Pitfalls in Clinical Recognition and a Novel Operative Approach for Hypertrophic Cardiomyopathy With Severe Outflow Obstruction Due to Anomalous Papillary Muscle

To the Editor:

The Brief Rapid Communication by Maron et al describing in greater detail a variant of hypertrophic obstructive cardiomyopathy significantly adds to our knowledge of this intriguing disease. The point is very well taken: hypertrophic obstructive cardiomyopathy is not one single entity! Every case has its very own problems, and there are hardly 2 cases alike. In their discussion, the authors raise the important question of whether nonsurgical techniques could possibly deal with this particular variant. Certainly, from their surgical illustrations, it appears as if transluminal techniques could be considered in this situation. All myocardium is supplied by coronary arteries, and if the supply arteries of the myectomy territory can be properly identified, it should be possible to deal with this problem by transluminal myocardial reduction with alcohol.

A number of cases of midventricular obstruction have now been successfully dealt with by nonsurgical myocardial reduction. In the cases presented by Maron et al, it is conceivable that such a procedure would be as effective or almost as effective as surgery. It would be most helpful to see the coronary angiogram performed before surgery in these 2 patients, but it is normally possible to cannulate the supply arteries in these cases. The issue that remains unresolved, however, is how much the papillary muscle itself contributes to the outflow tract gradient, because it is unlikely that this could be modified by nonsurgical techniques.

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Response

We appreciate Dr Sigwart’s interest in our observations regarding the surgical management of obstructive hypertrophic cardiomyopathy (HCM) in those patients for whom the anatomy of the left ventricle and mitral apparatus and the mechanism for the outflow gradient are unusual. The 2 patients in our report had a muscular form of marked subaortic obstruction produced by anomalous (and direct) insertion of a papillary muscle into a portion of anterior mitral leaflet without the interposition of chordae tendineae. However, as we emphasized in our article, although this anomaly can be defined clinically with echocardiography, this may be challenging, particularly when an uninvolved portion of the mitral valve produces substantial systolic anterior motion. Indeed, in our patients, anomalous papillary muscle insertion was initially identified by the operating surgeon (Dr Gordon Danielson) on direct visual inspection through the aortotomy. The novel and extensive muscular resection that followed was tailored precisely to the observed anatomy and ultimately achieved relief of both outflow obstruction and symptoms.

Dr Sigwart is largely responsible for the creation, design, and development of alcohol septal ablation as a treatment for patients with obstructive HCM to reduce the outflow gradient, as a potential alternative to the ventricular septal myotomy-myectomy procedure. We acknowledge these important contributions to the management of HCM. Nevertheless, it should be emphasized that myotomy-myectomy remains the time-honored (for over 40 years) standard treatment for that small but important subset of HCM patients with severe drug-refractory congestive symptoms and marked outflow obstruction at rest. Therefore, at this preliminary juncture, interventions such as alcohol septal ablation (or possibly dual-chamber pacing) should be regarded only as potential alternatives to surgery. Of course, this situation may change in ensuing years when more experience (and longer follow-up) becomes available for these newer treatment modalities.

We believe that the important but complex anomaly of the mitral apparatus in the 2 HCM cases cited in our report, and to which Dr Sigwart refers, may well have been exceedingly difficult to appreciate and manage prospectively in the context of an ablation procedure in the catheterization laboratory. On the other hand, at the time of myotomy-myectomy, it was possible for the operating surgeon to directly and accurately identify (and therefore effectively treat) this uncommon phenotypic expression of HCM. Indeed, it is just such a clinical circumstance that underlines the limitations of applying alcohol septal ablation techniques routinely to those patients with obstructive HCM who would otherwise be candidates for ventricular septal myotomy-myectomy.

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