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Reply to letter to the editor: "Pathophysiology of Non-compaction Remains Enigmatic"

LETTER TO THE EDITOR
REPLY

We would like to thank you for the opportunity to reply to the comments and concerns raised in the author's letter to the Editor and to give details about the patient population included in our study.[1] We would also like to thank the author for his interest in our paper and for taking the time to express their concerns.

In this letter to the editor, the author notes potential concerns about not mentioning comprehensive data about the clinical features of the included patients with left ventricular non-compaction (LVNC).

In this study, we retrospectively included patients with LVNC phenotype who fulfilled the morphologic criteria of LVNC and were free of other known cardiovascular or systemic diseases. As the author of the letter mentioned, LVNC can be complicated with heart failure; however, we excluded patients with decreased left ventricular ejection fraction (<50%), and none of them required heart failure treatment or device therapy. Intraventricular and inter-trabecular thrombi and thromboembolic events are considered the most specific complications for LVNC; however, these rare events are associated with reduced left ventricular ejection fraction and/or atrial fibrillation, and LVNC by itself is not a risk factor.[2,3] As none of the patients had atrial fibrillation and their left ventricular function was good, intraventricular thrombi were not visible, and none of them had previous thromboembolic events.

Regarding arrhythmias, 22 patients had previously documented arrhythmias (ventricular extrasystole, supraventricular extrasystole, non-sustained ventricular tachycardia, and atrioventricular reentrant tachycardia) and 6 patients had previous syncope but none of them needed oral antiarrhythmic treatment or had implantable cardioverter defibrillator.

We agree with the author that LVNC can be associated with genetic and neuromuscular disorders (NMD) and that genetic examination should be carried out in patients with LVNC. Genetic background was available in 27 patients and 5 of them had pathology of likely pathologic mutations of the titin or troponin T genes. These patients were fully asymptomatic, but their family history was positive for cardiomyopathy or sudden cardiac death. Two families were involved in our study (brother and sister and 2 sisters), but interestingly, the genetic examination did not reveal any pathologic or likely pathologic mutations. None of the included patients had other known diseases besides LVNC, and thus, neuromuscular disorders were not present and the genetic examination also did not reveal NMD causing mutations. Family history was positive for cardiomyopathies of sudden cardiac death in overall 19 patients. Further genetic examinations and family screening are in progress.

Late gadolinium enhancement is the strongest independent predictor of LVNC complications.[4] Sixty patients received contrast agents and none of them had late gadolinium enhancement.

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To summarize, in this study, we described sex-specific left ventricular characteristics of patients who fulfilled the morphologic criteria of LVNC and had good left ventricular function using CMR aiming to provide further information for the CMR morphologic diagnosis of LVNCszu02. We did not aim to describe the detailed background and clinical features of the patients due to the retrospective nature of our study. Although, we agree that clinical information is necessary for the diagnosis and risk stratification of patients with LVNC. Patient follow-up with the genetic examination, family screening, clinical examination, and cardiac imaging is in progress.

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