



EDITORIAL COMMENT

Should atrial function be routinely assessed in hypertrophic cardiomyopathy?

Deve a função auricular ser avaliada por rotina na miocardiopatia hipertrófica?



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Hypertrophic cardiomyopathy (HCM) remains an important cause of morbidity and mortality in the general population.¹ One of its most challenging features is a well-documented predisposition to atrial fibrillation (AF), which affects ~20% of HCM patients² and contributes to worse clinical outcomes, including heart failure (HF) and stroke.^{2,3}

Several – and not mutually-exclusive – pathophysiological contributors to AF have been described in HCM, including diastolic dysfunction, mitral regurgitation, ischemia and abnormal atrial histology (atrial myopathy). Clinical risk factors associated with AF include female sex, age, LA diameter and hypertension.⁴ An association of incident AF with pathogenic variants in *MYH7* (beta-myosin heavy chain) has been recently described.⁵

A correlation between atrial size and AF incidence is very well known in HCM and other cardiac diseases.² However, the risk of cardioembolic events is higher in HCM patients, which justifies the strong indication to anticoagulate once AF is detected (i.e. the CHADS-VASc score should not be used

to make decisions regarding anticoagulation in HCM).⁶ Due to the challenge of detecting small and infrequent paroxysms of AF, using the usual monitoring methodology of repeat Holters at regular intervals, some advocate starting anti-coagulation when left atrium (LA) dimensions are over a certain value (e.g. 45 mm of anteroposterior diameter) or alternatively perform more frequent (six-monthly) Holters.⁶ Similar to sudden cardiac death (SCD) risk estimation,⁶ LA diameter is the most used parameter of LA dimension for cardioembolic risk estimation. Limitations of this approach include the non-uniform remodeling of the atrium. Therefore, some authors have suggested a move toward using volumetric measures as an imaging biomarker of risk in HCM,⁷ in accordance with contemporary chamber quantification recommendations.⁸

In other clinical conditions, LA function evaluation has been reported to provide additional prognostic information beyond LA size and in predicting progression from paroxysmal to persistent AF.⁹ Normal reference values have been published for atrial strain and volumetrically derived emptying fraction parameters.⁹ These parameters are however not routinely assessed in clinical practice.

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In this edition of the Portuguese Journal of Cardiology, Teixeira et al.¹⁰ report an assessment of left atrial function in a small cohort of 53 patients with HCM, comparing 25 patients with paroxysmal AF (PAF) with 28 sex and aged matched patients without AF. The authors used atrial strain analysis derived from speckle-tracking and volumetric analysis from three-dimensional (3D) echocardiography. The main and interesting findings were the following: a) patients with PAF presented atrial emptying fractions (active and total atrial emptying fractions (AAEF and TAEF, respectively), measured by 3D echocardiography, smaller than individuals in the control group; b) AAEF showed an independent association with PAF, controlled for age, sex, presence of arterial hypertension and body mass index; c) atrial volumes were higher in the PAF cohort, but no difference was seen regarding the anteroposterior diameter of the LA; d) LA strain in all its components was reduced in the overall cohort compared to the published reference values, but with no difference seen between groups.

Similar reports on these parameters of deformation and function were previously published using echocardiography (and more recently magnetic resonance) and have also described a difference for volumetrically derived functional and/or strain parameters regarding incidence of AF in HCM. Some of these studies are cited in the article but others were published more recently.¹¹⁻¹³ Other studies have reported an association of strain with outcomes such as HF, stroke and death.^{14,15}

The strength of the study by Teixeira et al.¹⁰ lies in simultaneously¹⁰ addressing deformation measurements and parameters of LA function derived from 3D-echo volumetric assessment. The main limitations of the current study are its retrospective/cross-sectional nature, which does not allow for direct evidence of AAEF in prospectively predicting AF and the small size of the cohort, which might have prevented there being enough statistical power to detect a difference of strain measurements between cases and controls.

However, the findings are relevant enough to justify the design of well-powered prospective studies with the aim of studying the predictive value of volumetric and strain assessment of the LA in HCM, and also importantly the value of these parameters as predictors of cardioembolic events. With the relative ease of obtaining these parameters from the currently available post-processing software, a significant result from such studies might lead to the incorporation of at least some of these measurements in routine decision-making and contribute to reducing the burden of stroke. Similarly, the use of these parameters should be tested in new models of SCD prediction, given the known association with LA dimension. Finally, it would be interesting to explore genotype-atrial phenotype associations using these measurements, and the potential impact of genotype as a direct cause of atrial myopathy.

Conflicts of interest

The author has no conflicts of interest to declare.

References

- Lorenzini M, Anastasiou Z, O'Mahony C, et al. Mortality among referral patients with hypertrophic cardiomyopathy vs the general European population. *JAMA Cardiol.* 2020;5:73-80.
- Guttmann OP, Rahman MS, O'Mahony C, et al. Atrial fibrillation and thromboembolism in patients with hypertrophic cardiomyopathy: systematic review. *Heart.* 2014;100:465-72.
- Guttmann OP, Pavlou M, O'Mahony C, et al. Prediction of thrombo-embolic risk in patients with hypertrophic cardiomyopathy (HCM Risk-CVA). *Eur J Heart Fail.* 2015;17:837-45.
- Guttmann OP, Pavlou M, O'Mahony C, et al. Predictors of atrial fibrillation in hypertrophic cardiomyopathy. *Heart.* 2017;103:672-8.
- Lee SP, Ashley EA, Homburger J, et al. Incident atrial fibrillation is associated with MYH7 sarcomeric gene variation in hypertrophic cardiomyopathy. *Circ Heart Fail.* 2018;11:e005191.
- Authors/Task Force m, Elliott PM, Anastasakis A, et al. 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy: the task force for the diagnosis and management of hypertrophic cardiomyopathy of the European Society of Cardiology (ESC). *Eur Heart J.* 2014;35:2733-79.
- Hiemstra YL, Debonnaire P, Bootsma M, et al. Global longitudinal strain and left atrial volume index provide incremental prognostic value in patients with hypertrophic cardiomyopathy. *Circ Cardiovasc Imaging.* 2017;10.
- Kou S, Caballero L, Dulgheru R, et al. Echocardiographic reference ranges for normal cardiac chamber size: results from the NORRE study. *Eur Heart J Cardiovasc Imaging.* 2014;15:680-90.
- Haji K, Marwick TH. Clinical utility of echocardiographic strain and strain rate measurements. *Curr Cardiol Rep.* 2021;23:18.
- Teixeira KLM, Correia EB, Tressino CG. Echocardiographic assessment of atrial function in patients with hypertrophic cardiomyopathy with and without paroxysmal atrial fibrillation. *Rev Port Cardiol.* 2022;41:771-9.
- Nakao R, Nagao M, Higuchi S, et al. Relation of left atrial flow, volume, and strain to paroxysmal atrial fibrillation in patients with hypertrophic cardiomyopathy. *Am J Cardiol.* 2021.
- Raman B, Smillie RW, Mahmud M, et al. Incremental value of left atrial booster and reservoir strain in predicting atrial fibrillation in patients with hypertrophic cardiomyopathy: a cardiovascular magnetic resonance study. *J Cardiovasc Magn Reson.* 2021;23:109.
- Tayal B, Malahfji M, Buerger JM, et al. Hemodynamic determinants of left atrial strain in patients with hypertrophic cardiomyopathy: a combined echocardiography and CMR study. *PLoS One.* 2021;16:e0245934.
- Vasquez N, Ostrander BT, Lu DY, et al. Low left atrial strain is associated with adverse outcomes in hypertrophic cardiomyopathy patients. *J Am Soc Echocardiogr.* 2019;32:593-603, e1.
- Fujimoto K, Inoue K, Saito M, et al. Incremental value of left atrial active function measured by speckle tracking echocardiography in patients with hypertrophic cardiomyopathy. *Echocardiography.* 2018;35:1138-48.