Reconsidering left ventricular non-compaction: a separate phenotype with genetic heterogeneity?

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COMMENTARY

Excessive and conspicuous trabeculations are associated with deep recesses that communicate with the ventricular cavity but not with the coronary circulation in Left-Ventricular Non-Compaction (LVNC), a cardiac disease. LVNC is hypothesised to be caused by a failure of trabecular regression that occurs during normal embryonic development. Left-ventricular (LV) trabeculations are echocardiographically characterised as structures that move synchronously with ventricular contraction and have similar echogenicity as the myocardium. two and four.

According to clinical investigations, LVNC is frequently familial, with autosomal dominant inheritance being the most common kind. Mutations in ZASP, dystrobrevin, and tafazzin have all been associated to it. six and seven Progressive LV systolic dysfunction can occur throughout life, and it's linked to an increased risk of thromboembolism and ventricular arrhythmia [1].

The diagnosis of non-compaction of the ventricular myocardium (LVNC) has shifted from the autopsy table to a well-known cardiomyopathy. The growing interest in this intriguing cardiac phenotype is reflected in the exponential rise in publications. This raises the question of whether non-invasive diagnostic technology has improved the demarcation of the morphological appearance of the myocardium, or whether LVNC is over diagnosed and the diagnostic criteria are overly sensitive. The purpose of this article is to review developmental considerations, different diagnostic criteria, genetic considerations, outcomes, and therapeutic implications of this cardiomyopathic phenotype with genetic heterogeneity, and to determine whether LVNC is a distinct cardiomyopathy or a distinct myocardial phenotype of various underlying diseases.

Clinical research suggests that LVNC is frequently family, with autosomal dominant inheritance. Mutations in various genes have been related to it, including ZASP, 5 dystrobrevin and tafazzin. The condition can cause progressive LV systolic dysfunction over time and is linked to an increased risk of thromboembolism and ventricular arrhythmia [2].

Is myocardial non-compaction congenital or acquired?

LVNC is thought to indicate impaired/arrested compaction of the developing myocardium based on evolutionary changes during morphogenesis. However, whether LVNC can be acquired is a point of contention. 15 Serial echocardiographic investigations in which LVNC was not diagnosed on the initial echocardiogram but became apparent on subsequent exams have been used to refute the developmental hypothesis.

The weight of evidence suggests that LVNC is caused by a defect in early cardiac morphogenesis or a failure of full maturation of the compacted myocardium, based on developmental alterations

However, recent genetic advances enhance the possibility that LVNC can develop after birth. Dilated and hypertrophic cardiomyopathy have morphological traits that are not present at birth but develop over time. Mutations in sarcomere protein genes are present in patients with dilated cardiomyopathy, hypertrophic cardiomyopathy, and LVNC. The possibility of LVNC developing later in life is raised by this common genetic background.

Left ventricular non-compaction diagnostic criterion

For the diagnosis of LVNC, a variety of echocardiographic definitions have been presented. The first two are based on a research of less than 45 patients with a shared phenotype, while the third is extrapolated from a post-mortem study that looked at the number of conspicuous trabeculations. 4, 9, and 10 are all numbers that can be used to make a number of different combinations. Although all definitions seek to define the condition's morphology, their approaches differ significantly. In distinct echocardiographic views and at different depths of the left ventricle in end-diastole, researchers propose a method for evaluating trabeculation size in relation to compacted wall thickness. Jenni and colleagues 10 and 20 proposed a method for detecting two myocardial layers: compact and non-compact, in short-axis images of the left ventricle in end-systole. In this situation, LVNC is defined by the ratio between the two layers [3].

REFERENCES

1. Freedom RM, Yoo SJ, Perrin D et al. The morphological spectrum of ventricular noncompaction. Cardiol Young. 2005;15(4):345.
2. Ritter M, Oechslin E, Sütsch G et al. Isolated noncompaction of the myocardium in adults.Mayo Clin Proc 1997;72,(1):26-31.
3. Stöllberger C, Finsterer J, Blazek G. Left ventricular hypertrabeculation/noncompaction and association with additional cardiac abnormalities and neuromuscular disorders. Am J Cardiol . 2002;90(8):899-902.

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