Pediatr Radiol (2005) 35: 1147–1151
DOI 10.1007/s00247-005-1551-2

WORK IN PROGRESS

Fahad Alhabshan · Jeffrey F. Smallhorn
Fraser Golding · Norman Musewe
Robert M. Freedom · Shi-Joon Yoo

Extent of myocardial noncompaction: comparison between MRI and echocardiographic evaluation

Received: 18 March 2005 / Revised: 19 June 2005 / Accepted: 24 June 2005 / Published online: 6 August 2005

Abstract ***Background***: Noncompaction of the left ventricular myocardium is an important cause of cardiomyopathy. There is no clear consensus about its diagnostic criteria or the diagnostic test of choice. MRI is increasingly used in the pediatric cardiac field because of its superior and objective image quality. ***Objective***: To compare the echocardiographic and MRI findings in four patients with recently diagnosed ventricular noncompaction. ***Materials and methods***: We compared the extent of myocardial involvement shown at MRI and echocardiography in four individuals, two patients with echocardiographic diagnosis of left ventricular noncompaction, and two family members of one of the patients. ***Results***: In all patients, MRI showed wider area of involvement than echocardiography. A definite diagnosis was entertained in only two patients by echocardiography but in all by MRI. Cine imaging was diagnostic of the disease in all patients. Black-blood pool imaging with double-inversion recovery sequence also helped to visualize the abnormal areas by showing slow flow artifacts in the four- and two-chamber images. ***Conclusion***: MRI provided better delineation of the extent of the abnormal trabeculation in patients with noncompaction of the left ventricular myocardium. It was particularly useful when the myocardial involvement was subtle, as in the asymptomatic family members.

Keywords Noncompaction · MRI · Echocardiography

F. Alhabshan · J. F. Smallhorn · F. Golding · N. Musewe
R. M. Freedom
Department of Pediatrics, Division of Cardiology,
Hospital for Sick Children, University of Toronto,
Toronto, Ontario, Canada

S.-J. Yoo (✉)
Department of Diagnostic Imaging, Division of Cardiology,
Hospital for Sick Children, University of Toronto,
555 University Ave, M5G1X8 Toronto, Ontario, Canada
E-mail: shi-joon.yoo@sickkids.ca
Tel.: +1-416-8136037
Fax: +1-416-8137591

Introduction

Noncompaction of the left ventricular (LV) myocardium is an uncommon disorder characterized by increased trabeculations and deep intertrabecular recesses [1]. It can present as an isolated disorder or it can be associated with other congenital cardiac malformations. This condition is being diagnosed with increasing frequency. This is probably a result of increasing awareness of the characteristic echocardiographic features. Furthermore, there is more familiarity with some of the clinical manifestations, including ventricular dysfunction, subsequent heart failure, life-threatening arrhythmia and thromboembolism [1–6]. This myocardial disorder is still considered an unclassified cardiomyopathy by WHO definition [7].

Echocardiography has been used as the main diagnostic tool for this disease [1, 8, 9]. Recently, it has been suggested that MRI can be used as an alternative diagnostic method with a better yield [10–13]. The aim of this study was to compare the echocardiographic and MRI findings in four patients with recently diagnosed ventricular noncompaction.

Materials and methods

The study population consisted of two patients with a suspected diagnosis of LV myocardial noncompaction and two family members of one of the patients. All four underwent echocardiography and MRI within a 12-week period.

The first patient, patient 1, was a 17-year-old girl who presented with syncope and was found to have dilated LV with increased trabeculations on echocardiography. The second patient, patient 2, was a 17-year-old boy who had been treated for congestive heart failure since 4 months of age. His condition was initially believed to be secondary to viral myocarditis; however, the diagnosis was changed to familial dilated cardiomyopathy when his sister was

diagnosed with a similar disease. His 12-year-old sister, patient 3, was diagnosed as having a dilated cardiomyopathy when she presented with tachycardia at 1 month of age, at which time she was observed to have mild LV dilatation on echocardiography. Another sibling, a 14-year-old asymptomatic boy, patient 4, was screened at the age of 2 years for dilated cardiomyopathy and also found to have mild dilatation of his LV.

Transthoracic echocardiographic studies were performed using an HP Sono 5500 (Hewlett Packard, Andover, Mass.) or ATL HDI 5000 system (Philips Ultrasound, Bothell, Wash.). Videotaped echocardiograms were blindly reviewed by two experienced pediatric cardiologists. MRI studies were performed using 1.5 T scanner (GE CV/i; General Electric Medical Systems, Milwaukee, Wis.) by using electrocardiographic-gated white-blood pool, cine imaging with steady-state free precession sequences and static black-blood pool imaging with double-inversion recovery sequences. Electronic MR images were blindly reviewed at an MRI workstation by an experienced cardiac radiologist.

The reviewers were asked to describe the pattern and extent of the LV trabeculations as well as the ratio of the compacted to the noncompacted myocardial thickness. For the assessment of the extent of involvement, the LV wall was divided into nine segments: one apical, four mid-ventricular and four basal (septal, anterior, lateral, and inferior) [3]. Each reviewer was asked to describe whether the degree of trabeculations was heavy, moderate or few. The LV function was evaluated by measuring the ejection fraction by M-mode from the parasternal long-axis echocardiographic images, and by measuring the LV end-systolic and end-diastolic volumes from the short-axis MR images. The final diagnosis of myocardial noncompaction was entertained when the maximum ratio of the thickness between the noncompacted and compacted layers in end systole was ≥2:1 by either echocardiography or MRI.

Results

There was no significant difference between the findings of the two cardiologists. A definite diagnosis of myocardial noncompaction of the LV was entertained in only two patients by echocardiography but in all by MRI (Table 1). In patients 3 and 4, who were siblings of patient 2, echocardiography underestimated the degree of LV trabeculations, the ratio of noncompacted to compacted layer, and the extent of involvement (Fig. 1). Echocardiography was sensitive in depicting abnormalities of the apex and posterior wall of the LV, but it

Table 1 Comparison between the echocardiographic (*Echo*) and MRI findings (*NC/C* noncompacted/compacted, + disease present, *0* disease not present)

Patient	Trabeculations		NC/C ratio		LV size		LV ejection fraction (%)		Diagnosis	
	Echo	MRI	Echo	MRI	Echo[a]	MRI[b]	Echo	MRI	Echo	MRI
1	Moderate	Heavy	2.9:1	3.3:1	4.6	88	53	56	+	+
2	Heavy	Heavy	3:1	3:1	6.1	116	56	53	+	+
3	Few	Moderate	1.8:1	2:1	4.8	91	53	55	0	+
4	Few	Moderate	1.2:1	2:1	4.7	96	53	63	0	+

[a]Left ventricular end-diastolic dimension in centimeters; normal value 3.5–5 cm
[b]Left ventricular end-diastolic volume index in milliliters per meter squared; normal value 44–89 ml/m^2

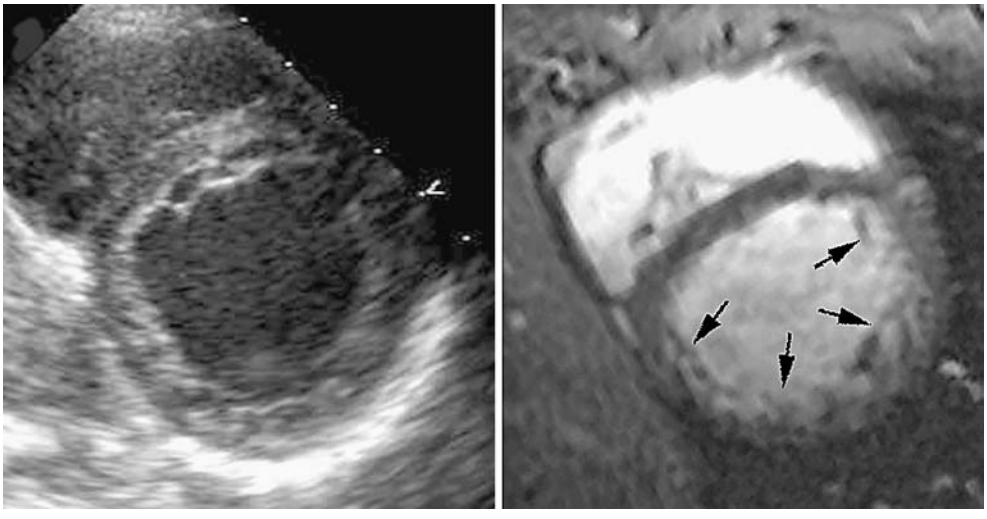

Fig. 1 Short-axis echocardiogram and cine MR image from patient 4. Echocardiogram was reported as normal. In the cine MR image, myocardial noncompaction is evident (*arrows*)

Table 2 Comparison between MRI and echocardiographic (*Echo*) findings of abnormal trabeculation on the different LV segments (+ number of patients in whom abnormal trabeculation was observed, *0* none of the patients)

LV segment	Echo	MRI
Basal septum	0	0
Basal anterior wall	0	+
Basal lateral wall	+	+ +
Basal inferior wall	+	+ +
Mid-ventricular septum	0	+ +
Mid-ventricular anterior wall	+	+ + + +
Mid-ventricular lateral wall	+ +	+ + + +
Mid-ventricular inferior wall	+ +	+ + + +
Apex	+ + + +	+ + + +

underestimated the involvement of the mid-septal region in all (Table 2). Unlike echocardiography, MRI clearly demonstrated not only the heavy but also the fine lacy trabeculations (Fig. 2). Compared to echocardiography, MRI showed a higher ratio of the noncompacted to compacted myocardium in all but one patient (Table 1). MRI revealed trabeculations that mainly involved the apical part of the LV, with a variable degree of extension toward the ventricular base (Fig. 2). In both long- and short-axis views, a thin interrupted layer of tissue demarcated the endocardial side of the noncompacted myocardial layer. Thick and thin trabeculations traversed the space between this thin layer and the compacted myocardium. The hypertrabeculated area showed slow flow artifact in double-inversion recovery images in all patients (Fig. 3). All four patients had low normal ejection fraction in the range 53–56%. The LV end diastolic dimension was in the upper range of normal for three patients, while the fourth had a dilated LV. There was a good correlation between the echocardiographic and MRI measurements.

Discussion

Myocardial noncompaction of the LV is characterized by abnormally prominent trabeculations and intervening recesses in the endocardial side of the myocardium. However, there is a wide variation in the extent and depth of myocardial involvement. In addition, the clinical presentation varies considerably, with some patients showing no clinical symptoms and others presenting with severe heart failure or arrhythmias [3–5]. It has

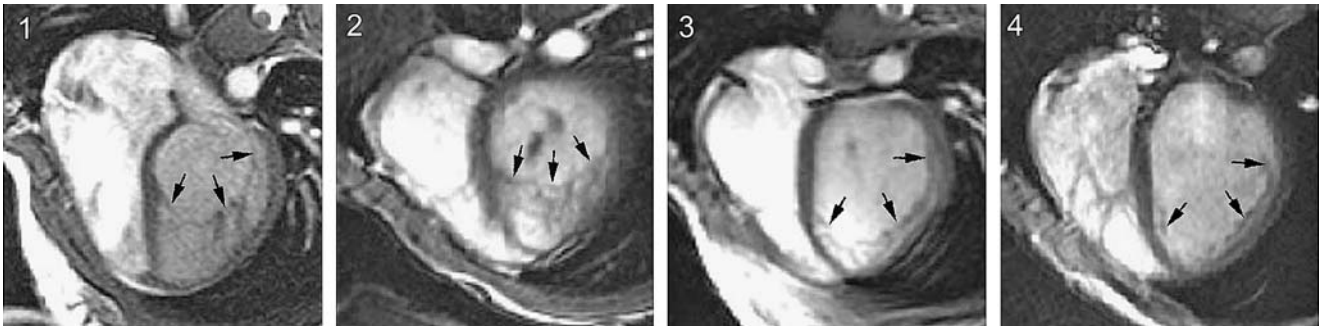

Fig. 2 Cine MR images in horizontal long-axis plane from patients 1–4 show that myocardial noncompaction involved the apical part of the LV in all patients and that there was variable degree of extension toward the ventricular base. Note that a thin interrupted membranous layer (*arrows*) delineates the endocardial aspect of the noncompacted layer. Free wall involvement extends well to the atrioventricular junction in patient 1

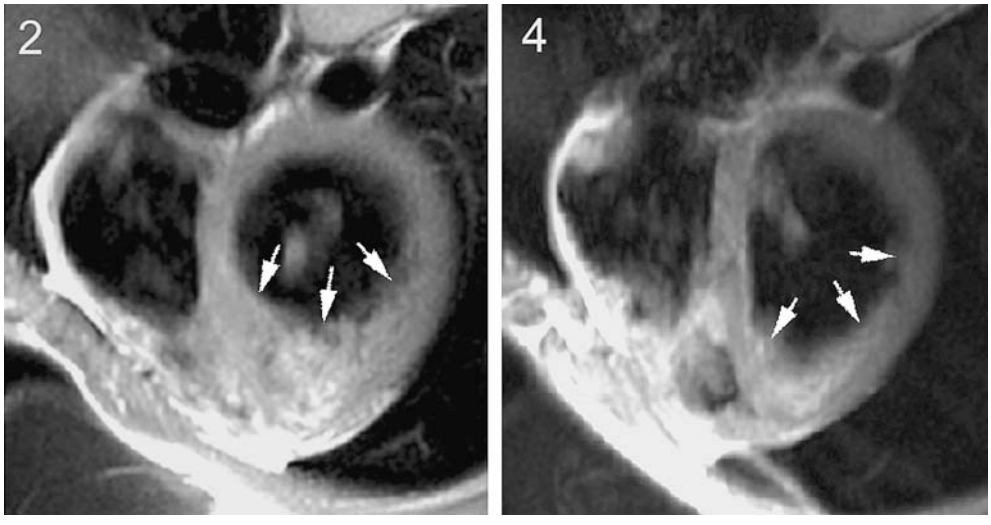

Fig. 3 Static black-blood pool MR images with double-inversion recovery sequence from patients 2 and 4. Note the high signal in the LV from slow flow artifact between the trabeculation (*arrows*). Compare with the cine MR images shown in Fig. 2

been described both as an isolated abnormality and in association with other cardiac morphological abnormalities [6]. The morphological features of hearts with LV noncompaction appeared to be variable. Three basic features have been described: (1) extensive spongy transformation of the LV myocardium, (2) prominent coarse trabeculations of the ventricular wall with deep recesses of the ventricular cavity, and (3) dysplastic appearance of the myocardium with thinned myocardium and excessive trabeculations [6].

The MRI findings in our four patients are interesting in terms of morphology and pattern of involvement. The process invariably involved the LV apex, with a variable degree of extension toward the cardiac base along the free wall and ventricular septum (Table 2). Similar to the pathological description, the MRI appearance of the involved area showed prominent coarse trabeculations with deep intertrabecular recesses traversing between the compacted layer of the myocardium and the LV cavity. The thick trabeculations appeared thinner toward the LV cavity (Fig. 2). The space between the trabeculations invariably contained high signal intensity of slow flow artifact on black-blood images obtained by using double-inversion recovery sequences (Fig. 3). The sluggish blood flow in the intertrabecular spaces might explain why formation of a thrombus can complicate myocardial noncompaction.

The above features were present in all three members of the family with a variable degree of involvement. The difference among them was the extent of involvement and the depth of the space between the innermost layer and compacted myocardium. The mildest form noticed was in patient 4, who was entirely asymptomatic. His echocardiographic study was performed prior to his MRI and was interpreted as being normal. However, his MRI unequivocally demonstrated that he had LV noncompaction. This observation indicates that it is more appropriate to use MRI as a screening tool for family members of patients with an established diagnosis of ventricular noncompaction. It also indicates that the disease can have different presentations within one family.

Echocardiography has been the main diagnostic tool for the diagnosis of noncompaction. However, echocardiographic diagnosis of this uncommon disease requires experience as well as a high index of suspicion. Many reported cases have been misdiagnosed and labeled as dilated or hypertrophic cardiomyopathy [4]. The diagnosis can also be challenging in patients with difficult echocardiographic windows. Color Doppler and contrast agents can facilitate the diagnosis by demonstrating flow in the deep intertrabecular recesses [8, 14–16].

There is no clear consensus about the diagnostic criteria for this disease. Chin et al. [1] described a quantitative approach to diagnosing noncompaction using a trabeculation peak to trough ratio, but it has not been used widely in clinical practice. Oechslin et al. [3] and Jenni et al. [8] introduced different echocardiographic diagnostic criteria for this disease based on the findings of a two-layer myocardium, with a noncompacted to compacted ratio of more than 2:1, along with evidence of color Doppler flow within the trabeculations. Pignatelli et al. [5] proposed slightly different diagnostic criteria by including patients with structural heart disease and using a ratio of 1.4:1 between the noncompacted and the compacted myocardium. Our experience raises the question as to whether the diagnosis of myocardial noncompaction should be made based mainly on the ratio between the noncompacted and compacted layers. We think that the diagnosis should be suspected whenever echocardiography or MRI demonstrates hypertrabeculation with two-layered myocardium even if the ratio is less than 2:1. This should be differentiated from other forms of cardiomyopathies. In dilated cardiomyopathy, the myocardium is usually thin with few thin trabeculations. In hypertrophic cardiomyopathy, it is usually thick with thick trabeculations but without two distinct layers of compacted and noncompacted myocardium. This study showed that MRI is superior to echocardiography in defining the morphology and extent of myocardial noncompaction. It showed that myocardial noncompaction involves the ventricular apex, the LV free wall and the interventricular septum. We found that fast cine imaging with steady-state free precession sequence and static black-blood imaging with double-inversion recovery sequence are adequate in the evaluation of myocardial noncompaction. There was a good correlation between the hypertrabeculated areas shown at cine imaging and the areas showing slow flow artifact at static black-blood imaging.

In summary, MRI is a very useful tool in the diagnosis of suspected left ventricular myocardial noncompaction. It was found to be superior to echocardiography in screening family members of patients with an established diagnosis. The disease can have different presentations among family members. Slow flow artifact at black-blood imaging by using double-inversion recovery is helpful in defining the disease extent.

This study was limited by the small number of patients. Further prospective study is necessary to correlate the extent of the disease involvement and the severity of the clinical symptoms related to myocardial dysfunction.

References

1. Chin TK, Perloff JK, Williams RG, et al (1990) Isolated noncompaction of left ventricular myocardium. A study of eight cases. Circulation 82:507–513
2. Ritter M, Oechslin E, Sutsch G, et al (1997) Isolated noncompaction of the myocardium in adults. Mayo Clin Proc 72:26–31
3. Oechslin EN, Attenhofer Jost CH, Rojas JR, et al (2000) Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. J Am Coll Cardiol 36:493–500
4. Ichida F, Hamamichi Y, Miyawaki T, et al (1999) Clinical features of isolated noncompaction of the ventricular myocardium: long-term clinical course, hemodynamic properties, and genetic background. J Am Coll Cardiol 34:233–240

5. Pignatelli RH, McMahon CJ, Dreyer WJ, et al (2003) Clinical characterization of left ventricular noncompaction in children: a relatively common form of cardiomyopathy. Circulation 108:2672–2678
6. Stollberger C, Finsterer J (2004) Left ventricular hypertrabeculation/noncompaction. J Am Soc Echocardiogr 17:91–100
7. Richardson P, McKenna W, Bristow M, et al (1996) Report of the 1995 World Health Organization/International Society and Federation of Cardiology Task Force on the Definition and Classification of cardiomyopathies. Circulation 93:841–842
8. Jenni R, Oechslin E, Schneider J, et al (2001) Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. Heart 86:666–671
9. Sengupta PP, Mohan JC, Mehta V, et al (2004) Comparison of echocardiographic features of noncompaction of the left ventricle in adults versus idiopathic dilated cardiomyopathy in adults. Am J Cardiol 94:389–391
10. McCrohon JA, Richmond DR, Pennell DJ, et al (2002) Images in cardiovascular medicine. Isolated noncompaction of the myocardium: a rarity or missed diagnosis? Circulation 106:e22–23
11. Daimon Y, Watanabe S, Takeda S, et al (2002) Two-layered appearance of noncompaction of the ventricular myocardium on magnetic resonance imaging. Circ J 66:619–621
12. Borreguero LJ, Corti R, de Soria RF, et al (2002) Images in cardiovascular medicine. Diagnosis of isolated noncompaction of the myocardium by magnetic resonance imaging. Circulation 105:E177–178
13. Korcyk D, Edwards CC, Armstrong G, et al (2004) Contrast-enhanced cardiac magnetic resonance in a patient with familial isolated ventricular non-compaction. J Cardiovasc Magn Reson 6:569–576
14. Baumhakel M, Janzen I, Kindermann M, et al (2002) Images in cardiovascular medicine. Cardiac imaging in isolated noncompaction of ventricular myocardium. Circulation 106:e16–17
15. Koo BK, Choi D, Ha JW, et al (2002) Isolated noncompaction of the ventricular myocardium: contrast echocardiographic findings and review of the literature. Echocardiography 19:153–156
16. Lowery MH, Martel JA, Zambrano JP, et al (2003) Noncompaction of the ventricular myocardium: the use of contrast-enhanced echocardiography in diagnosis. J Am Soc Echocardiogr 16:94–96