Letters to the Editor

Importance of Sudden Cardiac Death Risk Stratification in Hypertrophic Cardiomyopathy

La importancia de la estratificación de riesgo de muerte súbita en la miocardiopatía hipertrófica

To the Editor,

We read with great interest the work of Sarrias et al,[1] which described the progress of a group of patients with hypertrophic cardiomyopathy (HCM) with implantable cardioverter-defibrillator (ICD) to prevent the risk of sudden cardiac death. We would like to congratulate the authors for producing a current multicenter registry that reflects the difficulty of evaluating ICD implantation in this condition. The authors concluded that ICD implantation as primary prevention (PP) had an extremely low appropriate therapy rate, and that the risk stratification criteria had poor predictive value, and should therefore be revised. However, we believe that these conclusions should be put into context. Firstly, the guidelines available before publication of the article suggested ICD implantation as PP in patients with 2 or more risk factors (ESC/AHA 2003 guideline),[2] and subsequently in those with at least 1 major risk factor (ACC/AHA/ESC 2006 prevention of sudden death[3] and ACC/AHA 2011 HCM).[4] The effectiveness and safety of these recommendations have been evaluated in prospective registries that observed an annual appropriate shock rate of 2.3% to 4% in PP,[5] slightly above that of the registry of Sarrias et al (1.7%).[1] However, those data were based on small series, selected from large centers, and did not study the role of individual risk factors in other circumstances (advanced age or populations from nonspecialized clinics). In addition, we continue to learn about HCM due to new techniques such as genetics, which have helped to classify some patients' ventricular hypertrophy, not as HCM, but as Fabry disease or Danon disease, which have different characteristics, prognoses, and sudden cardiac death risk from HCM.

Secondly, the registry has a mean follow-up of 3.3 years. Other registries have observed appropriate shocks up to 9 to 10 years after implantation,[6] which is why, given the young age of these patients and the long at-risk period, the 2003 HCM guidelines emphasized the need for long-term follow-up in ICD effectiveness analyses. Thirdly, the study had few patients, despite including 3 hospitals (69 patients in total, 70% as PP), which may have influenced the low event rate. The guidelines' recommendations are based on previous registries with larger patient numbers: 334 patients (92% as PP)[6] and 506 patients (76% as PP).[5] We must bear in mind that evaluations based on short follow-up times and small populations may lead to erroneous conclusions.

The article also highlights the high percentage of inappropriate shocks (13%), related mainly to supraventricular arrhythmias in ICDs implanted from 1996 to 2012. While this percentage is lower than that in older registries (27% reported by Maron et al),[5] it is similar to that in recent registries (16% reported by O'Mahony et al).[6] This is probably related to improvement of supraventricular arrhythmia discrimination algorithms and antitachycardia therapies with the latest ICDs, leading to a decrease in the number of therapies.

This year the ESC published guidelines on the diagnosis and management of HCM.[7] They found that previous algorithms based on binary variables did not take into account the effect size of individual risk factors, and so had a modest discriminatory power between high and low risk. Therefore, sudden cardiac death risk assessment using HCM Risk-SCD[8] is recommended. This model divides patients into 3 risk groups according to age, family history of sudden death, syncope, left ventricular outflow tract gradient, maximal left ventricular wall thickness, left atrial diameter, and nonsustained ventricular tachycardia, and offers better discriminatory power than previous algorithms. We hope that this new tool casts some light on the doubts raised by Sarrias et al,[1] as there is still much to learn about HCM.

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