

Ventricular non-compaction—a frequently ignored finding?

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This editorial refers to ‘Diagnosis of left ventricular non-compaction in patients with left ventricular systolic dysfunction: time for a reappraisal of diagnostic criteria?’ by S.K. Kohli et al., on page 89

The investigation by Kohli and his colleagues¹ reveals startling findings. In a population of almost 200 patients referred to a dedicated clinic investigating cardiac failure in the UK, almost one-quarter were found to satisfy the currently accepted diagnostic criteria for ventricular non-compaction, or hypertrabeculation to give the finding its alternative name.² Significantly, these criteria were also satisfied by five of 60 control subjects, of whom 30 were black, with four of these showing the features of non-compaction. The authors rightly conclude that the current criteria used for diagnosis of non-compaction are too sensitive, particularly amongst the black population. The alternative conclusion is that non-compaction, or hypertrabeculation, is simply a variant of normal maturation of the ventricular myocardium, with only the most severe forms producing a pathological entity. Both interpretations beg the question as to why the entity is now being recognized with such frequency, having come to clinical attention as recently as 1990.³

It is also the case that examples of what we would now call non-compaction were recognized prior to the important study of Chin et al.³ Dusek and his colleagues,⁴ in 1975, are usually credited with being the first to describe ‘spongy myocardium’, but most of the hearts they described had associated congenital malformations, and not all would now be classed as non-compaction. Anton Becker and I, however, certainly illustrated an example of unequivocal non-compaction in our Atlas ‘Cardiac Pathology’,⁵ although we had no idea at the time that this entity would become known either as non-compaction, or as hypertrabeculation.

Since being brought to attention by Chin and colleagues,³ the condition has generated considerable controversy, with any who wrote suggesting that the entity might represent persistence of the trabecular meshwork seen during embryonic development of the heart being roundly attacked by Stollberger and her colleagues, who insist that the lesion should be considered as

hypertrabeculation,^{2,6} although they now recognize the need to add ‘non-compaction’ to their descriptions, producing as a result the remarkably awkward entity of ‘left ventricular non-compaction/hypertrabeculation’.⁷ To my eyes, as a morphologist with an interest in cardiac development, the similarity between florid examples of non-compaction and the situation seen in the developing heart is striking.⁸ Indeed, my re-introduction to the entity came through a request from Steffen Petersen, a talented young cardiologist working in Oxford, to look at an image produced using magnetic resonance imaging from the relative of a patient known to have hypertrophic cardiomyopathy. The presence of non-compaction was unequivocal, yet this individual was at the time a commercial airline pilot. Determining the clinical significance of such findings in a symptomless person, who had no abnormal findings on testing, is a huge dilemma.⁹ The study of Kohli and his colleagues¹ shows that such problems are going to increase as our techniques for imaging the ventricular myocardium continue to improve. Should the five persons within their control group be concerned that, currently, they fulfil the diagnostic criteria for non-compaction or hypertrabeculation? And what is the significance of the fact that, of these otherwise normal individuals, three fulfil the criteria established both by Chin and colleagues³ and by Jenni and associates,¹⁰ but only two fulfil the criteria for hypertrabeculation established by Stollberger and her co-workers.^{2,6} It is also noteworthy that there were similar discrepancies amongst the patients with heart failure with regard to the diagnostic criteria. Of 47 patients fulfilling one or other sets of rules, almost four-fifths satisfied those established by Chin and colleagues,³ almost two-thirds fulfilling those drawn up by Jenni and associates,¹⁰ and only just over half those set up by Stollberger and her co-workers.^{2,6} Significantly, however, there was overlap of all sets of rules in almost one-third of this population of 47 patients drawn from nearly 200 patients referred for investigations of cardiac failure. Ventricular non-compaction, therefore, or hypertrabeculation for those who prefer this term,⁷ is certainly much more common than previously believed.

The investigation of Kohli and his associates,¹ therefore, shows that there is much work to be done on this fascinating entity.

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There is work for the developmental anatomists, since we still do not know what happens to the trabecular layer seen in the developing heart.⁸ This extensive trabecular meshwork is seen prior to the development of a thickened compact ventricular layer, and prior to the need for coronary arteries to nourish the compact layer. In my opinion, it is most unlikely that the trabeculations themselves become compacted to form the compact layer, so 'non-compaction' could prove to be just as inappropriate as 'hypertrabeculation' to describe the entity. Another important outcome from the study of Kohli and colleagues¹ is their finding that the abnormal layer of the ventricular wall can take the form of a spongy zone, seen in one-fifth, a meshwork, found in almost half, or become evident as hypertrabeculations, as seen in the remaining patients. Thus, we need to know what happens to the trabecular meshwork that is seen during the early stages of development of the ventricular walls. We also need to know whether persistence of the abnormal layer, in its various guises, is present at birth. Kohli and his colleagues¹ found that the frequency of an abnormal layer decreased with age in white patients, but not in blacks—another interesting finding. Perhaps the most important need, nonetheless, is to determine when persistence of the abnormal ventricular layer, in whatever anatomic form, becomes of clinical significance. It is almost certainly the case that many persons have lived a trouble-free life with an extensive trabecular layer, and that it has gone unnoticed. Increasingly now we are beginning to observe undue persistence of the trabecular layer in the setting of congenital cardiac malformations. In some lesions, this is not unexpected, as for example in the so-called 'Swiss-cheese' ventricular septal defect, where the non-compaction involves not only the ventricular apexes, but also the septum. Non-compaction is also now recognized as being an associated finding in patients with left isomerism and those with Ebstein's malformation. The important study of Kohli and colleagues,¹ therefore, raises more questions than it answers. That is no more than we should expect from truly innovative investigations.

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