Reply to: Consider the genetic and myopathic background, familial occurrence, and alternative definitions of left ventricular hypertrabeculation/noncompaction

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We would like to thank Prof. Stöllberger for her letter.

Regarding the echocardiographic (echo) definition of noncompaction cardiomyopathy (NCCM), we have shown that NCCM does involve the papillary muscles, especially the base of the muscle [1] (Fig. 3A and attached videos 1–4). Using echo, it is routine to visualize the papillary muscle and measure the non-compacted/compacted layer thickness ratio at its distal point.

In addition, our echo observations were confirmed by operative findings of friable papillary muscles documented by Chung et al. [2].

Stacey et al confirmed similar findings to ours of papillary muscle abnormalities by magnetic resonance imaging that predisposes to mitral regurgitation (MR) in patients with NCCM [3].

Regarding the comment on the left ventricle being thin walled, we could not find this description in our manuscript.

In patient number 4 with postoperative arrhythmia, there has been no documented stroke or embolism, and the child was discharged with hypoxic ischemic encephalopathy, then lost to follow up. We attribute ventricular arrhythmias to the NCCM.

We did not conduct genetic testing on this cohort, but none of them had skeletal myopathy. Some patients reported having familial myopathy and NCCM [4].

The controversies and pitfalls in the diagnosis of NCCM were extensively discussed, and there were many diagnostic criteria; however, echocardiography remains the most utilized method.

It has been well documented by us and others [4,5] that in NCCM, ejection fraction (EF) improves with or without treatment and then deteriorates later (relapses and remissions).

This feature is one of the few clinical signs that distinguishes NCCM from other types of cardiomyopathy.

The patients' echocardiographies were reviewed by two authors from Khartoum and Riyadh. Patients with EF below 45% were omitted from the study in order to exclude left ventricle/mitral valve annular dilatation as a cause of secondary MR. Those with EF > 45% are unlikely to have secondary MR, and as we are assuming a new

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mechanism for MR, we needed to exclude other causes of this pathology.

Although we agree with Prof. Stöllberger that this disease needs to be more accurately defined, we have no doubt that it causes distinct echo changes on the mitral valve leading to MR.

References

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