



## EDITORIAL COMMENT

# Compacting knowledge in left ventricular non-compaction



## Compactar o conhecimento na não compactação ventricular esquerda

Alexandra Toste, Nuno Cardim\*

Multimodality Cardiac Imaging Department, Hospital da Luz, Lisbon, Portugal

Available online 14 September 2016

Left ventricular non-compaction (LVNC) is morphologically characterized by excessive trabeculation of the left ventricular (LV) walls with deep intertrabecular recesses that communicate with the ventricular cavity but not with the coronary circulation.<sup>1</sup>

It is a relatively recently described anomaly, still with many uncertainties and the subject of intense controversy and debate: there is no consensus regarding disease pathophysiology, classification or diagnostic criteria. Additionally, it is sometimes doubted whether this entity really exists as a single disease.

Though it has been thought to be a congenital heart disease resulting from incomplete morphogenesis of the endomyocardium, another theory argues that it is possible to acquire LVNC during life. Currently, it is recognized that physiological and pathophysiological processes associated with increased LV preload and afterload (including pregnancy, sports, chronic renal failure, sickle cell disease and heart valve disease) may lead to a cardiac morphology that also fulfills criteria for LVNC.<sup>2</sup>

LVNC is classified as a genetic cardiomyopathy by the American Heart Association,<sup>3</sup> whereas the European Society of Cardiology includes LVNC in the group of unclassified cardiomyopathies.<sup>4</sup>

However, the most important and debated point of controversy concerns LVNC diagnostic criteria. The most

frequently used criteria are those of Jenni,<sup>5</sup> who proposed the following: (1) the existence of two layers, one thin and compacted (C) and the other noncompacted (NC), and a ratio of NC/C >2 when measured in end-systole in parasternal short-axis view; (2) absence of other structural cardiac anomalies; (3) numerous, excessively prominent trabeculations and deep intertrabecular recesses; and (4) intertrabecular spaces perfused by blood flow demonstrated by color Doppler. Chin et al.<sup>6</sup> suggest as a diagnostic criterion an X-to-Y ratio  $\leq 0.5$ , where X is the distance between the epicardial surface and trough of a trabecular recess and Y is the distance from the epicardial surface to the peak of the trabeculation, as measured in images of the LV apex and free wall in end-diastole. Stöllberger et al.<sup>7</sup> propose as diagnostic criteria the existence of more than three trabeculations protruding from the LV wall, apical to the papillary muscles, visible in a single image plane, and intertrabecular spaces perfused from the ventricular cavity, visualized by color Doppler.

Cardiac magnetic resonance (CMR), with its excellent spatial resolution and good visualization of the LV apex, also has an important role in diagnosing LVNC. Petersen et al.<sup>8</sup> suggested as a criterion an NC/C ratio of >2.3 in diastole. On the other hand, Jacquier et al.<sup>9</sup> calculated the LV trabecular mass using steady-state free precession short-axis views and proposed a cut-off of LV trabecular mass above 20% of the total LV mass as predictive of LVNC. More recently, Captur et al.<sup>10</sup> described a new CMR tool, fractal analysis, which summarizes global LV trabecular complexity as a continuous variable termed fractal dimension; a fractal dimension  $\geq 1.3$  gave the optimal prediction for patients with LVNC.

DOI of original article:

<http://dx.doi.org/10.1016/j.repc.2016.04.009>

\* Corresponding author.

E-mail address: [ncardim@hospitaldaluz.pt](mailto:ncardim@hospitaldaluz.pt) (N. Cardim).

<http://dx.doi.org/10.1016/j.repc.2016.08.003>

0870-2551/© 2016 Sociedade Portuguesa de Cardiologia. Published by Elsevier España, S.L.U. All rights reserved.

The multiplicity of diagnostic criteria reflects the fact that none of them are fully satisfactory. In fact, in some cases we may be classifying as pathological a physiological adaptation to different load conditions. On the other hand, Kohli et al.<sup>1</sup> report that of 199 patients with LV systolic dysfunction, 23.6% met at least one of three echocardiographic definitions of LVNC, and suggest that the current criteria lack specificity. LV dilatation and the increased sphericity typical of dilated cardiomyopathy can make trabeculae more evident and in this way lead to overdiagnosis of LVNC.

The broad clinical spectrum of disease ranges from asymptomatic patients, with no family history, normal LV ejection fraction, normal ECG, normal exercise capacity and excellent prognosis (in whom it is questionable if they really have LVNC) to the other extreme, of patients presenting with chronic heart failure, thromboembolic events, severe ventricular arrhythmias and sudden cardiac death.

It is clear that an intense research effort is still needed to achieve a better understanding of the pathophysiology of LVNC, perhaps expanding knowledge of the disease to other cardiac chambers, such as the atria. A proper characterization of atrial mechanics could be important in detecting subclinical dysfunction in LVNC and also in differentiating between pathological and physiological conditions. Furthermore, as atrial enlargement and/or dysfunction are recognized outcome predictors in many heart diseases, it would be interesting to assess their prognostic power in LVNC.

In this issue of the *Portuguese Journal of Cardiology*, Nemes et al.<sup>11</sup> studied right atrial (RA) deformation in LVNC patients with three-dimensional speckle tracking echocardiography (3D-STE). Although they identified atrial volume differences between LVNC patients and controls, they were unable to detect significant differences in RA deformation parameters between the two groups, suggesting that RA mechanics is preserved in LVNC.

These results are in strong contrast to a previous study from the same group,<sup>12</sup> also with 3D-STE in LVNC patients but focusing on the left atrium (LA). In that study, the authors found (together with increased LA volumes and decreased LA emptying fractions) a significant reduction in LA deformation parameters in LVNC patients, showing significant deterioration in all the different components of LA function in LVNC.

This work represents one more step towards a better understanding of LVNC, suggesting that RA dysfunction plays a minor role in LVNC and indicating that future investigation in this disease should remain focused mainly on the left heart chambers.

To summarize, this paper is useful to compact knowledge in left ventricular non-compaction. Please, study the left heart in a left ventricular disease!

## Conflicts of interest

The authors have no conflicts of interest to declare.

## References

1. Kohli SK, Pantazis AA, Shah JS, 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.
2. Gati S, Rajani R, Carr-White G, et al. Adult left ventricular noncompaction: reappraisal of current diagnostic imaging modalities. *JACC: Cardiovascular Imaging*. 2014;7:1266–73.
3. Maron BJ, Towbin JA, Thiene G, 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 Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. *Circulation*. 2006;113:1807–16.
4. Elliott P, Andersson B, Arbustini E, et al. Classification of the cardiomyopathies: a position statement from the European Society Of Cardiology Working Group on Myocardial and Pericardial Diseases. *Eur Heart J*. 2008;29:270–6.
5. Jenni R, Oechslin E, Schneider J, et al. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: a step towards classification as a distinct cardiomyopathy. *Heart*. 2001;86:666–71.
6. Chin TK, Perloff JK, Williams RG, et al. Isolated noncompaction of left ventricular myocardium. A study of eight cases. *Circulation*. 1990;82:507–13.
7. 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.
8. Petersen SE, Selvanayagam JB, Wiesmann F, et al. Left ventricular non-compaction: insights from cardiovascular magnetic resonance imaging. *J Am Coll Cardiol*. 2005;46:101–5.
9. Jacquier A, Thuny F, Jop B, 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;31:1098–104.
10. Captur G, Muthurangu V, Cook C, et al. Quantification of left ventricular trabeculae using fractal analysis. *J Cardiovasc Magn Reson*. 2013;15:36.
11. Nemes A, Domsik P, Kalapos A. Right atrial deformation analysis in isolated left ventricular noncompaction – insights from the three-dimensional speckle tracking echocardiographic MAGYAR-Path Study. *Rev Port Cardiol*. 2016;35:515–21.
12. Nemes A, Piros GA, Domsik P, et al. Left atrial volumetric and strain analysis by three-dimensional speckle tracking echocardiography in noncompaction cardiomyopathy – results from the MAGYAR-Path Study. *Hellenic J Cardiol*. 2016;57:23–9.