

Hypertrophic Cardiomyopathy in Argentina: Centers of Excellence in Hypertrophic Cardiomyopathy in Latin America? What is the Meaning of Late Enhancement?

Miocardopatía hipertrófica en la Argentina: ¿Centros de excelencia en miocardopatía hipertrófica en América Latina? ¿Cuál es el significado del realce tardío?

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Since its initial description by Teare, hypertrophic cardiomyopathy (HCM) has fascinated and surprised many by its enigmatic hemodynamics, extremely variable phenotypic expression and sometimes unpredictable clinical manifestations, though the understanding of its natural evolution has been evaluated in several observational studies by different groups worldwide. (1) Today we know that most patients can have an excellent quality of life, a normal life expectancy and treatment with drug therapy, and symptomatic patients in advanced functional class, with left ventricular outflow tract obstruction, benefit from myectomy and alcohol septal ablation, improving functional class and, in some cases, preventing mortality. (2, 3)

The work of Costabel et al. of the Instituto Cardiovascular de Buenos Aires (ICBA) (4) published in this issue of the Journal is unprecedented, since it shows a large series of 259 patients in Latin America with median longitudinal mid-term follow-up of 3.5 years (2-10 years), performed by a group of physicians dedicated to the management of patients with HCM. Most of this experience is consistent with that previously described by different groups, but this work shows a fact that deserves to be rescued. Interestingly, it shows a high prevalence of apical HCM (27%), similar to that described in Japan (5) and not previously described in the Western world, raising the need for the characterization of the natural evolution of this subgroup of patients in Argentina and their genotypic correlation, which might indicate the path to follow with these patients.

Documented as a disease with high risk of peripheral and central embolism, this series shows a total embolism rate of 1.15% (3/259 patients) with two peripheral events and one stroke, slightly less than in some previous studies, showing a stroke rate of 1.4% (0.24% per year), (6, 7) although the ICBA population is relatively young (56 ± 16.8 years), with atrial fibrillation (5.4%), relatively small left atria [38 mm (28-48)] and under anticoagulation therapy. It should

be acknowledged that in previous series central embolisms are the leading cause of death in patients older than 70 years and that it is essential to lower the anticoagulation threshold in patients with large left atria (> 45 mm), paroxysmal or permanent supraventricular arrhythmias, and over 60 years of age. (6) The use of the CHA₂DS₂-VASc score is not recommended to guide anticoagulation therapy in these patients. (8)

The management of symptomatic patients with HCM has progressed in the last six decades due to the efforts focused on establishing the pathophysiology and developing the necessary skills to improve prognosis.

The creation of a center of excellence for the management of patients with HCM in Latin America represents a challenge and the group of ICBA seems to have faced the task, which is encouraging and inspiring for the whole region; however, the study shows some aspects of the long way to go in order to become a center of excellence at the highest level worldwide. Although the results are encouraging and of increasing experience, it has still a very limited number of myectomies (15 cases) and alcohol septal ablations (22 cases), with an ablation: myectomy ratio of 1.5: 1. The main centers of the world show an inverted myectomy: ablation relationship of approximately 10: 1. The results achieved in the main centers of HCM are impressive. (9)

Although surgical myectomy mortality data in ICBA is unknown, the mortality rate in the two main centers of global reference is close to 0%, with an approximate series of 130-140 or more cases per year. (9) This is only possible with continuous work, increasing the number of surgical cases and with the dedication of one or at most two surgeons to treat these patients with pre-established perioperative management protocols. We wish to see the growth of ICBA as HCM management center and know their mid- and long term surgical results.

In this issue of the Argentine Journal of Cardiol-

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ogy, Deviggiano et al. (10) reproduce the information obtained from studies with cardiac magnetic resonance (CMR) in HCM, showing late gadolinium enhancement (LGE) in two thirds of patients, associated with maximum myocardial thickness but not with ventricular mass, and more common in the areas of hypertrophy. This finding confirms that HCM is a fibrosing disease and, above all, located at the basal and mid inferior septum and at the mid-anterior septal level.

Late gadolinium enhancement was observed in 30 (71%) patients with HCM and 141/672 (21%) of the segments evaluated. Its distribution was predominantly intramyocardial (n=103; 73%) and, less frequently, subendocardial (n=24; 17%), epicardial (n=10; 7%) and transmural (n=4; 3%).

Although several studies have promoted LGE as a possible marker for risk of sudden death, its extent is unequivocally associated with cardiovascular mortality, but not with sudden death, (11-13) so European guidelines do not support its use for such purpose. (8)

Late gadolinium enhancement can provide prognostic information and clinical practice guidelines support the use of MRI in all patients with HCM and in first-degree relatives, especially performed in centers with experience in this imaging technique. (8) However, there is still need to close the gap between guideline recommendations and clinical practice with studies that substantiate the value of MRI and its impact on clinical decision-making, upon which many questions still remain unanswered and, more importantly, if these variables are undeniably associated with prognosis and type of outcomes. The answer to these questions will be probably helped by the results of an ongoing registry with more than 2,750 patients from four countries, assessing LGE in this extensive series. (14)

Another missing link in almost all HCM registries is the study of coronary artery disease in the so-called HCM-unrelated mortality. Few prospectively studies have evaluated the impact of coronary artery disease on HCM-unrelated mortality; however, a study from the Mayo Clinic showed poor prognosis in this subgroup of patients. (15) The proper characterization of this subset of cases is a debt with clinical cardiology.

Conflicts of interest

None declared.

(See authors' conflicts of interest forms in the website/Supplementary material).

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