# What do cardiologists expect from Echocardiography in the evaluation of non-compacted myocardium

***O que o cardiologista espera do Ecocardiograma na avaliação de miocárdio não compactado***

*Luciano Nastari*[1]

[1]*Assistant Physician - Clinical Unit Cardiomyopathies and Aortic Diseases - Instituto do Coração - HCFMUSP (InCor), São Paulo, SP, Brazil.*

Echocardiography, the first-line diagnostic screening method for non-compacted myocardium (NCM), is easy to perform and features low cost and low risk for patients. It is also widely available in clinical practice. However, it is also an operator-dependent method, with limitations including a dependent echocardiographic window, primary focus of visualizing the ventricular apex, and operator difficulty accurately distinguishing between compacted and non-compacted ventricular layers.

Another important limitation of echocardiography is the presence of greater trabeculations among healthy populations of Black people, athletes, and pregnant women, which may overestimate diagnoses. Thus, some authors determined diagnostic criteria for NCM by echocardiography. The currently most accepted criteria were proposed by Chin et al.[1], who analyzed five men with NCM by analyzing the parasternal view, short and apical axes, left ventricular (LV) free wall in maximum diastole, distance from the epicardial surface to the trabecular recess (X), and distance from the epicardial surface to the peak of the trabeculae (Y). The NCM diagnosis is made when $X/Y < 0.5$.

Other criteria were proposed by Jenni et al.[2], who studied 34 patients (25 men) with NCM and analyzed the location of the trabeculae in the apical, medial, and inferior medial LV walls and LV wall thicknesses of the compacted (C) versus non-compacted (NC) regions. Using this classification, the NCM diagnosis is made when $NC/C > 2$ in the short-axis view at the end of systole.

Stöllberger et al.[3] and Kohli et al.[4] analyzed 62 patients (49 men) with NCM and identified defined it as the presence of more than three trabeculations in the LV wall in the apical area of the papillary muscles and visible in a single apical four-chamber plane. On color Doppler, intertrabecular spaces are filled with blood from the ventricular cavity.

Paterick et al.[5] recently proposed the diagnosis of NCM as NCM/compacted myocardium > 2, with parasternal short-axis view measurements taken at end-diastole. This criterion showed a good correlation with cardiac magnetic resonance findings and, according to the authors, provided more accurate NCM and myocardium layer thickness measurements. However, this requires validation, additional confirmation, and comparison with other populations with structural heart disease before it can be accepted as a viable diagnostic option.

Another study[4] compared 199 patients, 143 of whom had an LV ejection fraction (LVEF) < 40%, referred to an accredited heart failure assessment center with 60 healthy volunteers (30 Black). The authors used the three echocardiographic definitions to identify NCM, assessed trabeculation number and size as well as the relative thickness of the non-compacted layer whenever possible. The three echocardiographic definitions showed a poor correlation, with only 29.8% of patients fulfilling all three criteria. Also, 8.3% of the normal controls met the criteria for NCM. Five subjects in the control group (four Black, one White) met at least one diagnostic criterion for NCM. This result emphasizes the limitation of echocardiographic criteria for diagnosing NCM, especially in Black individuals, leading to its overdiagnosis. In that study, if the control group included individuals with heart failure, these results could have been even more discrepant. The study findings suggest that current echocardiographic diagnostic criteria are very sensitive and result in NCM overdiagnosis in patients with LV systolic dysfunction. This seems particularly true in the Black population.

Extensive and detailed studies including normal populations of different races are necessary to determine the upper limits of normal trabecular patterns to avoid unnecessary investigations and treatments in patients and their relatives. Hypertrabeculation can also be a transient phenomenon. A study[6] of 102 primiparous pregnant women undergoing serial echocardiographic tests showed that 26 developed increased trabeculations during pregnancy (8 women met NCM criteria). These patients were followed up for a mean two years, with total trabeculation reductions occurring in 19 women and marked trabeculation reductions occurring in the remaining group. The inability to distinguish noncompaction from hypertrabeculation cardiomyopathy has significant clinical implications.

Despite the increasingly frequent diagnosis of NCM, the echocardiographic criteria are based on studies of limited numbers of patients and different methods. The point in the cardiac cycle at which NCM and compacted myocardium thickness are measured directly influences the relationship between the two evaluated layers. Myocardial thickness peaks in systole and troughs in diastole, directly affecting the relationship between NCM and compacted myocardium. In addition, we must consider the echocardiographic view in which these measurements are taken. Most criteria suggest

## Keywords

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**Mailing Address: Luciano Nastari •**
Av. Dr. Avenida Enéas de Carvalho Aguiar, 44, Cerqueira Cesar, CEP: 05403-000 – São Paulo, SP, Brazil.
E-mail: luciano-nastari@uol.com.br
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that these measurements be taken in the parasternal short-axis view; however, in daily clinical practice, they are often taken in the apical two- and four-chamber views.[7] Finally, some studies showed that a considerable number of young athletes met NCM diagnostic criteria, highlighting the non-specificity of the current diagnostic criteria for highly trained athletes.[8]

Thus, consensus is lacking about which classification is the most efficient for diagnosing NCM. In this situation, patients fulfilling all three diagnostic criteria would be ideal. In current clinical practice, the Jenni et al.[2] criteria are the most widely used, but increased ventricular trabeculation (without the criteria defined above) and echocardiography findings suggestive of possible NCM are also common; thus, the clinician is expected to perform another imaging test to confirm the diagnosis.

Another important factor worth consideration is the underestimation of familial NCM disease since echocardiography is used to screen for it and has a high misdiagnosis rate. The trabeculae are better assessed with echocardiographic contrast and three-dimensional echocardiography. Some recent publications[9–11] with small sample sizes compared NCM patients with healthy and dilated cardiomyopathy subjects. Thus, subclinical myocardial impairment was identified by speckle tracking[10] in patients with a preserved LVEF and NCM versus normal subjects. Other authors identified LV longitudinal strain reductions in the mid and apical regions and normal motility in the basal region, unlike patients with dilated cardiomyopathy, who presented reduced motility in the three regions in apical sections. The authors suggested that longitudinal strain may contribute to the differentiation of cases of dilated cardiomyopathy, therefore reducing NCM overdiagnosis.[9–11]

As for the current guidelines, we have the following recommendations and evidence levels.[12]

**Recommendation level IIb; evidence level B**

- In individuals with suspected NCM, diagnostic criteria on echocardiography or magnetic resonance imaging may be reasonable for establishing the diagnosis when measured as an effective NC/C.
- In individuals with suspected NCM and ventricular arrhythmias, magnetic resonance imaging or other advanced cardiac imaging tests may be able to establish the diagnosis and perform the risk stratification.[12]

## Conflict of interest

The author declares that he has no conflict of interest

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