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EDITORIAL COMMENT

When and How to Agree in Disagreeing on the Diagnosis of Noncompaction by Echocardiography?*

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Left ventricular hypertrabeculation (LVHT), also known as noncompaction, is a clinical entity that has gained great visibility after its initial description in 1984 (1). Its relevance relates to the fact that it might have significant clinical implications, including malignant arrhythmias and sudden death. One of the most important challenges regarding LVHT is the clear differentiation of this condition from anatomic and structural variants that are present in the normal heart (2). Several investigators have tried to describe criteria that could help in the correct diagnosis. Echocardiography has been extensively used, but no uniformity has been achieved. From the initial studies of Chin et al. (3) and also Jenni et al. (4), several other studies have been published describing echocardiographic criteria. More recently, the introduction of cardiac magnetic resonance (CMR) (5), computed tomography scanning (6), and 3-dimensional echocardiography (7) have added accuracy to the diagnosis, but again, no unanimous criteria has so far been accepted by the scientific community.

One of the problems with the correct diagnosis of LVHT is related to the fact that the overdiagnosis of the condition has been observed in many laboratories (8), because even though the existing echocardiographic and CMR criteria are useful in diagnosing LVHT in patients with clear phenotypic expression, the precise sensitivity and specificity of the criteria are still unknown, particularly in milder forms of LVHT, where the distinction from normal variants can be more difficult. This is quite relevant because it might create significant repercussions to the patient who is misdiagnosed, not only from a psychological standpoint, but even professionally (e.g., athletes). In this regard, the use of CMR has had an increasing role in diagnosing this condition, especially as the availability and experience with this technique increase, and the structural and functional features of left ventricular (LV) noncompaction are better defined. A small CMR study demonstrated that noncompaction is present in 91% of the LV apical segments, and 78% of mid-cavity segments of healthy subjects, and also in athletes and patients with hypertrophic and dilated cardiomyopathies, hypertensive heart disease, and aortic stenosis, but the relative thickness of the noncompacted layer, though measured in diastole, was significantly smaller than in patients with LV noncompaction (9). These findings suggest that LVHT may represent an extreme of a continuous spectrum of the physiological compaction process during embryogenesis. These data need to be confirmed in larger populations, including different age groups and ethnicities, in order to establish more sensitive and specific diagnostic criteria.

Therefore, the need to have a technique that can reliably and reproducibly diagnose the condition is urgent.

SEE PAGE 1252

In this issue of *iJACC*, the study by Stöllberger et al. (10) on intraobserver agreement of LVHT among different experienced observers and laboratories was determined in a group of 51 patients with a previous diagnosis of LVHT and 49 controls. The fact should be highlighted that these observers included the first author and 2 others with more than 20 years' experience specifically with this topic, including several scientific publications on the subject. The authors showed that even the use of pre-defined

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criteria yielded disagreement in 35% of the cases, and even after mutual review, 11% of the cases were still questionable. These results clearly demonstrate the need to have further investigation in this field and expand the use of other imaging modalities in order to reduce the disagreement observed. One of the obvious conclusions from this study is that in case of doubt, the threshold to use other imaging modalities, such as CMR, should be very low. This may help to improve the accuracy of diagnosing correctly LVHT. Although the study involved a small population of patients, and some other methodological aspects could be discussed, the observers were very experienced echocardiographers, and the results can certainly be translated into the real-world practice.

It is also relevant to add that several other characteristics of LVHT still need to be better clarified, namely its natural history, prognosis, and management. Taken into consideration the rarity of this condition, improving our understanding of LVHT will only be possible through the development of multicenter studies, including registries, involving larger populations.

The current study is an important alert to the echocardiographic community to take extra care in the assessment of patients with suspected LVHT because the level of disagreement is still very high. This is particularly relevant at a moment in time where we do not yet have a widely tested and accepted gold standard for the diagnosis of LVHT. In addition, there is a lack of sufficient pathological validation of imaging methods, so that at the present time, there is simply no in vivo gold standard for the diagnosis, and some cases by necessity remain unresolved.

This is certainly another area in cardiology in which the need to organize joint studies is mandatory in order to get enough sample power to make the results of the studies powerful enough to support strong recommendations in the guidelines.

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