Left ventricular non-compaction (LVNC) is historically considered to be a rare form of cardiomyopathy characterized by excessive and prominent trabeculations associated with deep recesses that communicate with the ventricular cavity.2 Given that prominent trabeculations are a normal feature of the intrauterine developing myocardium, LVNC is postulated to be caused by an arrest of trabecular regression that occurs during normal embryonic development.2 Nevertheless, there has been no proof of an arrest in embryonic endomyocardial morphogenesis. Moreover, both cases of de novo acquired LVNC and cases of non-compaction disappearance have been reported in the literature,3,4 calling into question the developmental hypothesis.

The diagnosis of LVNC is based on the identification of a two-layer myocardium, which is thin and compacted adjacent to the epicardium and non-compacted (NC) near the endocardium.5 However, even if frequently overlooked in the past, the inner surface of the ventricles is not smooth and prominent LV trabeculations are present even in normal hearts as shown both at post-mortem inspection6 and by imaging.6 This raises the question of the distinction between normal (or normal variants) and abnormal trabeculations given that, rather than being an all-or-none phenomenon, the extent of myocardial compaction may be a continuous trait within the population with areas of non-compaction occurring even in healthy subjects. The crucial point is to distinguish between normal trabeculations and LVNC.

With regard to echocardiography, three main criteria have been proposed. Chin et al.7 recommend the calculus of a X to Y ratio, where X is the distance from the epicardial surface to the trough of the trabecular recess, and Y is the distance from the epicardial surface to the peak of trabeculae. A diagnosis of LVNC requires an end-diastolic X/Y ratio of up to 0.5. Jenni et al.8 advise to measure the maximal thickness of NC and compacted (C) myocardial layer: in the presence of colour Doppler evidence of deeply perfused inter-trabecular recesses, an end-systolic NC/C ratio ≥ 2 in adults is considered diagnostic for LVNC. Stöllberger and Finnter define LVNC as ≥ 3 trabeculations of the same echogenicity as the myocardium protruding from the LV wall in one imaging plane, located apically (but not connected) to the papillary muscles and surrounded by inter-trabecular spaces perfused from the ventricular cavity. Not surprisingly, this inhomogeneity of definitions creates some confusion. Even worse, the reproducibility of making measurements to diagnose LVNC by accepted criteria is poor4 and the concordance between the three echocardiographic criteria is limited.9 Moreover, up to 8% of healthy individuals fulfil one or more diagnostic criteria for LVNC,9 suggesting that the present criteria could be too sensitive (especially among black population) and lead to overdiagnosis.

More recently, cardiac magnetic resonance (CMR) has emerged as a useful tool in the evaluation of this disease. Its high spatial resolution allows adequate delineation of myocardial wall and trabeculations. A diastolic ratio of NC/C > 2.3 has been proposed for distinguishing pathological LVNC from the degrees of non-compaction observed in healthy, dilated, and hypertrophied hearts.10 Nevertheless, Kawel et al.11 had shown that up to 43% of subjects free of cardiac diseases or hypertension exhibit an NC/C > 2.3 in ≥ 1 region and 6% in > 2 regions, suggesting a low specificity of the NC/C ratio alone. Moreover, the NC/C ratio varies using different imaging planes,11 indicating the need for a standardized measurement. Finally, in a Chinese adult population with dilated cardiomyopathy (DCM) 6 ± 2.5 out of 16 segments were NC on CMR.12 The study by Marchal et al.13 adds an important tessera in our understanding the mutual relationship between trabeculae and LV geometry. The authors studied by CMR 82 patients with DCM; 62% of segments examined were trabeculated and 3% had an NC/C ratio (expressed in the study as segmental trabeculations index) > 2.3. Moreover, the global amount of trabeculation (global trabeculations index, expressed as the ratio of the sum of total trabeculated layer thickness to the sum of total compacted thickness) was positively correlated with the LV sphericity index (i.e. the ratio between LV width and length). These findings suggest that ‘concealed’ trabeculations could become manifest when LV modifies its geometry in heart failure and increases its sphericity. Likewise, this could explain the cases of disappearance of hypertrabeculation in DCM patients after LV geometry improvement.3 Therefore, hypertrabeculation should be evaluated with cautiousness, given that it could represent a normal variant in subjects free of cardiac disease11 and could be magnified in patients with DCM by LV shape remodelling.13 It is unknown if the method proposed by Jacquier et al.14 who measure separately the global and the trabeculated LV mass and diagnose LVNC when the trabeculated mass is > 20% of the global mass, will improve the diagnostic accuracy of CMR.
In summary, universally accepted definition of LVNC is still lacking both on echocardiography and on CMR. Normal variants and LV shape should be take into account when evaluating hypertrabeculation in order to minimize the risk of overdiagnose LVNC, which remains an infrequent myocardial disorder.

References