LETTERS TO THE EDITOR

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Normal left ventricular twist in patients with non-compaction cardiomyopathy, or in normal subjects with hypertrabeculation?

We read with great interest the article by Pacileo et al., recently published in the European Journal of Echocardiography. The authors described normal left ventricular (LV) twist in 14 patients with non-compaction cardiomyopathy (NCCM). This finding is in sharp contrast to severely impaired LV twist described in larger series by us and others. Also, an abnormal LV motion pattern called ‘rigid body rotation’ (systolic basal and apical LV rotation in the same direction, possibly related to the absence or abnormal functioning of the LV fibre helices) was described by us and others. In this respect the unexpectedly high (normal) LV ejection fraction is also in sharp contrast to impaired LV ejection fraction (ranging from 40 to 45%) in NCCM described by us and others.

It should be noted that the diagnosis of NCCM still represents a major dilemma more than two decades after the first proposed diagnostic criteria. It should be well recognized that the criss-crossing meshwork of thin muscle bundles at the LV apical third and thick muscle bundles aligning the myocardial wall are to some extent normal structures. Up to 70% of hearts display at least one prominent LV trabeculation, with two or more present in 36%. In a study by Kohli et al., 8% of normal controls had one or more of the ‘echocardiographic criteria’ for NCCM. Also, subjects with chronic pressure or volume overload such as endurance athletes or patients with valvular heart disease, and blacks are known to have a high incidence of prominent trabeculations.

Therefore, it cannot be excluded that the finding of a normal LV twist in NCCM by Pacileo et al. may be flawed by erroneous inclusion of normal subjects with hypertrabeculation as NCCM patients in their study. Do the authors have additional evidence for NCCM in their patients in terms of genetics, magnetic resonance imaging, “rigid body rotation”, or a severely abnormal non-compaction/compaction ratio?

References

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LETTERS TO THE EDITOR

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Normal left ventricular twist in patients with non-compaction cardiomyopathy, or in normal subjects with hypertrabeculation? Reply

We are grateful to Dr Van Dalen for his letter dealing with the results and the characteristics of our study population. However, an expert reader will certainly recognize that we have carefully followed the proposed inclusion criteria for left ventricular (LV) non-compaction (NC). Indeed, as we reported, all our LVNC patients met the criteria proposed by Jenny et al. (i) absence of coexisting cardiac abnormalities; (ii) NC/C ratio at end-systole >2 and (iii) evidence on colour Doppler of deep perfused intertrabecular recesses.

As reported by us and others, the significant correlation between left ventricular ejection fraction (EF) and twist supports the evidence that twist plays a major role in LV mechanics. As a consequence, in order to compare the ‘twist-pattern’ of cardiomyopathy (CM) patients with preserved EF with that of patients with dilated CM (as a paradigm of CM with impaired EF), we intentionally selected LVNC patients with preserved ejection fraction. As far as we know, impaired pump function is not a criterion for the diagnosis of LVNC. Furthermore, in LVNC patients histopathological abnormalities are mainly confined to subendocardial fibres, whereas ventricular twist, owing to the longer radius, is more significantly governed by the subepicardial fibres. These pathophysiological considerations support the evidence that, in the presence of normal EF, the LV twist pattern is not reduced in LVNC patients. Accordingly, one of the two LVNC patients reported by Udink ten Cate et al. in the presence of preserved LVEF, showed increased LV twist.