LETTERS TO THE EDITOR

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Normal systolic function in hypertrophic cardiomyopathy: reality or myth?

We read with great interest the article by Ganame et al.,1 published in the European Heart Journal. By using strain rate imaging technique, the authors found that the myocardial systolic deformation was significantly and inhomogeneously reduced in children with hypertrophic cardiomyopathy (HCM). The main clinical message of the aforementioned study is that in HCM although left ventricular ejection fraction is normal or supranormal, a subclinical myofibrillar systolic dysfunction exists. A crucial matter is whether this systolic dysfunction precedes the development of hypertrophy. In an experimental study published a decade ago by Geisterfer-Lowrance et al.,2 it was clearly demonstrated that in a mouse model of familial HCM, cardiac dysfunction, by means of reduced myofibrillar shortening, was found to precede histopathological changes, i.e. myocyte disarray, hypertrophy, and fibrosis, and notably when ECG was still normal. Therefore, the classic aspect that myocardial systolic function in HCM is normal or hyperdynamic is rather a ‘myth’. On the other hand, alterations in relaxation pattern detected by pulsed-wave Doppler in healthy first-degree relatives of patients with HCM,3 and reduced myocardial contraction and relaxation velocities detected by tissue Doppler imaging in patients genotype positive for familial HCM, but with no hypertrophy and with a normal ECG,4 have been described.

Which one of the above two myocardial functions precedes cannot easily be determined. In our opinion, ‘pure’ systolic or diastolic dysfunction does not exist since the two phenomena of cardiac cycle are closely interdependent and additionally, myocardial relaxation process involves both systole (systolic relaxation) and diastole (diastolic relaxation). In the current era, we think that the traditionally referred disease phenotype, i.e. hypertrophy, should be reconsidered, taking into account the myocardial systolic or diastolic subclinical abnormalities consistent or not with an abnormal ECG, in the absence of hypertrophy.

Another interesting point of the study is that the reduction in systolic myocardial function was related to maximal wall thickness and decreased exercise capacity. It would be very interesting if the authors provided data on how reduced systolic deformation affected patients’ blood pressure response on exercise. The authors also found that the patients who developed ventricular tachycardia had more prominent reduced systolic deformation parameters and higher values of septal E/E’ ratio compared with patients with no ventricular tachycardia. Since this cohort was quite small, multivariate analysis in order to predict the prognostic significance of the above parameters was not performed. In a study by our group, a septal E/E’ > 15 was found to be an independent predictor of adverse outcome in 96 adult patients with HCM.5

In conclusion, in the view of recently developed techniques, such as tissue Doppler and cardiac MRI, the secrets of HCM have become obvious and familiar to the medical community.

References

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