Unsolved issues of left ventricular hypertrabeculation/noncompaction

With interest we read the article by Aras et al. on a 78-year old woman with isolated left ventricular hypertrabeculation/noncompaction (LVHT) but preserved systolic function. The paper raises the following concerns.

What is meant by the term "isolated"? If "isolated" means without any cardiac symptoms or signs or abnormal instrumental investigations, LVHT of the presented patient was not "isolated". Palpitations, exertional dyspnea, ventricular tachycardia, and impaired relaxation pattern are arguments against "isolated" LVHT. The term "isolated" LVHT should be abandoned since LVHT is associated with other cardiac abnormalities in the majority of the cases and since the term does not contribute to the understanding of LVHT.

Carvedilol was obviously given for ventricular tachycardia. It is not mentioned, however, if this therapy was effective. No results of a follow-up Holter are presented. Why was no intra-cardial defibrillator considered? It is well known that LVHT is associated with an increased risk of malignant rhythm abnormalities or even sudden cardiac death.

The authors claim that LVHT is a congenital disorder. Why was no other pathogenetic concept considered? Though congenital occurrence is quite likely in the majority of the cases, there are rare single cases, in which LVHT developed during life.

Apical hypertrophy was diagnosed 5 years before the detection of LVHT. Was LVHT overlooked during this period or did LVHT develop in addition to apical hypertrophy?

The authors cite Oechslin’s study as that with the highest number of LVHT patients. The authors ignore Stöllberger’s study on 77 LVHT patients.

The applied echocardiographic diagnostic criteria for LVHT are only one of at least three different definitions of LVHT. Why did the authors choose this particular definition, and do hypertrabeculations also fulfill the other two criteria for LVHT? Was the ratio noncompacted to compacted myocardial layer measured at end-systole or end-diastole? According to Oechslin’s definition this has to be done at end-systole. According to previous experiences, assessment of the noncompacted layer is difficult at end-systole. Was inter-observer agreement achieved?

Since LVHT also can be visualized on cardiac magnetic resonance imaging, it is desirable to present the results of this investigation, particularly if it confirmed the diagnosis.

Did the morphology of LVHT change during the 2-year follow-up, as has been reported in a patient in whom left ventricular ejection fraction increased and left ventricular volume, end-diastolic pressure, and extension of the noncompaction decreased upon pharmacological therapy?

The oldest LVHT patient is not 83 years but 84 and 94 years, respectively (unpublished data).

LVHT has been described to occur familiarly. Thus, information about the presence or absence of LVHT in the first degree relatives of the described patient is warranted.

We agree that an LVHT prevalence of 0.014% is too low. In our own series we calculated a prevalence of 0.25%.

Why was acetyl-salicylic acid given? Whether LVHT is associated with embolism and requires anticoagulation or not is under debate. In a series of 62 patients the frequency of embolism was not significantly increased as compared to controls.

To our understanding acetyl-salicylic acid is not indicated in the presented patient.

Up to 82% of the patients with LVHT suffer from a neuromuscular disorder. LVHT has been reported in association with dystrophinopathies, myotonic dystrophy 1, dystrobrevinopathy, Pompe’s disease, myo-adenylate deaminase deficiency, mitochondrialopathy, cypher gene mutations, Friedreich ataxia, Barth syndrome, Charcot–Marie–Tooth

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disease, and various other rare genetic disorders.\textsuperscript{2,15–18} Was the history of the patient and her relatives positive for neuromuscular disease? Was the patient and her relatives also neurologically investigated?

Overall, pathogenesis, visualization on cardiac MRI, thromboembolic risk, prevalence, follow-up investigations, familiarity, and neuromuscular involvement remain issues, which are frequently insufficiently addressed when reporting single patients or a group of patients with LVHT.

References


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