Letters to the Editor

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Left ventricular outflow tract obstruction and sudden death in hypertrophic cardiomyopathy

We thank Maron et al. for their editorial on our paper.1,2 Hypertrophic cardiomyopathy (HCM) is a capricious disorder, characterized by heterogeneity at all levels. Together with its relative rarity, this has resulted in a dearth of evidence-based practice. The authors suggest that cardiologists rely on guidance from a few major centers and imply that our paper is out of step with expert consensus. Unfortunately, in their effort to prevent overenthusiastic interpretation of our study, the authors create the impression that there are widely divergent approaches to risk stratification even within the expert community.

It is accepted that the annual sudden death risk for most asymptomatic patients with mild-to-moderate hypertrophy is very low. There is a general agreement that particular clinical features are associated with an increased risk.3 The challenge is the relatively low-positive predictive accuracy of these risk factors. One solution is to model survival as a function of the number of risk markers.4 In our experience, this approach identifies a cohort of patients with an annual sudden death risk of 3% or more. In an abstract quoted by the authors, 13% (not 40% as stated) of patients with one or more than one risk factors and an ICD had appropriate shocks—the annual risk was 4%, very similar to that predicted by our model.5

The authors suggest that we promulgate the idea that only patients with two or more risk factors should be considered for an ICD. In our original paper, we have stated, ‘...Therapeutic recommendations for patients with single risk factors remain speculative, and further work on individual risk factors is necessary to determine if and when prophylactic therapy is indicated.’4 We recognize that there are circumstances in which implantation of an ICD in patients with a solitary risk factor might be appropriate—for example, in the presence of a very malignant family history—but it is important to acknowledge that the decision to implant is not based on robust data.

The observation that resting left ventricular outflow tract obstruction (LVOTO) increases with gradient severity and is higher in the presence of other risk markers. The question raised by this and previous publications is how should we clinically respond to a large resting gradient?

LVOTO differs from most other risk factors in that it is modifiable. In all patients with moderate-to-severe LVOTO, careful evaluation of symptoms and exercise tolerance identifies those individuals in whom invasive gradient reduction is appropriate. When no symptomatic indications for intervention exist, we believe that the current evidence base supports consideration of an ICD in patients with severe LVOTO and other clinically significant risk factors. At present, we do not believe that the data support the use of potentially harmful invasive techniques to reduce gradients in asymptomatic patients.

In summary, we believe that it is more useful to patients to establish the common ground between experts. Surely it is time for physicians with an interest in this disease to work together to design prospective studies.

References

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Left ventricular outflow tract obstruction and sudden death in hypertrophic cardiomyopathy: reply

We have read with interest the accompanying letter from Elliott et al., which is a response to our editorial,1 which itself was commissioned as a response to the Elliott et al.2 paper in the June 5 issue of the Journal. Surprisingly, however, the authors have not even attempted to answer or rebut the substantial clinical points raised in our editorial, which we assumed was the justification for their additional correspondence.

The central issue of this controversy, as we see it, is that Elliott et al. published a paper that could well be misleading to the practising cardiovascular community and lead to widespread application and excessive as well as unnecessary numbers of defibrillators implanted in patients with obstructive hypertrophic cardiomyopathy (HCM). Indeed, the authors’ promotion of left ventricular outflow obstruction as a strong independent risk factor for sudden death could

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