The Role of Operative Treatment in Patients with Idiopathic Hypertrophic Subaortic Stenosis

Over the past dozen years our understanding of the disease spectrum embracing idiopathic hypertrophic subaortic stenosis (IHSS) has advanced considerably. Originally, it was considered that all patients with IHSS had a dynamic form of subaortic stenosis. Subsequently it was recognized that left ventricular outflow obstruction was only one manifestation, and an inconstant one, of a disease that is basically a cardiomyopathy characterized by nonuniform ventricular hypertrophy and a bizarre orientation of myocardial cells.1-5 Interestingly, the cardiomyopathic process nearly always involves the interventricular septum to a disproportionately greater extent than the free wall of the left ventricle5-8; hence, the recently proposed name of asymmetric septal hypertrophy, or ASH.5,9 In some patients the disproportionately hypertrophied septum is associated with severe obstruction of left ventricular outflow under resting basal conditions. In other patients, however, obstruction may appear only after provocative maneuvers, and in still others, no obstruction can be demonstrated under any conditions.

The majority of patients with this type of cardiomyopathy have no symptoms or are only mildly limited, and in such patients no therapy is necessary. Other patients may be severely symptomatic but respond well to the administration of propranolol. Finally, a relatively small proportion of patients, probably under 10%, have severe symptoms in spite of maximal medical therapy. In this Institute an operation designed to abolish outflow obstruction is recommended to such patients if a large pressure gradient is demonstrated within the left ventricle, either at rest or following provocation. The benefit provided by operation, however, ultimately depends on whether or not symptoms are due mainly to outflow obstruction or to the cardiomyopathic process per se. Some investigators hold the latter view, and consider that operation has no role in the therapy of patients with this disease.10 Our experience is at variance with such a belief.

On the basis of recent pathologic, microscopic, and echocardiographic studies performed in collaboration with Dr. Barry Maron, we have identified patients in whom the cardiomyopathic process appears to be distributed throughout the left ventricular myocardium. This wide distribution, however, seems to be characteristic of severely symptomatic patients who do not have obstruction3-7; symptoms in these patients can be attributed largely to decreased contractility and decreased diastolic compliance. In contrast, in severely symptomatic patients who have left ventricular outflow obstruction, the more bizarre cellular manifestations of the cardiomyopathic process are largely confined to the interventricular septum; we believe symptoms in these patients are due mainly to outflow obstruction and the resulting increase in ventricular pressure work.

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This belief is reinforced by the results of the operative experience with IHSS at this Institute which dates from 1960 and now includes 68 patients.11, 12 Preoperatively, all patients had large intraventricular pressure gradients at rest or with provocation, and were in functional classes III or IV (New York Heart Association). The operative procedure employed is transaortic ventriculomyotomy and myectomy. Through an incision in the ascending aorta, the muscle fibers in the hypertrophied septum are doubly incised, split by digital pressure, and a modest amount of tissue between the two myotomies is resected. Operative mortality using this technique has been 9%. Fifty-three of the patients have now been studied by left heart catheterization six or more months postoperatively. No resting gradient was present in 50 patients, and each of the remaining three had gradients of 10 mm Hg. In no patient did a gradient greater than 50 mm Hg appear with isoproterenol administration or the Valsalva maneuver. The left ventricular end-diastolic pressure was lower in most patients in whom it was abnormally elevated preoperatively, and symptomatic improvement occurred in all; of 55 patients followed one year or more, 24 are asymptomatic (Class I), and 31 manifest only mild symptoms (Class II). Comparable results have been reported by other groups utilizing a similar surgical method.13, 14 Of significance, all patients but one have maintained their increased functional capacity during follow-up periods ranging from one to 13 years (mean 4.5 years).

An important question relating to the long-term efficacy of operation is whether the gradient eventually returns. Ten patients have had left heart catheterization in the early (6-12 month) postoperative period and again three to seven years later (mean 5.1 yrs). All ten patients had severe obstruction at rest preoperatively; no resting gradient was recorded in any patient at late study. In addition, it recently has been shown that the intraventricular pressure gradient in patients with IHSS can be estimated very closely by echocardiography.15 Utilizing this technique, an additional 16 patients have been studied two to 11 years after operation (mean 6.5 years), and again, none has evidenced recurrent outflow obstruction.

Thus, in this Institute the operative results demonstrate that septal myotomy and myectomy has been extremely effective in relieving outflow obstruction, that obstruction has not recurred during postoperative follow-up periods up to 11 years, and that symptomatic improvement has been achieved consistently and is long-lasting. That operative elimination of the intraventricular pressure gradient is accompanied by symptomatic improvement indicates that left ventricular outflow obstruction is a major cause of symptoms in such patients. Moreover, the prolonged symptomatic benefit provided by operation also suggests that the cardiomyopathic process in patients with obstruction is either not progressive, or that the rate of progression is very slow. Accordingly, we continue to recommend myectomy to severely symptomatic patients with outflow obstruction in whom propranolol administration is ineffective.

In IHSS, left ventricular outflow obstruction results from abnormal forward motion of the anterior mitral leaflet during systole, an event that results in apposition of the leaflet with the markedly hypertrophied interventricular septum.5, 18-20 Recognizing this mechanism of obstruction, Cooley and others recently have recommended excision and prosthetic replacement of the mitral valve as the operative method of choice.17 The procedure was carried out in nine patients, and in the seven survivors good symptomatic and hemodynamic improvement was reported early after operation (median follow-up period, about six months).

One may question on several counts, however, the desirability of this operation in IHSS. Although obstruction is produced by the abnormal forward movement of the anterior mitral leaflet during systole, the mitral valve itself is intrinsically normal. The primary cause of the abnormal leaflet motion, and thereby the obstruction, would appear to be in some way related to the massively hypertrophied septum. Evidence for this statement includes the observation that septal myectomy abolishes the outflow gradient by restoring normal systolic movement of the mitral valve, a phenomenon demonstrable by echocardiography immediately after resection, as well as much later in the postoperative period.16 Mitral regurgitation frequently is observed in patients with IHSS, and appears to be another consequence of the abnormal leaflet position during systole; myectomy would be expected, and has been demonstrated to abolish the regurgitation.11-13, 20

Thus, mitral valve replacement, although effective therapy, does not attack the primary cause of the obstruction in patients with IHSS. Additionally, it generally is recognized that in patients undergoing mitral valve replacement for rheumatic heart disease, complications related to the prosthesis are
most frequent in those individuals in whom the left ventricular cavity is small. In IHSS the ventricular cavity is slit-like during systole and its shape grossly different than normal. A high incidence of mechanical and embolic problems related to a prosthetic valve, even one of low profile, might be anticipated in such patients. Moreover, patients with prosthetic mitral valves must be permanently anticoagulated, and are thus exposed to the complications and inconvenience of this therapy. Finally, many of the patients operated upon are young, and the fate of prosthetic valves over periods longer than eight to ten years is unknown.

Several other unresolved questions relating to the role of operation in IHSS are of importance, and deserve mention. While we have been gratified by the symptomatic relief afforded by operation in essentially all of the patients surviving operation, it is our current clinical impression that patients who derive most benefit are those whose predominant symptoms preoperatively are angina, presyncope, or syncope; patients whose predominant symptoms are severe fatigue, dyspnea on exertion without angina, or congestive heart failure, although improved by operation, may not be symptom free. These clinical impressions are being subjected to test by exercise studies now in progress, which hopefully will determine whether subgroups of patients can be defined whose responses to operation are different. In addition, operation was initially carried out only in those patients in whom a large gradient was present at rest. However, we recently have extended operative indications to include patients without resting gradients, if large gradients appear following provocative maneuvers. Many of the patients in this subgroup have also benefited from operation, but objective demonstration of the extent of improvement, and the more precise definition of patients who will or will not benefit from the operative procedure, still remain to be determined.

In summary, operative treatment produces symptomatic and hemodynamic benefit in essentially all patients with IHSS and outflow obstruction, and operation is indicated when symptoms are not relieved by nonoperative therapy. The long term results in this Institute indicate that septal myotomy and myectomy is the preferable operative procedure, since 1) outflow obstruction is invariably abolished, 2) recurrence of obstruction has not been observed over prolonged follow-up periods, 3) symptomatic improvement occurs uniformly and is long lasting, and 4) these results are accomplished without exposing the patient to the complications of a prosthetic valve.

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References


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