Response to Letter Regarding Article, “Mortality and Sudden Death in Pediatric Left Ventricular Noncompaction in a Tertiary Referral Center”

We thank Drs Stollberger and Finsterer for their letter. They raise many important questions that warrant further discussion and evaluation. Although many of the specific questions remain beyond the scope of the original article, we would like to provide clarification to some of the inquiries solicited.

As suggested in the query, it is well documented that left ventricular noncompaction (LVNC) can be associated with congenital heart disease (CHD) or neuromuscular disorders. We chose to exclude them from our analysis because it has been shown that their prognosis and outcomes are not uniform and indeed worse than those without associated disease. Although the morphology may not be different between isolated and nonisolated LVNC, for the clinician, separation of the entities as such may hold value in the ability to cleanly prognosticate. Indeed, literature has frequently separated the 2 and assessment of isolated noncompaction is commonplace. As suggested, however, analysis of patients with associated CHD and neuromuscular disorders remains important, and that evaluation is ongoing at our institution.

The query also appropriately brings to light the ongoing challenge surrounding criteria for the definitive diagnosis of LVNC. This has historically been based on echocardiographic parameters, although MRI criteria have been suggested of late. As an institution, we have used criteria reported by Jenni et al., which are based on a noncompacted to compacted ratio of the myocardium in excess of 2:1 at end systole. The predominant location of pathology was in the midlateral, midinferior, and apical regions, and all diagnoses were made postuterine (no intrauterine diagnoses included). We fully recognize that there is debate regarding diagnostic criteria, and other centers may use different parameters (California criteria, Vienna criteria, Milwaukee criteria, etc.). From a standpoint of the current study, we thought it most important to be consistent and clearly define the criteria used. Currently, we believe an accurate diagnosis of pathology requires integration of clinical variables (including possible genetic testing) as well as morphological measurements. As such, ongoing study and follow-up of existing cardiac registries will be critical to the understanding of disease.

There are limited data about the clinical management of children with LVNC, particularly regarding activity restrictions. As stated in the article, our practice has been to activity restrict any patient with abnormal cardiac dimensions, cardiac dysfunction, or arrhythmias. We do not actively restrict patients with normal cardiac dimensions and function without evidence of arrhythmias. However, the current dataset is not robust enough to make overarching recommendations regarding activity restrictions at this time.

Overall, we agree that there continues to be a number of unresolved issues concerning LVNC in children and global final conclusions should not be drawn at this time. However, our data suggest that LVNC has a relatively high incidence of cardiac death and is strongly associated with life-threatening arrhythmias in children. As such, continued follow-up and life-long monitoring remain essential. We would welcome a comparative study between pediatric and adult LVNC as suggested.

Disclosures

None.

References

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