gradient-echo sequence boosted in T1 in 14 patients, 12/15 minutes after intravenous drip administration of a dose of gadolinium 0.04 mmol/kg.

Analysis of MRI images

The analysis was done in a workstation (ViewForum; Philips Medical Systems) with specific software. End-diastolic volume (LVEDV) and end-systolic volume, end-diastolic diameter (LVEDD) and end-systolic diameter, ejection fraction (LVEF), cardiac mass and LV trabeculations were determined.

End-diastolic phase (ED) and end-systolic phase (ES) were established in short axis cine MRI images which completely cover the LV, subsequently the epicardium and the endocardium with the inclusion of the papillary muscles

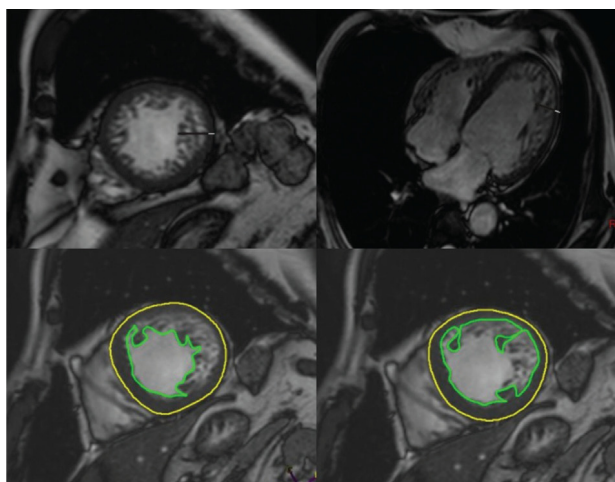


Fig. 1. Short axis cine MRI images acquired at the end of diastole.

A and B. Short axis cine MRI images and four chambers that show the quantization of the thickness of the non-compacted (black line) and compacted (white line) myocardial layer. **C.** Measurement of the left ventricular trabecular mass. The internal line that delimits the endocardium includes the papillary muscles and the trabecular myocardium. **D.** Measurement of the compacted left ventricular mass. The internal line that delimits the endocardium includes the papillary muscles and excludes the trabecular myocardium.

were semiautomatically demarcated. Trabeculation was defined as the myocardium that protruded towards the left ventricular cavity in the end-diastole. The diastolic thickness of NC and C myocardium was measured over the major trabecular section, in the short axis cine MRI images. The distribution of NC myocardium was carried out with the model of 17 myocardial segments from the AHA classification (15).

The determination of the global left ventricular mass (global LV mass) was done with the inclusion of the papillary muscles and the trabecular myocardium whereas for the determination of the compacted left ventricular mass (compacted LV mass) that inclusion was excluded. The left ventricular trabecular mass (LVTM) was calculated by subtracting the compacted LV mass to the global LV mass (Figure 1). The LVTM percentage in relation with the global LV mass (LVTM %) was also calculated (14).

The determination of the ventricular volumes and the LVEF was done with the inclusion of the papillary muscles and the exclusion of the trabecular myocardium (16). The value LVEF $< 50\%$ was considered as ventricular dysfunction. All the determinations were adjusted to the corporal surface.

The presence of hyperintense myocardial areas after the administration of gadolinium was considered a positive delay-enhancement.

Analysis of echocardiographic images

After the cardiac MRI, 5 echocardiographies were repeated. The studies were done with equipment Toshiba Xario, using transducers of 3.0 MHz.

Two classifications were used. Jenny et al. (11) that takes into account the absence of other congenital abnormalities, a mean (NC/C) of end-diastole > 2 , and the objectification of the intertrabecular flow by Doppler color; and Stolberg et al. (12) that takes into account the visualization of perfused intertrabecular spaces from the ventricular cavity in the

Doppler color added to the presence of more than three trabeculations which protrude from the left ventricular wall at the apical level, in front of the papillary muscles, visible in an echocardiographic plane.

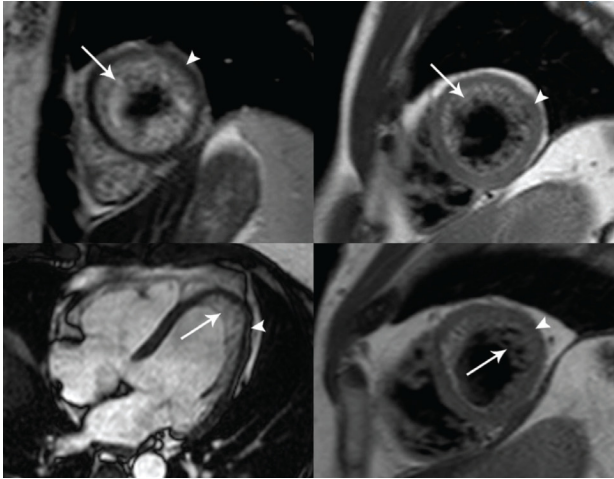


Fig. 2. **A.** Short axis proton density image of patient 4. **B.** Short axis proton density image of his/her brother, patient 5. **C** and **D.** Cine MRI image in four chambers and short axis proton density image in patient 13. The arrows show the non-compacted myocardium and the tops of the arrows show the compacted myocardium

Statistical analysis

The quantitative variables were expressed as mean \pm DS. Student's t-test was used in order to compare the variables between both groups. The significance level was established in $p < 0.05$. The correlation between 2 constant variables was calculated using the linear regression model. The comparison of nominal or dichotomous variables was done with the Chi-square test. The analyses were done using StatsDirect statistical software (Version 2.6.5, Altrincham, UK).

RESULTS

70 % of the population were men and the mean age was 49.6 ± 13.8 . The coronary anatomy was known in 7 patients; 5 had ventricular dysfunction and 2 had preserved ventricular function. Only one had mild coronary disease in the left anterior descending artery. One patient had maternal history of NCC and 3 showed family histories of dilated cardiomyopathy and sudden death. Two patients with perfusion defects in gamma camera and 4 patients derived due to IDC had normal coronary arteries. Chart 1 shows the characteristics of the studied population.

Left ventricular involvement

The average thickness of NC myocardium and C myocardium was 13.1 ± 3.3 mm and 3.6 ± 0.6 mm respectively. Only one patient had C myocardial thickness higher than 5 mm. The average number of segments with absence of compaction was 8.2 ± 1.2 . The more affected segments were the apex of the heart and the lateral segments at the apical and mid-ventricular level. In none of the cases, the basal

anteroseptal and inferoseptal segments were affected. There were no differences statistically significant among the patients with and without left ventricular dysfunction according to age, sex, mean NC/C and LVTM %. LVEDD, LVEDV, global LV mass, compacted LV mass and LVTM were significantly increased in the group of patients with ventricular dysfunction (Chart 2).

LVEF had a negative linear correlation with the LVTM/m² ($R = 0.67$, $p = 0.001$), LVEDV/m² ($R = 0.77$, $p < 0.001$) and LVEDD/m² end-diastolic diameter ($R = 0.71$, $p < 0.001$). There was also a positive linear correlation between LVTM/m² and LVEDV/m² ($R = 0.76$, $p < 0.001$) (Figure 2). 4 of the 14 patients that received gadolinium presented delay-enhancement (28%).

In 5 of the 18 patients with a previous echocardiogram, NC myocardium was correctly recognized. Doppler echocardiography was repeated in 5 patients, three of them were symptomatic with ventricular dysfunction. Intertrabecular flow in 5 patients and a myocardial mean NC/C > 2 in 2 of them were objectified (Figure 3).

DEBATE

NCC is a less frequent pathology gathered within the genetic primary cardiomyopathies (17). Although NCC can be associated to several congenital heart defects (18-20), it is usually evident in an isolated way. At the beginning, the prevalence of NCC was very low, $< 0.3\%$ (3). Recent publications show a prevalence of 23% in patients with heart failure (10).

NCC is originated due to the interruption of the myocardial compaction process that happens between the 4th and 8th weeks of gestation, moment in which the embryonic recesses are gradually converted into C myocardium from the epicardium to the endocardium, from the septum to the free wall and from the base to the apex (21). This explains the predominance of NC myocardium at the apex level and in the left ventricular lateral part, and also the absence of the medial and basal septal affection (13, 22, 23).

Several publications confirm the family factor of NCC. The genetic pattern of NCC involves gene mutation related to the mitochondrial function, as protein G 4.5, and gene mutation related to the synthesis of protein cypher/ZASP and lamin A/C protein (5-7). These mutations take part in the development of different cardiomyopathies such as hypertrophic and dilated cardiomyopathies; that is why it is possible to find different kinds of cardiomyopathies within the same family (4-7, 24). What was mentioned supports the family screening in carriers of NCC.

In the studied population, the large clinical pleomorphism of NCC is identified. The volume overload that takes place immediately after birth makes NCC to be confused with peripartum cardiomyopathy (25). The anomalous development of

Table 1. Clinical characteristics, reasons for the request and findings of Cardiac MRI

Patient	Sex	Age	Study request	Clinic	NC mm	C Mm	Mean (NC/C)	Location	LVEDD mm	LVEDV ml/m ²	LVEF %	LVTM %
1	F	43	Cardiac mass	Asymptomatic	9.4	3.5	2.69	7, 10-17	46.3	57.0	59.2	42.0
2	M	66	IDC	Class I-II dyspnea	12.1	3.4	3.54	7, 10-13, 15-17	70.0	126.0	25.4	38.5
3	M	68	PFO.ISA	AP	8.7	2.9	3.00	7, 10-13,15-17	53.1	61.4	56.5	35.3
4	F	35	PPCM	Asymptomatic	12.4	3.0	4.13	7, 10-17	47.0	72.5	48.4	48.1
5	M	34	Screening	Asymptomatic	13.1	3.7	3.54	7, 10-17	51.8	75.5	40.0	42.8
6	M	66	RPD	Class I-II dyspnea	11.2	2.6	4.31	10-17	63.1	106.6	45.1	30.9
7	M	58	IDC	AP	12.3	4.3	2.86	7, 10-12, 15, 17	63.0	97.6	37.5	34.9
8	M	40	PFO	Asymptomatic	16.3	3.0	5.43	7, 10-17	58.5	103.6	69.2	33.9
9	M	50	Medical checkup	Asymptomatic	15.8	3.8	4.16	7, 10-13, 15-17	58.0	74.2	54.6	30.2
10	F	42	Medical checkup	Asymptomatic	19.5	3.5	5.57	1, 12, 13, 15-17	53.7	74.9	53.0	40.2
11	F	48	RPD	AP	13.3	3.5	3.80	7, 10-13, 15-17	57.5	91.6	50.0	36.6
12	F	55	Screening	Asymptomatic	9.3	3.6	2.58	5-7, 10-12	48.6	53.1	65.5	28.6
13	M	53	NCC-CT	AP	10.4	4.4	2.63	7, 10-13, 15-17	56.3	71.5	52.6	30.6
14	M	42	NCC-Echo	Asymptomatic	14.8	5.4	2.74	7, 8, 10-17	59.0	89.9	52.6	37.9
15	M	16	Screening	Asymptomatic	9.3	3.4	2.74	1, 7, 11-13, 15-17	54.1	82.0	59.0	29.4
16	M	59	IDC	Class III-IV dyspnea	16.2	4.8	3.38	11-13, 15-17	79.7	181.5	9.9	33.7
17	F	35	PPCM	Asymptomatic	15.8	4.4	3.59	7, 10-17	63.2	124.8	18.5	42.7
18	M	69	Cardiac hydatid disease	Class II-III dyspnea	13.4	3.4	3.94	10-13, 15-17	70.5	146.2	34.6	47.1
19	M	60	IDC	Class I-II dyspnea	19.9	3.5	5.64	1, 4-7, 10-17	68.1	127.7	34.9	39.8
20	M	54	Ventricular arrhythmia	Asymptomatic	9.1	3.4	2.68	1, 7, 11-13, 15-17	60.9	93.8	66	37.8

Cardiac MRI: Cardiac Magnetic Resonance Imaging. M: Male. F: Female. NC: Non-compacted myocardium. C: Compacted myocardium. AP: Angina Pectoris. RPD: Reversible Perfusion Defect. IDC: Idiopathic Dilated Cardiomyopathy. PFO: Permeable Foramen Ovale. ISA: Interatrial Septal Aneurysm. PPCM: PeriPartum CardioMyopathy. LVEDD: Left Ventricular End-Diastolic Diameter. LVEDV: Left Ventricular End-Diastolic Volume. LVEF: Left Ventricular Ejection Fraction. LVTM%: LVTM percentage in relation to the global left ventricular mass. The distribution of NC myocardium was carried out with the model of 17 myocardial segments from AHA classification (15).

Table 2. Comparison of patients with and without left ventricular dysfunction

Variables	Normo-function n=11	Dysfunction N=9	P
Male	63.6%	77.8%	0.425
Age	46.4 (±12.9)	53.5 (±14.6)	0.264
LVEDD (mm)	55 (±4.5)	64 (±9.8)	0.014
LVEDV/sup	77.6 (±16.2)	117.6 (±34.3)	0.002
LVEF (%)	58.5 (±6.5)	32.7 (±12.5)	<0.001
Number of affected segments	8 (±1.2)	8.3 (±2.1)	0.752
Thickness of NC myocardium (mm)	12.4 (±3.6)	14 (±2.7)	0.302
Thickness of C myocardium (mm)	3.6 (±0.8)	3.6 (±0.7)	0.917
Mean MN/MC	3.4 (±1.1)	3.8 (±0.7)	0.369
Global LV mass (gr/m ²)	77.3 (±20)	118 (±22.7)	0.013
Compacted LV mass (gr/m ²)	49.9 (±13.7)	72 (±17.4)	0.025
LVTM (gr/m ²)	27.4 (±7.6)	46 (±8.9)	<0.001
LVTM (%)	34.8 (±4.6)	39.8 (±5.9)	0.052

LV: Left ventricle. LVEDD: Left Ventricular End-Diastolic Diameter. LVEDV: Left Ventricular End-Diastolic Volume. LVEF: Left Ventricular Ejection Fraction. NC: Non-compacted. C: Compacted. Global LV mass: Global Left Ventricular mass. Compacted LV mass: Compacted Left Ventricular mass. LVTM: Left Ventricular Trabecular Mass. LVTM%: LVTM percentage in relation to the global LV mass.

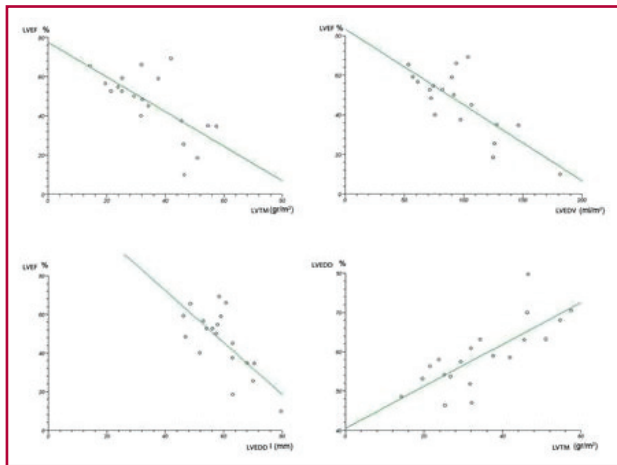


Fig. 3. The graphs show the linear relationship between: **A.** The increase of LVTM and the decrease of LVEF. **B.** The increase of LVEDV/ m^2 and the decrease of LVEF. **C.** The increase of LVEDD/ m^2 and the decrease of LVEF. **D.** The increase of LVTM/ m^2 and the increase of LVEDD.

LV: left ventricle; LVEF: left ventricular ejection fraction; LVTM: left ventricular trabecular mass; LVEDV: left ventricular end-diastolic volume; LVEDD: left ventricular end-diastolic diameter; LVTM: left ventricular trabecular mass.

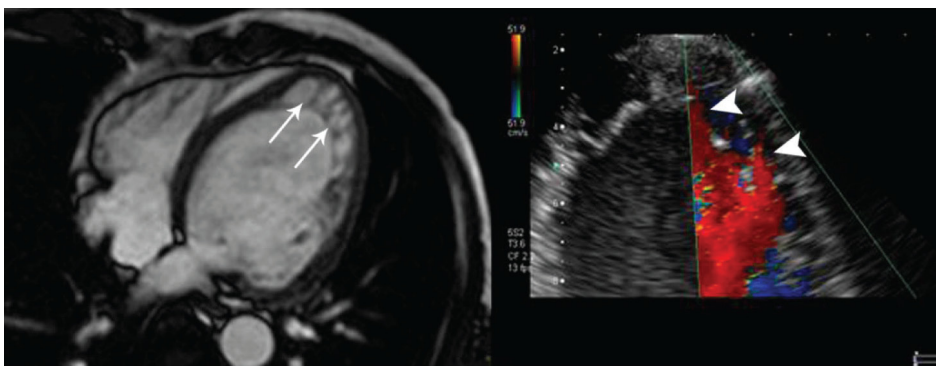


Fig. 3. A. Cine MRI image in 4 chambers of patient 2. The arrows show the non-compacted myocardium. **B.** Four-chamber color Doppler echocardiography. The tops of the arrows show the intertrabecular flow.

microcirculation in the non-compacted areas would be related to the presence of angina pectoris and perfusion defects in absence of coronary disease (26). Literature also describes the association of NCC with interatrial septal aneurysm and permeable foramen ovale (27).

Petersen et al. (13) established the first diagnostic criterion of cardiac MRI, showing that a mean NC/C > 2.3 differentiated correctly NCC from minor forms of compaction present in normal individuals, sportmen, patients with hypertrophic and dilated cardiomyopathy, and aortic stenosis. The inclusion of NC myocardium inside the ventricular cavity is the more efficient and reproducible way of ventricular volume measurements and LVEF (16). Recently, Jacquier et al. (14) included the second diagnostic criterion of cardiac MRI. LVTM % > 20 differentiated correctly the NCC trabeculation in normal individuals, patients with hypertrophic and dilated cardiomyopathy. When comparing this diagnostic criterion with that of Petersen et al. (13) a sensitivity and a specificity of 78.5% and 72.2% respectively were obtained (14). The determination of LVTM has the limitation of including the blood volume placed in intertrabecular recesses.

The similarity in the parameters of cardiac MRI among patients who attended due to family screening and those that did not, is in favour of the pathology presence in our population (unknown data).

A thickness of N myocardium < 5 mm in NC areas reinforces pathology diagnosis; this information is usually not considered in the literature.

The presence of ventricular dysfunction does not coincide with mean NC/C or with the percentage of NC myocardium, but with global LV mass, compacted LV mass, LVTM, LVEDV and LVEDD, that is, it works as a dilated cardiomyopathy. On the other side, it is probable that cardiac MRI is looking with a magnifying glass subtle ways or in early stages and/or non-pathological hypertrabeculation, not clear in echocardiography. Nowadays we do not have imaginology parameters that predict the evolution towards symptomatic ventricular dysfunction in young people. Our results agree with those of Belager et al. (28), who found a relation between the increase of the NC myocardial area and the mean NC/N, and the decrease of the systolic function through echocardiographic evaluation, in patients with several degrees of myocardial trabeculation with no requirement of having to fulfill with the presence of the mean NC/C > 2 in the end-systole. Authors considered that NCC has a presentation spectrum which goes from mild to severe myocardial trabeculation (28).

The absence of genetic analysis in our population makes us suppose that patients with ventricular dysfunction could have genes related to dilated cardiomyopathy or have a genetic pattern similar to those who present preserved systolic function,

but with different phenotypic expression due to the influence of environmental factors. As it happened with hypertrophic cardiomyopathy, it is time to incorporate the genetic determination inside the risk stratification in patients with NCC (29), especially in those with preserved systolic function. The answer to the questionings that arised from our observation will be revealed through longitudinal studies with genetic mapping.

The classification of Chin et al. (11) considers the mean C/NC in the end-diastole, taking a value of ≤ 0.5 , without taking into account the evaluation due to Doppler of intertrabecular flow. The most used classification is Jenny et al. (12), which uses a mean NC/C of end-systole > 2 plus the objectification of intertrabecular flow due to Doppler color. The classification of Stolberg et al. (3) adds to that of Jenny et al. (12) the presence of more than three trabeculations that protrude in the left ventricular wall at the apical level, in front of the papillary muscles, which are visible at an echocardiographic plane; this criterion is taking increasing interest.

A patient can have some of these diagnostic criteria but not all of them. The presence of intertrabecular flow is a characteristic, clear and easy to find sign (12), but not the measurement of thickness in both myocardial layers, since each classification measures them in different phases of cardiac cycle. The measurement of NC myocardium in end-systole is difficult due to the combination of trabeculae and the decrease of intertrabecular recesses that produce the cardiac contraction.

Kohli et al. (10) showed that only 29.8% of the patients with NCC gathered the echocardiographic diagnostic criteria of different classifications. These findings coincide with the minor detection of the mean in end-systole between NC/C myocardium < 2 than the presence of intertrabecular flow in individuals who had a second Doppler echocardiogram. On the other hand, the absence of recognition in non-compaction areas in previous echocardiograms could be related to the used equipment. Considering the echocardiographic suspicion of NCC, the execution of a cardiac MRI could clarify the diagnosis.

Clinical implications

The first publications evaluated severe ways of NCC with high incidence of heart failure, thromboembolic events, arrhythmias, and sudden death (3, 11). However, NCC has a more favourable evolution, with a survival free of cardiac transplant or death of 97% after 46 months of monitoring (24).

Although the several ways of presentation and the limited knowledge of the pathology, cardiologists should suspect the presence of NCC in patients with dilated cardiomyopathy of non-established etiology, even without severe worsening of LVEF, as well as, in those that have nuclear studies with perfusion defects dissociated from a coronary pathology.

Limitations

Although NCC is a less predominant affection and that its results come from an only centre, the execution of multicenter studies with longitudinal monitoring in a major number of patients is necessary to validate our findings.

Owing to the fact that there has not been genetic determination in the studied population, the NCC finding in direct family members suggests the genetic pattern of this entity.

CONCLUSIONS

Cardiac MRI through a mean NC/C > 2.3 and a LVTM $\% > 20$ recognizes correctly NCC. There are two ways of presenting the pathology, a subtle or mild with preserved systolic function and another associated to a ventricular dysfunction that behaves as dilated cardiomyopathy. The uneven echocardiographic criteria and the low suspicion rate would be the determining factors of NCC sub-diagnosis in the clinical practice.

RESUMEN

Evaluación de la miocardiopatía no compactada con resonancia magnética cardíaca en pacientes con función sistólica del ventrículo izquierdo conservada y disminuida

Introducción

La miocardiopatía no compactada (MNC) es un trastorno genético que se caracteriza por la presencia de una extensa capa de miocardio trabeculado con recesos intertrabeculares comunicados con la cavidad ventricular. Si bien su prevalencia es mayor en poblaciones sintomáticas con disfunción ventricular, las nuevas modalidades diagnósticas incrementaron su detección en pacientes asintomáticos con función sistólica conservada. No obstante, la patología permanece subdiagnosticada debido al bajo índice de sospecha sumado al uso de clasificaciones ecocardiográficas con criterios diagnósticos dispares. Se establecieron dos criterios diagnósticos de resonancia magnética cardíaca (RMC) que reconocen correctamente esta entidad.

Objetivo

Evaluar las características clínicas y morfológicas de los pacientes con MNC con disfunción sistólica y sin ella evaluados por resonancia magnética cardíaca (RMC).

Material y métodos

Se incluyeron en forma retrospectiva 20 pacientes con diagnóstico de MNC. Se determinaron: volumen de fin de diástole (VFDVI) y de sístole, diámetro de fin de diástole (DFDVI) y de sístole, fracción de eyección (FEVI), masa cardíaca y trabeculaciones del ventrículo izquierdo (VI). La distribución del miocardio NC se llevó a cabo con el modelo de 17 segmentos miocárdicos.

Resultados

El espesor medio del miocardio NC y el miocardio C fue de $13,1 \pm 3,3$ mm y de $3,6 \pm 0,6$ mm, respectivamente.

El DFDVI, el VFDVI, la masa global, compactada y trabeculada del VI estuvieron incrementados en forma significativa en el grupo de pacientes con disfunción ventricular. Hubo una correlación positiva y una relación lineal entre el DFDVI y la M_{TVI} (g/m²): $r = 0,76$; $r^2 = 0,59$; $p < 0,001$.

Conclusiones

Hallamos dos formas de presentación de la patología, una sutil con función sistólica conservada y otra asociada con disfunción ventricular que se comporta como la miocardiopatía dilatada.

Palabras clave > Miocardiopatía no compactada - Resonancia magnética cardíaca - Trabeculaciones - Miocardiopatías - Ecocardiograma

BIBLIOGRAPHY

- Weiford BC, Subbarao VD y Mulhern KM. Non-compaction of the Ventricular Myocardium. *Circulation* 2004; 109: 2965-71.
- Chiribiri A, Leuzzi S, Salvetti I, Patané S, Bonamini R, Trevi GP et al. Isolated noncompaction of the right ventricular myocardium in the adulthood?. *Int J Cardiol.* 2009; 134: 17-9.
- Oechslin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R. Long-term follow-up of 34 adults with isolated left ventricular noncompaction a distinct cardiomyopathy with poor prognosis. *J Am Coll Cardiol* 2000; 36: 493-500.
- Monserrat L, Hermida-Prieto M y Castro-Beiras A. Avances en miocardiopatía dilatada idiopática: del genotipo al fenotipo clínico. *Rev Esp Cardiol Supl.* 2007; 7: 2F-13F.
- Neustein HB, Lurie PR, Dahms B, Takahashi M. An X-linked recessive cardiomyopathy with abnormal mitochondria. *Pediatrics* 1979; 64: 24-9.
- Vatta M, Mohapatra B, Jimenez S, Sanchez X, Faulkner G, Perles Z et al. Mutations in Cypher/ZASP in patients with dilated cardiomyopathy and left ventricular non-compaction. *J Am Coll Cardiol* 2003; 42: 2014-27.
- Hermida-Prieto M, Monserrat L, Castro-Beiras A, Laredo R, Soler R, Peteiro J, et al. Familial dilated cardiomyopathy and isolated left ventricular noncompaction associated with lamin A/C gene mutations. *Am J Cardiol.* 2004; 94: 50-4.
- Stollberger C, Finsterer J, Blazek G. Left ventricular hypertrabeculation/noncompaction and association with additional cardiac abnormalities and neuromuscular disorders. *Am J Cardiol* 2002; 90:899-902.
- Stöllbergera C, Blazekb G, Winkler-Dworakc M y Finsterera J. Diferencias de sexo en la ausencia de compactación ventricular izquierda con y sin trastornos neuromusculares. *Rev Esp Cardiol.* 2008; 61(2): 130-6.
- Kohli SK, Pantazis AA, Shah JS, Adeyemi B, Jackson G, McKenna WJ et al. Diagnosis of left-ventricular non-compaction in patients with left-ventricular systolic dysfunction: time for a reappraisal of diagnostic criteria? *Eur Heart J* 2008; 29: 89-95.
- Chin TK, Perloff JK, Williams RG, Jue K, Mohrmann R. Isolated noncompaction of left ventricular myocardium. A study of eight cases. *Circulation* 1990; 82 : 507-13.
- Jenni R, Oechslin E, Schneider J, Attenhofer Jost C, Kaufmann PA. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. *Heart* 2001; 86: 666-71.
- Petersen SE, Selvanayagam JB, Wiesmann F, Robson MD, Francis JM, Anderson RH et al. Left Ventricular Non-Compaction Insights From Cardiovascular Magnetic Resonance Imaging. *J Am Coll Cardiol* 2005; 46:101-105.
- Jacquier A, Thuny F, Jop B, Giorgi R, Cohen F, Gaubert JY et al. Measurement of trabeculated left ventricular mass using cardiac magnetic resonance imaging in the diagnosis of left ventricular non-compaction. *Eur Heart J.* 2010; 31: 1098-1104
- Cerqueira MD, Weissman NJ, Dilsizian V, Jacobs AK, Kaul S, Laskey WK et al. Standardized Myocardial Segmentation and Nomenclature for Tomographic Imaging of the Heart: A Statement for Healthcare Professionals From the Cardiac Imaging Committee of the Council on Clinical Cardiology of the American Heart Association. *Circulation* 2002; 105: 539-42.
- Papavassiliu T, Kuhl HP, Schroder M, Suselbeck T, Bondarenko O, Bohm CK et al. Effect of endocardial trabeculae on left ventricular measurements and measurement reproducibility at cardiovascular MR imaging. *Radiology* 2005; 236: 57-64.
- Maron BJ, Towbin JA, Thiene G, Antzelevitch C, Corrado D, Arnett D, et al. Contemporary definitions and classification of the cardiomyopathies: an american heart association scientific statement from the council on clinical cardiology, heart failure, and transplantation committee; quality of care and outcome research and functional genomics and translational biology interdisciplinary working groups; and council on epidemiology and prevention. *Circulation.* 2006; 113: 1807-16.
- Ichida F, Tsubata S, Bowles KR, Haneda N, Uese K, Miyawaki T et al. Novel gene mutations in patients with left ventricular non-compaction or Barth syndrome. *Circulation* 2001; 103: 1256-63.
- Ilercil A, Barack J, Malone MA, Barold SS, Herweg B. Association of noncompaction of left ventricular myocardium with Ebstein's anomaly. *Echocardiography.* 2006; 23: 432-3.
- Bellet S, Gouley BA. Congenital heart disease with multiple cardiac anomalies: report of a case showing aortic atresia. *Am J Med Sci.* 1932; 183: 458-65
- Weiford BC, Subbarao VD y Mulhern KM. Non-compaction of the Ventricular Myocardium. *Circulation* 2004; 109: 2965-71.
- Dursun M, Agayev A, Nisli K, Ertugrul T, Onur I, Oflaz H et al. MR imaging features of ventricular noncompaction: emphasis on distribution and pattern of fibrosis. *Eur J Radiol.* 2010; 74: 147-51.
- Calvillo P, Martí-Bonmatí L, Chaustre F, Roldán I, Mora V, Peláez A et al. Cuantificación de la función, perfusión y realce tardío del ventrículo izquierdo no compactado mediante resonancia magnética. *Radiología.* 2009; 51(1): 45-56.
- Murphy RT, Thaman R, Blanes JG, Ward D, Sevdalis E, Papa E et al. Natural history and familial characteristics of isolated left ventricular non-compaction. *Eur Heart J.* 2005; 26: 1143-4.
- Bahl A, Swamy A, Sharma Y, Kumar N. Isolated noncompaction of left ventricle presenting as peripartum cardiomyopathy. *Int J Cardiol.* 2006; 109: 422-3.
- Sato Y, Matsumoto N, Matsuo S, Kunimasa T, Yoda S, Tani S et al. Myocardial perfusion abnormality and necrosis in a patient with isolated noncompaction of the ventricular myocardium: evaluation by myocardial perfusion SPECT and magnetic resonance imaging. *Int J Cardiol.* 2007 21; 120: 24-6.
- Monserrat L, Hermida-Prieto M, Fernandez X, Rodríguez I, Dumont C, Cazón L, Margarita G. Cuesta MG et al. Mutation in the alpha-cardiac actin gene associated with apical hypertrophic cardiomyopathy, left ventricular non-compaction, and septal defects. *Eur Heart J.* 2007; 28: 1953-61.
- Dabarian AL, Mady C, Rochitte CE, Shiozaki AA, Lemos PA, Salemi VM et al. An unusual case of angina pectoris: a patient with isolated non-compaction of the left ventricular myocardium. *Eur J Echocardiogr.* 2008; 9: 728-30.
- Monserrat L. Miocardiopatía no compactada: una enfermedad en búsqueda de criterios. *Rev Esp Cardiol.* 2008; 61: 112-5.

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