EDITORIAL

The "Real World" of Hypertrophic Cardiomyopathy

El "mundo real" de la miocardiopatía hipertrófica

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With a prevalence of 0.2% or 1:500 people, hypertrophic cardiomyopathy (HCM) is the most common congenital form of heart disease. (1) While the prevalence in the general population is high, many clinicians may perceive it to be relatively uncommon. The apparent disparity between perception in clinical practice and the actual prevalence may be a result of the heterogeneity in the disease resulting in under-diagnosis. The hallmark of HCM is unexplained left ventricular (LV) hypertrophy without dilation of the ventricular cavity. (1) The increased maximum wall thickness (>15mm) may be inappropriately attributed to hypertensive heart disease or "athletes heart". The majority of current data regarding the prevalence and clinical characteristics of HCM comes from large HCM centers in Europe, North America and Asia, with little data from South America. (2). In this issue of the Journal Arias et al. (3) report on the clinical characteristics of HCM in the community setting of Buenos Aires.

The authors retrospectively reviewed admissions to a single hospital over 6 years and identified over 140 patients with echocardiographic or cardiac MRI findings consistent with HCM. The echocardiographic and MRI criteria used were consistent with current recommended guidelines including maximal wall thickness (MWT) ≥ 15mm, resting or dynamic gradient ≥ 30 mmHg or characteristic phenotype without underlying cardiovascular disease. Hypertrophy cardiomyopathy has been identified in all ages (4) but the authors found an older median age of 66 yrs, with a slight predominance in women (52%). Previous epidemiological data suggests a male predominance in the younger population (5); however, recent data (6-7) echoes the results of this study indicating women are often older at the time of diagnosis. In contrast to previously reported data, the majority of patients in this study had a clinical history of hypertension (59%); however, most patients had asymmetric hypertrophy inconsistent with hypertensive heart disease. Since its earliest description (8) HCM has been linked to LV obstruction, and consistent with previous reports (9) the authors found that 2/3 of the patients had dynamic outflow obstruction. As with most contemporary descriptions of HCM, the authors found that the majority of patients had asymmetric septal hypertrophy and a low prevalence of apical HCM.

Traditionally, the etiology of syncope has been attributed to either hemodynamic derangements or arrhythmia. The hemodynamic mechanisms include LVOT obstruction, inappropriate vasodilation and impaired ventricular filling associated with diastolic dysfunction. Arias et al. report a direct correlation between LVMWT and syncope. Left ventricular wall thickness may contribute to diastolic dysfunction and provide greater substrate for ventricular arrhythmias thereby contributing to a higher incidence of syncope.

The authors report a good correlation between MRI and echocardiographic measurements in this population. While a relatively small percentage of patients (22%) underwent cardiac MRI, the demonstration of MRI as a valid tool in this population is of significant importance. As MRI continues to have a larger role in the diagnosis and risk stratification of HCM patients (10) this observation confirms its utility in this region.

Current data demonstrates a correlation between LVOT obstruction and heart failure symptoms. (11) Interestingly, the authors demonstrated a novel association between LVOT obstruction and anginal symptoms, an association not previously reported. However, this finding must be considered under the scope of a relatively high incidence of coronary artery disease described in the study population.

The authors observed higher rates of mortality in this population (2.8%/yr), compared to 0.5%/yr in recent evidence, which was attributed to aggressive treatment with state-of-the-art therapies. (12) However the study confirmed the relatively low mortality associated with HCM and the association with a normal life-span. In this study, 63% of deaths were the result of cardiovascular causes, which is high compared to

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previous reports [13]. Although analysis of the typical "high risk" features associated with HCM, including family history of HCM related sudden cardiac death (SCD), unexplained syncope, abnormal blood pressure response to exercise and frequent episodes of non-sustained ventricular tachycardia (NSVT) were outside the scope of this study, recent data suggests that these risk factors are less prognostic in the population of HCM patients represented in this study (>60 yrs of age). (13) In this population, hospitalization was the only independent predictor of mortality, which may be a result of older age or may represent a unique clinical feature of this population.

The findings by Arias et al. mirror previously reported clinical characteristics of HCM worldwide. Thus, they demonstrate a significant variability in clinical presentation and anatomy of hypertrophic cardiomyopathy. More importantly, this data provides a unique insight into the clinical characteristics of HCM in the community setting of Buenos Aires and confirms the heterogeneous yet ubiquitous presentation of hypertrophic cardiomyopathy regardless of the geographic region studied. Future studies should focus on confirming the traditional risk factors for sudden cardiac death, include younger patients and assess adherence to guideline recommended therapies in this region.

Conflicts of interest

None declared

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