Choice of Imaging Methods in Hypertrophic Cardiomyopathy

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You must choose ... But choose wisely!

(A line from the movie Indiana Jones and the Lost Cup)

In January 2019, it will be the 61st anniversary of what was considered the first description of hypertrophic cardiomyopathy (HCM) by the publication of a series of necropsy cases by Dr. Donald Teare in the British Heart Journal in 1958.[1]

However, the first clinical description of HCM was performed by the brilliant Dr. Eugene Braunwald and colleagues – particularly Dr. Andrew Glenn Morrow. These authors publish three manuscripts detailing the clinical, hemodynamic aspects and treatment of this heart disease between 1958 and 1964. This comprehensive study, containing 213 pages with 176 figures and 191 references, details several aspects of HCM recognized until now, which he called idiopathic hypertrophic subaortic stenosis, which we (and he) now refer to as "hypertrophic cardiomyopathy."[2-4]

It should be noted that in the first 25 years of HCM, the diagnosis could only be established by combining physical examinations, electrocardiogram and invasive hemodynamic studies, identifying only the forms of hypertrophy that caused left ventricular flow obstruction.[3] Only at the beginning of the 1970s, the first echocardiogram in HCM was published. It non-invasively described ventricular hypertrophy.[5]

With the technological development of medicine, new diagnostic methods and a variety of imaging modalities has emerged, which can be used to evaluate cardiac structure and function; determine the presence and severity of dynamic obstruction; and identify mitral valve abnormalities and the severity of mitral regurgitation, as well as myocardial ischemia, fibrosis and cardiac metabolism. Moreover, imaging is used to guide treatment, screening, pre-clinical diagnosis and to detect phenocopies.[6]

In the study of cardiomyopathies, Doppler echocardiography, cardiac magnetic resonance imaging (CMRI) and nuclear medicine provided important developments in the study and knowledge of these diseases.

Keywords

Cardiomyopathy,Hypertrophic; Discrete Subaortic Stenosis; Diagnostic Imaging; Electrocardiography/methods; Echocardiography/methods; Mitral Valve Insufficiency; Magnetic Resonance Spectroscopy/methods.

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Given the possibility of so many choices, in the cardiologists' clinical practice, how to choose the imaging methods in HCM?

Without a doubt, Doppler echocardiography is the main complementary method in HCM, as it allows to establish the diagnosis of ventricular hypertrophy, determine the location of hypertrophy (phenotypic forms), identify the presence and degree of left ventricular outflow tract obstruction, provide important information of atrial geometry and mitral valve disorders. It is recommended for the family screening of these patients.[6] Besides, it provides prognostic information via hypertrophy thickness and, presently, via incorporation of new strain, strain rate, speckle tracking and 3D techniques, which may support preclinical diagnosis and differential diagnosis of HCM.[7] The guidelines are consistent in that echocardiography is the method of choice for initial assessment and follow-up of patients with HCM.[8,9]

CMRI includes a range of modalities that provide detailed information on cardiac morphology, ventricular function and myocardial tissue characteristics, as well described in the review published by Fernandes et al.[10] However, its main limitation still lies in the lack of availability to public access, because of the costs, making it impossible to be offered to the whole population. CMRI is fundamental and irreplaceable in cases where hypertrophy is located in regions where echocardiography is less precise, with patients with poor echocardiographic windows or in the differential diagnosis of other cardiomyopathies.[11] In the stratification of sudden cardiac death, it is useful and may also be recommended for the identification of myocardial fibrosis to support the indication of an implantable cardiac defibrillator (ICD). However, it should be noted that, today, there is no consensus that the presence of this finding, alone, would be sufficient for the recommendation of the device.[8,9,12]

Differently, the use of nuclear medicine is more restricted according to the guidelines and, due to its high negative predictive value, it could be requested to rule out coronary artery disease (CAD) in patients of low clinical probability for CAD and with chest discomfort.[8,9] Although previous studies have shown important prognostic information, as well demonstrated by Ely et al.,[9] in the review published by this journal, the evaluation of the presence of microvascular ischemia reserve should not be routinely indicated in the evaluation of these patients.[9]

Authors' contributions

Manuscript writing: Antunes MO, Arteaga-Fernández E, Mady C; Critical revision of the manuscript as for important intellectual content: Arteaga-Fernández E, Mady C.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

Table 1 – Indications of complementary tests in hypertrophic cardiomyopathy (HCM)

Complementary test	Indication
Doppler echocardiography	For all patients at initial and repeated evaluation, when there is a change in symptoms or cardiovascular events In the family investigation of first-degree relatives after 12 years of age. Repeated every 12-18 months Associated with physical exertion in patients with resting gradient in LVOT < 50 mmHg, to identify latent obstruction and invasive treatment planning Microbubble echocardiography during transcoronary percutaneous septal hypertrophy reduction therapy Perioperative transesophageal echocardiography in patients undergoing septal myectomy, to support surgical strategy and to detect surgical complications
Magnetic resonance imaging	In the suspicion of HCM when the echocardiography is non-diagnostic In the investigation and quantification of myocardial fibrosis to assist the indication of cardiodesfibrillator, when stratification by conventional risk factors is inconclusive †
Nuclear medicine	To rule out coronary artery disease in low probability patients due to its high negative predictive value in selected cases It should not be requested as a routine procedure in patients with HCM

**Before this age, to investigate families with premature CSD history, athletes or intense or symptomatic physical training. After 21 years of age, every 5 years, it may be appropriate; † a The extent of myocardial fibrosis is associated with cardiovascular mortality, but, on its own, the current guidelines are conflicting and do not determine the indication of cardioverter defibrillator.*

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Academic Association

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