Letter by Stöllberger and Finsterer Regarding Article, “Mortality and Sudden Death in Pediatric Left Ventricular Noncompaction in a Tertiary Referral Center”

With interest we read the article by Brescia et al about mortality and sudden death in 242 children with left ventricular hypertrabeculation/noncompaction (LVHT). During a median follow-up period of 4 years cardiac dysfunction, repolarization abnormalities, arrhythmias, and presentation within the first year of life (infantile LVHT) were identified as risk factors for death or transplantation. Whereas some of these risk factors are similar to adult LVHT-patients, several issues should be clarified.

Why were patients with congenital heart disease excluded? LVHT has been frequently reported in association with congenital heart disease, even in the first description of LVHT by Grant in 1926. It has been shown that morphology of LVHT is not different between isolated and nonisolated LVHT. We miss a definition of metabolic syndrome and an explanation why these patients were excluded.

No information is given about the neurological comorbidity of the included patients. It has been shown in adults but also in children that neuromuscular disorders are frequently associated with LVHT, resulting in a worse prognosis than in cases without a neuromuscular disorder.

When reviewing the echocardiograms how did the authors assess a 2-layered structure of the endocardium? Most probably they did measure the myocardium—but at systole or at diastole? Were there interobserver discrepancies in the measurement of layer thickness?

Regarding the ECG abnormalities it would be interesting to know whether there were any cases with broadening of the QRS-complex or atrioventricular conduction disturbances because these abnormalities are frequent in adults with LVHT and may influence prognosis. How many of those who died suddenly had long-QT syndrome?

The incidence of death or transplantation in the 95 cases presenting in the first year of life (infantile LVHT) was 25%. Did the distribution of phenotypes differ between infantile and noninfantile LVHT patients? In how many of the infantile cases did problems during pregnancy or delivery occur? In how many cases was LVHT diagnosed already intrauterinely?

Thirty-one patients died and 13 were transplanted. Did pathoanatomic investigations of the hearts show any coronary abnormalities or endocardial fibrosis? What were the causes of death of the 16 patients who did not die from sudden cardiac death?

In how many patients did the phenotype of LVHT change during follow-up? Did LVHT disappear over time? Though only occasionally reported, disappearance of LVHT can occur.

How many patients developed strokes or embolism during follow-up? How many patients received oral anticoagulants?

Restrictions of physical activity are a further unresolved issue in LVHT. How many of the arrhythmias and sudden cardiac deaths were associated with physical activity?

If LVHT is explained only by interruption of the physiological compaction process, how do the authors explain acquired LVHT, a phenomenon repeatedly reported particularly in neuromuscular disorders?

Overall, there are a number of unsolved issues concerning LVHT in pediatric LVHT patients which need to be solved before making final conclusions. In particular, pediatric LVHT needs to be compared with adult LVHT.

Disclosures

None.

Claudia Stöllberger, MD
Josef Finsterer, MD, PhD
Rudolstiftung
Wien, Austria

References

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