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Response to the letter to the editor: unravel the genetic background of noncompaction before relating it with myocardial hypoperfusion

We would like to thank you for the opportunity to respond to the knowledgeable comments and concerns on our study by Prof. Stöllberger and Prof. Finsterer.[1]

There are many different opinions regarding the aetiology of noncompaction cardiomyopathy (NCM), but the authors agree that the development of epicardial coronary arteries is not affected in this type of cardiomyopathy. However, it has been shown that the noncompacted areas have insufficient intramyocardial vascular supply due to failure of microcirculation growth as part of the myocardial compaction process.[2–4] Histopathologic findings of NCM patients in most cases describe interstitial and subendocardial fibrosis, defined also by lack of coronary microvasculature.[5] As coronary artery disease was an exclusion criterion for our study, all patients had performed an either CT angiography or coronary angiography prior to study inclusion.

We understand the concern on our statement, that so far no studies have shown significant improvement by guideline based heart failure therapy. Li *et al*. have indeed studied the effect of beta-adrenergic blockers on possible reverse remodelling in 20 patients with NCM; however, only left ventricular mass reduction has been shown in 13 ± 6 months of follow-up, with no significant change in either left ventricle end-diastolic diameter or ejection fraction. N-terminal pro-B-type natriuretic peptide has been measured only at baseline.[6] Bertini *et al*. has shown that NCM patients with left bundle branch block with QRS duration of more than 150 ms or signs of left ventricular dyssynchrony to show some left ventricular reverse remodelling with resynchronization therapy.[7] We feel, however, that additional studies, especially in the light of newer heart failure therapies (i.e. sacubitril/valsartan), should be performed in order to show potential therapy-based improvement in NCM patients.

All the patients in our study had cMRI performed and fulfilled the accepted criteria for NCM, according to Petersen *et al*.—the end-diastolic ratio between noncompacted and compacted layers was greater than 2.3.[8] Regional distribution of ischaemia detected by SPECT has been reviewed and compared to echocardiography data (regions of noncompacted myocardium and longitudinal strain maps); however, both distributions have not been identical in most patients. Unfortunately, we could not obtain a detailed cMRI analysis with late gadolinium enhancement, which could be insightful to correlate regions of noncompacted myocardium and myocardial ischaemia in greater detail.

We reviewed the ECG data of our study patients and found only six patients with Q waves in ECG. Almost all patients (37/41 patients, all patients with signs of myocardial ischaemia) in both groups had repolarization abnormalities, mainly slight depression of ST segment or inverted T waves. In our study population, a CRT-D device was implanted in only four patients.

Unfortunately, in our study, we did not perform a genetic testing or neurological assessment, as it was not predefined in our protocol. We are thankful for raising this important point; we hope to be able to perform a more detailed study in the future, taking into consideration genetic background and association with neuromuscular disorders, as well as a long term follow-up in our study population.

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