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Editorial Comment

Surgical Versus Medical Therapy Of Hypertrophic Cardiomyopathy: Is the Perspective Changing?*

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One can hardly think of any disease entity that has been the subject of as much controversy as hypertrophic cardiomyopathy. The core of the problem dates back to the initial pathophysiologic interpretations of the intraventricular gradient with the conflicting concepts of outflow obstruction versus cavity obliteration (1–4). However, either as a consequence of this early and basic dilemma or independently, controversy has also touched on the semantics, diagnosis, genetics and, obviously, management of hypertrophic cardiomyopathy (5–16).

Surgical and medical therapy of hypertrophic cardiomyopathy. The rationale for surgery in hypertrophic cardiomyopathy is based on the early concepts of functional subaortic stenosis or dynamic obstruction impeding ejection out of the involved ventricle (1,2). A chunk of muscle is removed to increase the size of the outflow tract and decrease any significant impediment to flow (12). As the role of the anterior leaflet of the mitral valve in producing the obstruction became clearer (4,17), mitral valve replacement was proposed as the optimal approach to the mechanical correction of the problem (18). This idea was strongly challenged (19) and the indication for mitral valve replacement is currently restricted to a small group of patients in whom mitral regurgitation plays an important role (20). Thus, myotomy-myectomy remains the procedure of choice in the therapy of the obstructive type of hypertrophic cardiomyopathy (16).

The increased emphasis on the functional and sometimes episodic nature of the obstruction, the growing recognition of the nonobstructive forms of hypertrophic cardiomyopathy and the gradual acceptance that intraventricular gradients may be generated by "cavity obliteration" contributed to increased interest in the medical therapy of this entity (8,13,14,21). The enthusiasm for medical therapy was further enhanced by the realization that many of the symptoms of hypertrophic cardiomyopathy are related to abnormalities of diastolic function (22,23) and that arrhythmias were important determinants of prognosis (24,25). Beta-adrenergic blockers, calcium channel antagonists and antiarrhythmic agents are now widely used with proved effectiveness (16). Propranolol, verapamil and disopyramide are, respectively, the most popular agents in each group. Medical management is usually the preferred choice in the vast majority of patients with hypertrophic cardiomyopathy, at least as initial therapy. Surgery is reserved for the few severe and resistant cases that fail to respond or cannot tolerate the available pharmacologic agents.

The present study. In this issue of the Journal, Seiler et al. (26) report the results of a retrospective study of 139 patients diagnosed with hypertrophic cardiomyopathy over a period of approximately 30 years. Their focus is on the natural history of this entity, with attempts to compare the results of medical versus surgical therapy. In the process, the authors present a welcome review of numerous published reports with regard to annual mortality and cumulative survival rates in relation to the various therapeutic options. Their results are generally comparable with these published observations, which they have summarized in their Table 7, confirming that both medical therapy and surgical myotomy-myectomy tend to slightly decrease mortality and modestly improve cumulative survival rates.

The authors clearly state that the study is retrospective and that the groups of patients treated medically or surgically are not really comparable. They conclude, nevertheless, that "cumulative survival rate is significantly better in surgically than medically treated patients." In addition, they report an operative mortality rate of 0% while the generally accepted operative risk is about 5% to 8% (16). Thus, either they have an unusually superior or lucky surgical team or they have selected lower risk patients. Therefore, their results may not be representative and there are grounds for concern about possible bias favoring surgical therapy. In fact, 42 of their patients were sent directly to surgery, a practice contrary to the current trend of sending to surgery only selective patients with severe disease who fail to respond or cannot tolerate medical therapy.

Further, careful review of their data brings attention to the fact that in the medical group, patients receiving verapamil did better than those receiving propranolol, and in the the surgical group, those who received verapamil after operation had a better outcome. Thus, another possible conclusion may be that verapamil rather than surgery may be the therapeutic approach that would best improve survival. However, both such a conclusion and the authors' conclusions regarding surgery are strongly limited by the fact that the groups selected may be even less comparable than the authors state. Only vague information regarding the diagnostic criteria used in the selection of patients is mentioned (27) and no information regarding the presence or

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absence of arrhythmias or other factors that could affect prognosis in the various groups is provided (28). To attain more definitive conclusions, stricter definition of patients is necessary and all factors known to affect management and prognosis should be taken into consideration.

The identity and types of hypertrophic cardiomyopathy. Hypertrophic cardiomyopathy has fascinated, intrigued and confused physicians and investigators for more than 3 decades. It took about 2 decades after its recognition before a relative consensus was reached regarding its current nomenclature (5,6,8,10). In the meantime, so many different names have been proposed that, despite the extensive number of published reports, the amount of true knowledge of this entity was questioned: "the less known about a disease state, the more names are given to it" (6). Questions about the identity of hypertrophic cardiomyopathy and controversies surrounding it continued into the 1980s and persist until the present time (29,30). Difficulties in establishing satisfactory diagnostic criteria have also persisted. Various pathognomonic findings were proposed over the years, but did not withstand the test of time. Quantification of myocardial cellular disarray seems to be an accepted approach to establishing a pathologic diagnosis (16). However, the criteria currently used for clinical diagnosis remain somewhat vague and variable (27).

There is, nevertheless, a general tendency to think of subtypes of hypertrophic cardiomyopathy: obstructive or nonobstructive; asymmetric or concentric; subaortic, midventricular or apical; and hypertrophic cardiomyopathy with or without cavity obliteration (21,27,31–34). Thorough angiographic or echocardiographic imaging, or both, is needed, in addition to the hemodynamic findings for appropriate characterization of subtypes. Overlap is frequent and hypertrophic cardiomyopathy may represent a spectrum of disease rather than a single well defined entity (9). Difficulties encountered in defining and diagnosing hypertrophic cardiomyopathy will invariably affect any therapeutic trial. Although surgery may be clearly indicated in patients with true obstruction who do not respond to the available medical therapy, its role is questionable in patients who have an intraventricular gradient secondary to cavity obliteration. Thus, for definitive comparisons of therapeutic approaches to hypertrophic cardiomyopathy, prospective randomized trials in patients selected on the basis of strict and well defined diagnostic criteria are long overdue.

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