Letters to the Editor

Cardiologic and neurologic implications of left ventricular hypertrabeculation, also termed noncompaction

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Sir,

We read with interest the article by Freedom et al.¹ concerning the morphological spectrum of ventricular noncompaction. Some issues, addressed in the article, however, deserve comment.

Concerning the nomenclature of the described abnormality, we suggest it would be preferable to use the term “hypertrabeculation”.² As mentioned in the article, neither the etiology nor the pathogenesis of the abnormality is precisely known. There are indications, however, that the condition may be caused by several heterogeneous factors. Patients presenting with this abnormality show both genetic and morphological heterogeneity, and not only does it occur as a congenital malformation, but the feature can also develop or disappear during the life-time of the individual.³–⁵ Hypertrabeculation, being purely a morphologic description, would more precisely reflect the state of our current knowledge about this entity.

Freedom et al.¹ show a typical histological arrangement, characterized by trabeculations covered by extensive fibrous and elastic tissue. This pattern, however, is not always found in patients with this condition. Fat cells within the trabeculations, as well as a fibrous band separating the compacted from the noncompacted myocardial portions, have also been reported.⁶,⁷ It is not presently known if, and how, the pattern of fibrosis differs between compacted and noncompacted myocardium. And, concerning the morphology and location of the abnormality, the question remains to be answered why it is found most frequently in the left ventricular apex, and is extremely rare in the ventricular septum.

That the abnormality results from an embryonic developmental disturbance is but one of several pathogenetic hypotheses. Freedom et al.¹ state that they incline towards this hypothesis, based on observations of a study which looked for noncompaction by cardiac magnetic resonance imaging.⁸ Applying this technique, which is more sensitive than echocardiography, the investigators showed that non-compacted segments are not confined to patients with this abnormality, but can also be detected in the left ventricles of healthy volunteers, athletes, patients with hypertrophic cardiomyopathy, dilated cardiomyopathy, aortic stenosis and hypertensive heart disease.⁹ These interesting findings, however, provide no evidence in our opinion to support any pathogenetic hypothesis. On the other hand, observations of patients who have been shown to acquire this abnormality during their life-time raise the possibility that it may also develop as a compensatory response of a failing myocardium, possibly induced by activation of myocardial precursor cells to attempt renew an adequately functioning myocardium.¹⁰ Premature neglect of alternative pathogenetic hypothesis may impede research about the condition.

Research is further impeded by confusion regarding the criterions used for diagnosis. At least 3 different echocardiographic, and one pathological, definitions are currently available.²,⁷,⁹,¹⁰ Furthermore, cardiac magnetic resonance imaging and computed tomography are applied without anatomically confirmed diagnostic criterions. The confusion is also due to developments in cardiac imaging, since the visualisation of cardiac structures, such as the internal aspect of the left ventricle, previously not detectable non-invasively, raises the

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issue of new definitions for normality as opposed to abnormality.\(^8\)

The abnormality is frequently associated with neuromuscular disorders. The findings of patients with noncompacted myocardium may offer opportunity of detecting by histological and biochemical investigations previously unknown skeletal myopathies which also manifest in the heart. Attempts should be made to investigate the myocardium with methods applied in myology, particularly in cases with neuromuscular disorders of unknown aetiology. There is some evidence that histological abnormalities of the myocardium resemble those of the skeletal muscle in certain neuromuscular disorders, particularly in cases with predominant affection of the heart.\(^1\) Thus, histological evaluation of the myocardium may help to clarify the relation between myopathy and cardiomyopathy.

References


Response to Stöllberger and Finsterer

We thank Professors Stöllberger and Finsterer for their interest in our review. It is with sadness that we must respond without the support of Robert Freedom. As the readers of “Cardiology in the Young” are well aware, the encyclopaedic review of left ventricular non-compaction was one of Robert’s final projects prior to his untimely death.\(^1\) The topics that are raised by Stöllberger and Finsterer, in fact, were all discussed at length in the original review.\(^2\) Could it be that, as in Shakespeare’s Hamlet, “The lady doth protest too much, methinks”?\(^2\) We received a letter couched in similar vein subsequent to the publication of the report concerning the use of magnetic resonance imaging in the diagnosis of non-compaction prepared by Petersen et al.\(^3\) This exchange of letters has now been published.\(^4,5\) We predict, nonetheless, that like Canute, our Viennese colleagues will find it hard to turn back the ever-increasing tide of studies showing non-compaction to be a morphological trait rather than a specific cardiomyopathy, be the cardiomyopathy itself congenital or acquired. But Stöllberger and Finsterer are correct when they stress that the evidence for the trait being congenital has still to be established with certainty, and we fully accept that some patients may develop the manifestation of the trait during life. We wonder, however, whether the evidence is equally compelling for disappearance of the non-compacted layer during life?\(^2\)

It would be non-productive to answer each of their comments in turn, since it remains our belief that our opinions were stated with clarity by Robert Freedom in our initial review.\(^2\) We would emphasise that our opinion that the lesion represented failure of compaction of the embryonic myocardium was based more on examination of developing human hearts than on inferences made from the study of Petersen et al.\(^3\) We accept, nonetheless, that our embryological evidence to date is far from complete. We doubt very much that, despite their protestations, the term “hypertrabeculation” will replace the much more popular alternative of ventricular non-compaction. There is no question in our own minds that the arrangement seen in postnatal life is remarkably reminiscent of the
non-compacted arrangement of the left ventricular wall as illustrated in our review in the early stages of development of the human heart, but we accept fully that further investigation is needed finally to prove the hypothesis that most cases represent a congenital malformation. Disputation remains the lifeblood of science, and it is good that we continue to debate the origins of left ventricular non-compaction, since the one fact on which all agree is that the entity is now recognised with ever-increasing frequency.

Robert H. Anderson, Shi-Joon Yoo, Don Perrin  
Glenn Taylor, Steffen Petersen

References