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Editorial

Longitudinal Changes of Left Atrial Volume Index as a Prognosticator in Hypertrophic Cardiomyopathy

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▸ See the article "Role and Clinical Importance of Progressive Changes in Echocardiographic Parameters in Predicting Outcomes in Patients With Hypertrophic Cardiomyopathy" in volume 31 on page 85.

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Hypertrophic cardiomyopathy (HCM) is well known for its heterogenic and dynamic disease entity, in which echocardiography plays a major role in diagnosing and guiding clinical decisions along the course of the disease.[1)2)] There are several known HCM causative genetic mutations, yet phenotypical manifestations vary greatly by patient.[1)] As the 2020 ACC/AHA HCM guidelines suggest, pathophysiological components, such as "dynamic left ventricular (LV) outflow tract obstruction, mitral valve regurgitation, diastolic dysfunction, myocardial ischemia, arrhythmias, and autonomic dysfunction," result in a variety of clinical outcomes through complex interactions of the components.[3)] Therefore, some patients with causative genetic mutations remain stable and may be asymptomatic, requiring no medical intervention. The most severe clinical outcomes of fatal arrhythmia and sudden cardiac death particularly affect young patients. In some subsets of HCM patients, the heart appears to undergo adverse remodeling, resulting in overt dysfunction with significant hemodynamic impairment due to LV wall thinning and cavity enlargement.[1)3)4)] Using mavacamten, a myosin inhibitor recently approved by the FDA for obstructive-type HCM, serial echocardiography must be performed on a regular basis because this drug can decrease myocardial function.

As current guidelines suggest, it is recommended, even for asymptomatic patients, to regularly every one to two years re-evaluate the patient's heart for any changes or newly developed structures.[3)5)] Studies have demonstrated that progressive wall thinning and regressive left ventricular ejection fraction (LVEF) are associated with lethal ventricular arrhythmias and end-stage HCM.[3)] Other studies have suggested different parameters, such as changes in left atrial volume index (LAVI), to be clinically significant.[6)] However, even with the abundance of longitudinal echocardiographic data for HCM patients, there is no consensus on the clinical meaning of changes in echocardiographic predictors (e.g., LVEF, left ventricular outflow tract obstruction, LAVI, E/e′, tricuspid regurgitation velocity, and maximum wall thickness) over a certain period. Thus, the authors of this study attempted to re-evaluate certain echocardiographic predictors and retrospectively scrutinize the changes in patient echocardiograms throughout the course of the disease to determine which echocardiographic parameters are clinically useful as predictors of poor cardiovascular outcomes.

This study retrospectively used data of 162 patients from 2010 to 2017 who underwent follow-up transthoracic echocardiography (TTE) with a minimum one-year interval

without severe aortic stenosis or dual valve replacement. Most of the echocardiographic parameters associated with poor clinical outcomes were related to diastolic function of the left ventricle. As the pathophysiology of HCM involves disorganized thickening of the myocardium through genetic mutations in the sarcomere,[4] impairment of left ventricle relaxation, increased chamber stiffness, and decreased LA systolic function are thought to affect the diastolic function of the heart.[1] Although the current data did not show significant longitudinal changes on follow-up TTE, evaluating the changes in LVEF alone seems to have a limited value in predicting poor cardiovascular outcomes. Instead, the cross-sectional echocardiographic parameters of diastolic function, particularly LAVI from this study, have significant clinical value for predicting adverse cardiovascular events. In other words, changes in echocardiographic parameters over time may have limited clinical value, yet the hemodynamic state, ranging from enlarged LA to progressive pulmonary hypertension, throughout the course of the disease is worth scrutinizing.

HCM as a disease entity is vast, comprising asymptomatic patients with refractory heart failure or sudden cardiac death. This study supports the heterogenic and dynamic nature of HCM and suggests the need for a more individualized approach. As the authors mentioned in the limitations, this is a single-center, retrospective study without morphological subgroup analysis. The pathophysiology of HCM has a genetic component, and further multi-center studies with deep-phenotyping and multi-omics approaches along with regular follow-up echocardiography, including strain and 3-D parameters, may better guide subdivision of patients and further elucidate the natural progression of the disease. Tailoring therapy by individual would improve outcomes from this deadly genetic disease.

This study highlighted the importance of the complex and ever-changing hemodynamic state of HCM patients. Therefore, a prospective, longitudinal, multi-center study with bio-specimens for deep phenotyping could be a step toward precision medicine in HCM patients.

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Conflict of Interest

The authors have no financial conflicts of interest.

Author Contributions

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