Can cardiac magnetic resonance imaging resolve cardiac and neurological enigmas of left ventricular hypertrabeculation/non-compaction? Reply

Est-ce que l’IRM peut résoudre les énigmes cardioneurologiques d’un VG non compacté ? Réponse

We thank Drs Stöllberger and Finsterer for their comments, and take this opportunity to respond to and clarify important issues they have raised.

Some works suggest that current echocardiographic diagnostic criteria are too sensitive, resulting in over-diagnosis of left ventricular hypertrabeculation/non-compaction (LVNC), particularly in black individuals [1]. We agree that it would be interesting to assess the impact of cardiac magnetic resonance (CMR) imaging in this population, but all 16 of the patients in our study were white.

Drs. Stöllberger and Finsterer comment on the lack of neuromuscular disease in our patients. They have published a report demonstrating a high prevalence of neuromuscular disorders in the population with LVNC, and showed that this factor was predictive of death [2]. However, as in another series [1], this comorbidity was not detected in any of our small population despite a systematic clinical neurological examination and muscle enzyme serum dosage.

Six patients in our population had a family history of LVNC; four of the patients were relatives. Among these, we observed the same distribution of hypertrabeculation, which predominated at the lateral and apical levels but with different NC/C ratios. Moreover, they differed in respect to left ventricular function, which was decreased in the older patients. We can speculate that these patients have the same disease, but at different stages. All of the patients were screened for gene mutations, but we do not have the results at this time.

Regarding the frequent complications related to LVNC, one patient in our study had a left ventricular thrombus, but none had an embolic event, and three received oral anticoagulation. Of the 16 patients, 10 were treated for heart failure.

In our study, we observed some discrepancies between echo and CMR for the measurement of left ventricular ejection fraction (LVEF). This was observed especially in patients 1 and 15, in whom echo provided an overestimation. A possible explanation is the poor acoustic window in patient 15 and very large and extensive trabeculations in patient 1, making the measurement of LVEF difficult both by echo and by CMR.

The differentiation between LVNC and papillary muscles, false tendons or aberrant bands is sometimes difficult in the short-axis views, both by echo and CMR. In our work and in clinical practice, we tried to differentiate these structures using atypical echo views and several long-axis CMR slices.

In our study, we used only the maximal NC/C ratio in echo and CMR. We did not report the agreement between the two techniques regarding the segment where this measurement was made. Future studies are needed to answer to this question. However, we recently published a report showing that the percentage of the global trabeculated left ventricular mass measured by magnetic resonance imaging may be a better parameter to evaluate the extension of LVNC [3].

There is a lack of data regarding the correlation between the imaging and the pathological measurements. They could help to define stronger diagnostic criteria even if the difficult problem of the role of load conditions has to be taken into account. A pathological analysis was performed for the heart in three transplanted patients, but we have not yet done a comparison between echo and CMR.

Drs Stöllberger and Finsterer previously reported that LVNC may appear and disappear during a person’s lifetime [4,5]. In our opinion, this suggestion seems somewhat difficult to accept when we consider that LVNC is a congenital cardiomyopathy due to the persistence of the embryonic non-compact layer. However, as suggested by Elliot et al. [6], it is possible that normal left ventricular trabeculations may become more prominent in certain disease states, such as left ventricular hypertrophy, and that, in such circumstances, diagnostic criteria for LVNC may be fulfilled despite the absence of a unique pathology.

In conclusion, we share concerns with all of the issues raised in this letter. The fact that such a famous medical team in the field of LVNC still has questions demonstrates the important uncertainties relating to this “fascinating” disease.

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References


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