No re-appraisal of non-compaction diagnostic criteria without considering neurological co-morbidity and genetic heterogeneity

Methods, Results, and Conclusions of the interesting study by Kohli et al. raise the following concerns.

Irrespective of the diagnostic criteria applied, the prevalence of left ventricular hypertrabeculation/non-compaction (LVHT) is, compared with previous studies, extremely high. The sensitivity of the diagnostic criteria cannot explain this difference. The explanation that ‘the majority of normal controls fulfilling the LVHT criteria were black’ is not justified. LVHT in asymptomatic patients simply confirms that lone LVHT exists.

Which was the indication for echocardiography in the 43 patients without heart failure and the causes of heart failure in the remaining 156?

The statement that ‘no patient had signs of neuromuscular disease’ implies that all patients were seen by a neurologist, which is not mentioned in Methods section. Additionally, it is incredible that among 199 patients, including patients >80 years, none had neurological abnormalities. Since LVHT is frequently associated with neuromuscular disorders (NMDs) and NMDs often present with only subtle manifestations, neurological investigations are a must in LVHT patients.

Although it is currently assumed that LVHT is a congenital abnormality, it may also develop and disappear during life-time. In how many of the included patients was it possible to review previous echocardiographic recordings and to assess if the existence and pattern of LVHT have changed over time? How to interpret the observation that, at least in European whites, the prevalence of LVHT declines with age? Does this mean that patients with LVHT die earlier or that LVHT disappears with age? The authors themselves state that, regarding LVHT as a congenital abnormality, it should be independent of age.

To avoid over-diagnosing LVHT, we recommend to differentiate trabeculations from aberrant bands, false tendons, and abnormally inserting papillary muscles, to use atypical echocardiographic views to delineate these structures, and to consider differential diagnoses, like thrombi, apical hypertrophic cardiomyopathy, fibroma, obliterator processes, intra-myocardial haematomata, cardiac metastases, or intramyocardial abscesses.

The higher prevalence of LVHT with application of Chin’s and Jenni’s criteria than with our criteria may be due to the lack of anatomical landmarks and application of the short-axis view, in which parts of the papillary muscles may be easily mistaken for LVHT, in Chin’s and Jenni’s criteria. That LVHT was more prevalent in African blacks than in European whites is an interesting observation. Though LVHT has been reported in African Americans, there are lacking reports about LVHT in African blacks, possibly due to socio-economic reasons or due to biological or environmental differences between African blacks living in Africa and African blacks living outside. Considering the frequent association of LVHT with NMDs, the different frequency of LVHT in African blacks and European whites could be explained with the different prevalence of at least some of the NMDs in these ethnicities.

We agree with the authors that the current co-existence of different echocardiographic criteria should be overcome by a uniform definition. This also requires elucidation of the genetic heterogeneity of LVHT and of the association of LVHT with extra-cardiac disease, particularly NMD.

References

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Letters to the Editor
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doi:10.1093/eurheartj/ehn036
Online publish-ahead-of-print 12 February 2008

No re-appraisal of non-compaction diagnostic criteria without considering neurological co-morbidity and genetic heterogeneity: reply

We thank Drs Stollberger and Finsterer for their commentary on our paper. They raise a number of points.

First, they correctly observe that the prevalence of left ventricular non-compaction (LVNC) is much higher than that reported in previous series. They state that this cannot be explained by the sensitivity of current criteria, but do not present a counter-argument. Similarly, their statement that ‘the...