Treatment of Hypertrophic Cardiomyopathy

To the Editor:

In the excellent review of hypertrophic cardiomyopathy by Braunwald et al., the conclusions and recommendations in patient B that severe left ventricular hypertrophy (LVH) (≥3cm) is an absolute indication for an implantable cardioverter-defibrillator (ICD) warrants further comment.

The association of massive LVH and an increased risk of sudden death has been established by recent independent studies that had contradictory conclusions. Spirito et al. evaluated risk factors for sudden death derived from the history and echocardiogram, concluding that severe LVH (≥3cm) was sufficient to recommend an ICD. Elliott et al. performed a similar risk assessment, including markers of hemodynamic and electrical instability. They observed that patients with severe LVH were more likely to have other risk factors and that all patients who died suddenly had at least one of these risk markers. The fact that none of their patients with severe LVH but without other risk factors died suddenly led these authors to conclude that such patients did not require an ICD. In fact, although massive LVH is more frequent in young patients, it is also present in older patients, indicating that it is compatible with a normal survival.

Hypertrophic cardiomyopathy is a heterogeneous disease, and to establish absolute recommendations and guidelines based on just one specific clinical characteristic, independently of how striking it might be, does not seem wise. This is particularly true when the real increase in risk that it confers is not clearly established, when the clinical feature considered is subject to errors in its measurement, and when most of these patients are treated by cardiologists who may not have expertise in cardiomyopathy. In fact, no single disease feature or test is capable of stratifying risk in all patients. Although in the case described by Braunwald et al. the indication for an ICD may be correct, the message from their paper should have been less absolute. In our view, the recommendation for the cardiologist dealing with such a patient should be to use a study protocol that provides a broader characterization of the patient, includes markers of electrical (Holter) and hemodynamic (exercise) stability, and assesses multiple risk markers. Implantation of an ICD is not without complications and may carry important medical and socioeconomic implications, particularly in young patients.

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Response

We thank Dr. Penas-Lado and his colleagues for their comments on our article. Sudden cardiac death (SCD) is the most common cause of death in hypertrophic cardiomyopathy, and fortunately we now have the means, ie, implantation of an implantable cardioverter-defibrillator (ICD), to prevent this. In our brief review, we enumerate the several risk factors that may help to identify patients subject to this dreaded outcome.

The patient in question had three risk factors: (1) a family history of SCD; (2) extreme ventricular hypertrophy; and (3) relative youth. Our recommendation to implant an ICD into patient B was based on the entire clinical picture, not a single finding.

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