Operative Treatment in Hypertrophic Subaortic Stenosis

Techniques, and the Results of Pre and Postoperative Assessments in 83 Patients

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SUMMARY

The results of operative treatment in 83 patients with idiopathic hypertrophic subaortic stenosis (IHSS) are described. Most patients with the disease are asymptomatic, or derive satisfactory symptomatic improvement from nonoperative therapy: administration of propranolol, exercise limitation, control of arrhythmia, etc. Operation is required, however, in 10-15% of patients, those who remain severely symptomatic after nonoperative treatment or who become refractory to it. Operation relieves symptoms in IHSS by relieving obstruction to left ventricular outflow, and for a patient to be considered an operative candidate severe obstruction must be documented at left heart catheterization either under resting conditions or after provocative interventions.

All 83 patients were severely incapacitated — 58 in Class III and 24 in Class IV. Seventy had obstruction at rest (average gradient 96 mm Hg), and 13 had only provokable obstruction. At operation the hypertrophic interventricular septum was exposed via an aortotomy, and a vertical bar of muscle was resected between parallel myotomy incisions. There were six operative deaths (7%); no patient has died since 1970. Seven patients have died late after operation, five of them from causes unrelated to their heart disease or the operation.

All surviving patients describe symptomatic improvement. Fifty-two patients with obstruction at rest preoperatively (average gradient 95 mm Hg) have been studied postoperatively: no resting gradient was evident in 47, while in the remaining five the gradient was less than 25 mm Hg. Recurrence of obstruction has never been observed at late catheterization (21 pts) or late echocardiographic examination (37 pts). Obstruction could not be provoked postoperatively in ten of the 11 patients who had large gradients only with the Valsalva maneuver or isoproterenol administration preoperatively. Obstructed and provokable obstructed patients had similar symptomatic improvement after operation. A variety of rhythm and conduction abnormalities were observed both pre and postoperatively, and these are described in detail.

The results of operation in these 83 patients with IHSS demonstrate that gratifying symptomatic and hemodynamic improvement uniformly follows left ventriculomyotomy and myectomy. Relief of obstruction and amelioration of symptoms have proved to be long-lasting during postoperative observation periods extending to 14 years. Continued application of the operative procedure in properly selected patients appears to be indicated.

Additional Indexing Words:
Cardiomyopathy
Aortic stenosis
Obstructive cardiomyopathy
Diffuse subvalvular aortic stenosis
Asymmetric septal hypertrophy (ASH)
Muscular subaortic stenosis

Over the past fifteen years the study and treatment of patients with idiopathic hypertrophic subaortic stenosis (IHSS) has been one of the principal areas of interest to the physicians and surgeons of the National Heart and Lung Institute.

During this period the diagnosis of IHSS has been established in several hundred patients, and communications in this and other journals have described the results of studies characterizing the clinical, hemodynamic, angiographic, echocardiographic, pathologic, and genetic manifestations of the disease. Additionally, since 1960 operative treatment has been carried out in certain patients with IHSS, and the results of operation in the first 25 patients were presented in this journal in 1968. The operative experience with IHSS at the Institute has continued and, as of March 31, 1974, 83 patients had been
RESULTS OF OPERATION IN IHSS

operated upon. The present report summarizes the pertinent preoperative clinical and hemodynamic findings in these 83 patients and the results of continuing postoperative evaluations over intervals of 1–14 years are presented. The operative methods utilized to relieve obstruction in IHSS have been modified, and the techniques in current use are illustrated and described in detail.

General Considerations of Management in IHSS Patients

Relatively few patients with IHSS require operative treatment, and to place this mode of therapy in proper perspective, the general plans of management utilized in patients with IHSS are outlined.

It is important to note that IHSS constitutes a broad clinical and pathophysiologic spectrum, that with rare exception has asymmetric septal hypertrophy (ASH) as its characteristic feature. This spectrum embraces three general groups of patients: 1) those who have obstruction to left ventricular outflow under resting conditions (classical IHSS); 2) patients who have obstruction to left ventricular outflow only after provocative interventions; and 3) patients who have no obstruction either at rest or after provocative maneuvers. The clinical features and natural history of the third subgroup of patients, those without demonstrable outflow obstruction at any time, are just now being elucidated. Since much is still to be learned about this subgroup, and since operative treatment is not utilized in the treatment of these patients, they will not be considered further in the present communication.

A small minority of the patients with classic IHSS are totally asymptomatic,56,61 and these patients are usually identified when a precordial murmur, an abnormal ECG, or an abnormal roentgenogram of the heart is observed on a routine examination. Other asymptomatic patients may be detected when the families of known patients are examined and screened by echocardiography. In asymptomatic patients cardiac catheterization is not ordinarily indicated, and no therapy is instituted. Patients are examined periodically, however, and young patients are advised to refrain from unusually strenuous exercise and competitive athletics.

Most patients with IHSS come to medical attention because they are symptomatic. When the clinical manifestations of IHSS are observed in a symptomatic patient, it is usually desirable to substantiate the diagnosis and to determine the presence and severity of left ventricular outflow obstruction. This can usually be accomplished by means of echocardiography, and under ordinary circumstances left heart catheterization and selective angiography are not carried out at this stage of management. Treatment consists of the administration of propranolol, initially 50–160 mg daily, moderate restriction of physical activity, and weight reduction in obese patients. If symptoms suggest the occurrence of paroxysmal atrial fibrillation quinidine is indicated, and if this arrhythmia is documented, warfarin should also be administered because of the frequency with which systemic embolization occurs in patients with IHSS who develop atrial fibrillation.37 The frequency of syncope can be reduced in many patients by instructing them as to the mechanisms responsible for it and the circumstances under which it is likely to occur, e.g. with sudden assumption of the erect posture, after a large meal, during a paroxysm of coughing, while straining at stool, etc. With these general measures, modified as necessary, most patients will experience definite improvement, and may be managed successfully for prolonged periods at an acceptable balance of physical activity and symptomatology.

A relatively small proportion of patients with IHSS, approximately 10–15%, remain severely symptomatic after institution of the measures outlined above, or become refractory to treatment after an initial period of improvement. In such patients, operative treatment should be considered, unless some specific contraindication such as a serious intercurrent disease or extremely advanced age exists. The benefits provided by operation result from the relief of obstruction to left ventricular outflow, and the presence of severe obstruction must be documented by cardiac catheterization before the final decision as to operation can be made. Obviously, the study should relate to the patient’s current symptomatic state, and data obtained at a catheterization many months or years previously are not applicable. In most symptomatic patients a large systolic pressure gradient between the left ventricle and the aorta or a systemic artery is present when the patient is at rest under basal conditions. In other patients no gradient is evident under these conditions, but a large gradient appears with the application of one of the interventions known to provoke or intensify obstruction: execution of the Valsalva maneuver, administration of isoproterenol or amyl nitrite, etc. At the present time operation is recommended to symptomatic patients of both groups. Occasionally a patient is encountered who has severe obstruction at rest, but few or no symptoms attributable to heart disease. Currently, we do not believe that such patients should be operated upon, since the course of the disease in these circumstances is not well defined, and there is presently no evidence that “prophylactic” relief of obstruction will prolong survival. This concept of management is, of course, at direct variance with that applicable to children and young adults with the discrete forms of aortic stenosis.

Circulation, Volume 52, July 1975
Thus, the patient with IHSS who is an operative candidate: 1) is severely symptomatic, 2) is not significantly improved by optimal nonoperative treatment, 3) has severe obstruction to left ventricular outflow at rest or with provocative interventions, and 4) has no serious intercurrent disease.

The Patients Operated Upon

At the time of operation the 83 patients ranged in age from ten to 67 years (mean 41 years); 46 were men or boys and 37 were women or girls. In the text the patients are referred to by numbers indicating the chronologic order in which they were operated upon. The first operation was carried out in January 1960.

All patients described symptoms related to their heart disease, and the prevalence of specific symptoms is detailed in Table 1. One patient was in New York Heart Association functional Class II, 58 were in Class III, and 24 in Class IV. Each patient presented the physical findings characteristic of IHSS. Seventy-one patients had stable normal sinus rhythm at the time of operation, three were in stable atrial fibrillation, while nine were in normal sinus rhythm but had paroxysmal atrial fibrillation on numerous occasions. Electrocardiographic evidence of left ventricular hypertrophy was present in all patients. Conduction abnormalities of several types were noted preoperatively in 29 patients. Four patients had complete left bundle branch block (LBBB) preoperatively; one of these had had an unsuccessful operation performed previously at another hospital. One patient had complete right bundle branch block (RBBB), and another had left posterior fascicular block (LPFB). One patient demonstrated RBBB with alternating LPFB and left anterior fascicular block (LAFB). Twenty-two patients had electrocardiographic findings consistent with pure LAFB. In seven of these 22 patients the mean QRS axis was more leftward than −45°.

Cardiac Catheterization

This study was carried out preoperatively in all patients on one or more occasions. In a given patient the presence and magnitude of the systolic pressure gradient within the left ventricle was often different at sequential studies. Therefore, the data utilized in subsequent analyses are those obtained at the catheterization which most immediately preceded operation. In 70 patients studied under resting basal conditions a large systolic pressure gradient, ranging from 40 to 210 mm Hg (average 96 mm Hg), was recorded between the left ventricle and the aorta or a systemic artery; these patients are designated as obstructed. In 13 patients no gradient was present at rest in nine, while small gradients, 20 to 26 mm Hg, were recorded in the remaining four. In all these 13 patients, however, a large pressure gradient became evident with the application of one or more provocative interventions such as execution of the Valsalva maneuver or the administration of isoproterenol or amyl nitrate. These patients are designated as provocable obstructed. (Isoproterenol was usually administered intravenously in a dose sufficient to raise the heart rate 20–25%. When amyl nitrate was given the patient inhaled the vapors from one ampule of the drug.)

The left ventricular end-diastolic pressure was abnormally elevated (> 12 mm Hg) in 57 of the 83 patients; the abnormal values ranged from 15 to 45 mm Hg and averaged 23 mm Hg. The cardiac index at rest was normal (> 2.5 L/min/m²) in 56 of the 78 patients in whom this measurement was obtained; in all 78 patients the average index was 2.8 L/min/m². The arterial pressure was directly recorded before and after premature ventricular contractions in 81 patients preoperatively; the pulse pressure was not augmented in postextrasystolic beats in 79 of them (abnormal PVC response).

Pulmonary arterial pressure was measured preoperatively in 80 patients. The systolic pressure was abnormally elevated (> 40 mm Hg) in 27 patients; pulmonary hypertension was severe in seven of these 27 patients, and in them systolic pressures of 95, 80, 75, 68, 65, 63, and 60 mm Hg were recorded. Systolic gradients greater than 10 mm Hg were recorded within the right ventricular outflow tract in six patients; the gradients were 60, 35, 28, 21, 20 and 11 mm Hg.

Operative Methods

The operative method presently utilized to relieve outflow obstruction in IHSS is similar in principle to the technique of left ventriculotomy and myectomy described in 1964. Continuing experience with the operation, however, has resulted in the gradual adoption of an improved and probably more efficacious technique which is described.

If the patient is receiving cardioactive drugs, their administration is usually discontinued 48 to 72 hours before operation. In an occasional patient withdrawal of propranolol will result in severe exacerbation of precordial pain and/or tachycardia. In this event the drug is continued in lowest effective doses until 12 hours before operation. Similarly, continued digoxin administration may be required to control the rate of ventricular response in patients with atrial fibrillation.

Anesthesia is usually induced with morphine and maintained with halothane and a muscle relaxant. Halothane is preferred because its negative inotropic effect is known to lessen the severity of the outflow obstruction in this disease. Hypotension is an uncommon sequel of anesthesia, but if it occurs intermittent or continuous infusion of methoxamine or phenylephrine is indicated.

The patient is positioned supine, and a complete median sternotomy is made. The ascending aorta is freed to the origin of the innominate artery. A single venous cannula is passed into the right atrium; femoral arterial cannulation is usually carried out except in older patients. Cardiopulmonary bypass is instituted, after which the heart is elevated and a left ventricular drainage cannula inserted through an apical stab wound. The patient is then cooled to an esophageal temperature of 30°C, and the ascending aorta is occluded. A vertical aortotomy is made and extended obliquely to the aortic annulus in the noncoronary sinus of Valsalva (fig. 1). The normal aortic valve is retracted, and

Table 1

Preoperative Symptoms Described by All 83 Patients Undergoing Operation for Idiopathic Hypertrophic Subaortic Stenosis

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Percent of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dyspnea with effort</td>
<td>94%</td>
</tr>
<tr>
<td>Angina pectoris</td>
<td>81%</td>
</tr>
<tr>
<td>Syncope</td>
<td>71%</td>
</tr>
<tr>
<td>Orthopnoes or PND</td>
<td>59%</td>
</tr>
<tr>
<td>Paroxysmal arrhythmia</td>
<td>11%</td>
</tr>
<tr>
<td>Angina &amp; syncope &amp; failure</td>
<td>31%</td>
</tr>
</tbody>
</table>

Circulation, Volume 52, July 1975
RESULTS OF OPERATION IN IHSS

Figure 1

Operative exposure of the interventricular septum in preparation for left ventriculotomy and myectomy. After median sternotomy, bypass is instituted, general body hypothermia induced (30°C), and the aorta is opened vertically. The bulging hypertrophied septum is visible below the right coronary valve leaflet. A ridge of thickened white endocardium is always evident on the most prominent part of the septum, the site at which it is opposed by the anterior mitral leaflet during systole.

the ridge or bulge of hypertrophic muscle in the interventricular septum is visible below the base of the right coronary valve leaflet. Posteriorly, the anterior mitral leaflet is seen, and is often obviously thickened and opaque. The right coronary leaflet is then retracted with a special cloth-covered retractor, and, after the heart has arrested and become flaccid, much of the septum can be rotated anteriorly and into the operative field. On the most prominent aspect of the hypertrophied septum the endocardium is always seen to be thickened and heaped up into a transverse ridge of fibrous tissue (fig. 1); this endocardial scarring is the site at which the septum is contacted by the mitral leaflet during systole.

From an assortment of flat ribbon retractors, one is selected which will pass freely but snugly through the aortic annulus; in the usual adult a retractor 24 or 26 mm in width is appropriate. The retractor is passed through the aorta to the apex of the heart, displacing and protecting the anterior mitral leaflet and papillary muscles behind it (fig. 2). The tip of a #10 knife blade, attached to an angled handle, is then passed into the septum just below the base of the right coronary leaflet at a point 2–3 mm to the right of the commissure between the left and right coronary leaflets. The blade is inserted through the septum toward the apex for a distance of about 4 cm, and is then withdrawn as its cutting edge incises the septum with a sawing motion directed toward the ventricular lumen and the retractor (fig. 2). A second myotomy is made in the same manner, parallel to the first one and about 1 cm to the right of it. Both incisions are then palpated; at the most prominent part of the septum the incisions should be about 1.5 cm in depth, and, if necessary, the muscle fibers are split by digital pressure to achieve this depth. With a conventional knife a transverse incision is then made at the base of the right coronary leaflet connecting the proximal portions of the myotomies (fig. 3). A traction suture is placed in the end of the bar of muscle which has been isolated, and the bar can be freed from the surrounding septum for a variable distance by incision of its attachment under direct vision. This is usually not possible deep within the ventricle, however, and the suture is passed through the opening of a rectangular knife, the blade of which is 1 cm in length (fig. 4). As traction is made on the suture, the rectangular knife is pushed toward the apex, freeing the remainder of the muscle bar from the septum (fig. 5). The bar of muscle can usually be excised intact, but sometimes breaks or fragments. In this case the resection is completed with the special angled rongeur shown in figure 5.* An alternate method of resection, not utilizing the rectangular knife, is useful. The two vertical myotomies are made and connected transversely as described above. The muscle bar is then grasped with the angled rongeur and the instrument is pushed firmly toward the apex, peeling the

*Circulation, Volume 52, July 1975

The special retractors, knives, and the angled rongeur utilized in the operation are available from Codman & Shurtleff, Inc., Randolph, Mass. 02366.
After the heart has arrested and become flaccid, the right coronary leaflet is retracted with a cloth-covered retractor and the mitral leaflet and papillary muscles are protected with a flat ribbon retractor passed through the annulus to the apex. The first myotomy is made with an angled-handle knife just to the right of the commissure between the left and right coronary leaflets. The blade is inserted into the septum, in the long axis of the ventricle, for a distance of 4 cm and is withdrawn with a sawing motion directed toward the ventricular lumen and the retractor.

Figure 3

A second myotomy is made about 1 cm to the right (clockwise) of the first. The incisions are then deepened, if necessary, by digital splitting of the muscle fibers; the myotomies are usually 12–15 mm in depth at the most prominent aspect of the septum. A transverse incision is then made at the base of the valve leaflet connecting the proximal portions of the two myotomies.
RESULTS OF OPERATION IN IHSS

in two early patients (3 and 9), operated upon at a time when the relations of the conduction system to the interventricular septum were not fully appreciated and, inexplicably, in a recent patient, 67; all three patients have implanted pacemakers. Small interventricular septal defects were created at operation in two patients (9 and 55); in each the resulting left-to-right shunt is small, Qp/Qs 1.4 and 1.3, respectively. In three other adult patients the operation was uneventful, but interventricular septal defects were discovered when studies were carried out six months postoperatively. Each of these three patients had an illness characterized by chest pain, fever, and congestive heart failure 4–6 weeks after returning home, apparently at the time the defect occurred. It is postulated that in these patients myectomy caused infarction and subsequent late rupture of the septum, possibly because the resection compromised an

Figure 4

The bar of muscle isolated between the incisions is held by a traction suture (as shown) or a suitable clamp. The muscle is freed with the rectangular knife (designed by Dr. Edward B. Stinson) or with a special angled rongeur.

Figure 5

As traction is made on the muscle bar, the rectangular knife is pushed toward the apex, freeing the muscle bar from its anterior attachments to the septum. The apical portion of the resection is often more easily accomplished with the rongeur, which may be introduced via the aorta or, alternatively, via the apical stab wound. In the latter case the rongeur is positioned and directed by the left index finger passed through the valve ring.
MORROW ET AL.

atypical or diseased septal blood supply. In two of the three patients the left-to-right shunts are small, and no treatment has been necessary; in patient 35, in whom Qp/Qs was 2.4, congestive failure gradually developed, and the defect was uneventfully closed at a second operation carried out 3½ years after the initial one.

A postoperative complication led to late death in patient 59. His operation and early postoperative period seemed uneventful, but on the fourth postoperative day he suddenly became anuric. Abdominal aortography showed no filling of either renal artery, and the right renal artery was explored. It was found to be filled with thrombus, but neither gross nor microscopic examination indicated whether the material was embolic or whether it had formed in situ. Blood flow was restored to this kidney, but its function proved inadequate and, after 18 months of hemodialysis, renal transplantation was carried out. The patient ultimately died of infection related to immunosuppressive therapy. Autopsy disclosed no evidence of previous embolization to any organ. Thus, the question as to whether the renal arteries were selectively embolized or whether they thrombosed spontaneously cannot be resolved.

Six other patients have died late, eight months to 13 years after operation, and necropsy was carried out in five of them. Patient 1 was asymptomatic and working daily as a farmer. He died suddenly and unexpectedly, apparently of arrhythmia, 13 years after operation. Patient 26 died five years postoperatively; he

Table 2
Principal Causes of Hospital and Late Death in 13 Patients Undergoing Operation for Idiopathic Hypertrophic Subaortic Stenosis. Necropsy was Performed in 12 of the 13 Patients

<table>
<thead>
<tr>
<th>Pl. no./NYHA class*</th>
<th>Time postop</th>
<th>Cause</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hospital deaths</strong></td>
<td>Days</td>
<td></td>
</tr>
<tr>
<td>8/III</td>
<td>7</td>
<td>Arrhythmia</td>
</tr>
<tr>
<td>31/III</td>
<td>68</td>
<td>Anesthetic accident — pacemaker implantation</td>
</tr>
<tr>
<td>32/III</td>
<td>0</td>
<td>LV failure — low CO</td>
</tr>
<tr>
<td>39/IV</td>
<td>2</td>
<td>Low CO, after MV replacement</td>
</tr>
<tr>
<td>46/IV</td>
<td>0</td>
<td>Laceration of LV</td>
</tr>
<tr>
<td>49/IV</td>
<td>8</td>
<td>LV failure — low CO</td>
</tr>
<tr>
<td><strong>Late deaths</strong></td>
<td>Months</td>
<td></td>
</tr>
<tr>
<td>1/I</td>
<td>159</td>
<td>Arrhythmia</td>
</tr>
<tr>
<td>17/I</td>
<td>35</td>
<td>Suicide</td>
</tr>
<tr>
<td>18/II</td>
<td>60</td>
<td>Auto accident</td>
</tr>
<tr>
<td>22/III</td>
<td>8</td>
<td>Stroke</td>
</tr>
<tr>
<td>34/I</td>
<td>37</td>
<td>Acute MI, severe coronary atherosclerosis</td>
</tr>
<tr>
<td>39/II</td>
<td>20</td>
<td>Infection following renal transplantation</td>
</tr>
</tbody>
</table>

*Functional class immediately prior to death.

Abbreviations: CO = cardiac output; MI = myocardial infarction; MV = mitral valve.

Figure 6
Semidiagrammatic representation of the opened left ventricle illustrating the area and extent of septal resection and its relationships to the aortic valve leaflets and the adjacent membranous septum and conduction tissue.
RESULTS OF OPERATION IN IHSS

dveloped progressive right heart failure with massive edema and ascites, and was also a chronic alcoholic. At autopsy the left ventricle was found to be thin-walled and dilated, suggesting that alcoholic cardiomyopathy may have been primarily responsible for his terminal illness and death. In the four remaining patients death was unrelated to cardiomyopathy or the operation (table 2).

Distinct symptomatic improvement has been described by all patients. Fifty-nine presently surviving patients have now been followed for at least one year, and up to 14 years; the mean duration of follow-up in the entire group is 5.7 years. The functional classifications (NYHA) of these patients preoperatively and at the time of most recent examination are shown graphically in figure 7. The prevalence of specific symptoms before and after operation will be described in detail below, when the results of operation in patients with and without obstruction at rest are considered. In brief, however, improvement or abolition of dyspnea, angina, and left heart failure have occurred, and no patient has had syncope after operation. The single patient in class III is patient 9 who has complete heart block, a pacemaker, and operative ventricular septal defect. Her left-to-right shunt is small (1.4:1) but she has developed severe pulmonary hypertension (85/30 mm Hg). She is grossly obese and has also been shown to be a paranoid schizophrenic; no further operative treatment is contemplated.

Postoperative Hemodynamic Assessments

Cardiac catheterization has been performed postoperatively, on one or more occasions, in 63 patients. The initial postoperative study has usually been made at 6–9 months after operation, and in 21 patients later assessments have been made as well. In patients who have had more than one postoperative catheterization, the data presented are those obtained most recently. Also included are data obtained in six patients who subsequently died. The hemodynamic findings in patients with and without severe obstruction at rest preoperatively are noted separately.

Patients with Obstruction at Rest Preoperatively

The peak systolic pressure gradients between the left ventricle and a systemic artery recorded under basal conditions pre and postoperatively in the 52 obstructed patients who have been studied postoperatively are plotted in figure 8. Preoperatively, the gradients ranged from 40 to 175 mm Hg (average 95 mm Hg); postoperatively no resting gradient was evident in 47 patients, and in the other five the gradients were 25 mm Hg or less. The effects of the Valsalva maneuver were studied postoperatively in 46 obstructed patients (fig. 9). Under resting conditions 41 of the 46 patients had no gradient, and in 30 no gradient could be provoked by the maneuver. In the

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**Figure 7**

Functional classifications (New York Heart Association) of 59 surviving patients preoperatively and postoperatively. Only patients observed at least one year postoperatively are included.

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**Figure 8**

Systolic pressure gradients between the left ventricle and a systemic artery measured pre and postoperatively under resting basal conditions in 52 patients who had severe obstruction at rest preoperatively. The preoperative value is that measured immediately prior to operation, and the postoperative one is that recorded at most recent catheterization.
remaining 16 patients a gradient appeared, or an existing gradient was intensified by the Valsalva maneuver, but in only four patients did it exceed 50 mm Hg. Isoproterenol was administered to 32 patients who had had obstruction at rest preoperatively, and the results of this provocative intervention are also illustrated in figure 9. After operation no gradient could be provoked in 15 of the 32 patients, while in 17 patients a gradient appeared or an existing one was intensified by the drug. In only four patients did the gradient exceed 50 mm Hg. In nine patients a residual gradient at rest was found to be present at the first postoperative study (fig. 10). All nine were subsequently restudied, and in seven of the nine patients the residual obstruction evident early postoperatively was shown to have resolved completely at later study. Another 12 patients did not have intraventricular pressure gradients at the first postoperative study, but were also studied later; no gradient was ever evident at these subsequent studies. To elucidate further the long-term efficacy of operation, echocardiography was utilized to estimate the left ventricular outflow gradient47 in a total of 37 patients who were studied six months to 12 years after operation (mean 6.2 years); none showed recurrent outflow obstruction.

Pre and postoperative determinations of the left ventricular end-diastolic pressure (LVEDP) were made in 52 patients obstructed before operation. The pressure fell in 31 of the 34 patients in whom it was abnormally elevated (> 12 mm Hg) preoperatively, and became normal in 13 patients. The LVEDP increased in eight patients from a normal to an abnormal value, and in three patients LVEDP was abnormal preoperatively and did not change postoperatively.

No systematic change in cardiac index was observed following operation, and index was usually normal both before and after operation.

The pulmonary arterial (PA) pressure was measured pre- and postoperatively in 41 of the obstructed patients, and 14 of these had a systemic PA pressure which was abnormally elevated (> 40 mm Hg). Postoperatively, all 14 demonstrated a reduction in pressure, and in nine the PA pressure fell to normal. As noted above, patient 9 has developed progressive pulmonary hypertension apparently due to the ventricular septal defect created at operation.

Patients without Severe Obstruction at Rest Preoperatively

Eleven of the 13 patients who demonstrated severe obstruction only after provocative interventions preoperatively have been studied postoperatively. No resting gradient was present in any patient. The effects of the Valsalva maneuver pre- and postoperatively in all 11 patients are shown in figure 11. Preoperatively, eight of the 11 patients had no gradient at rest, and in the other three the gradients were 20, 20, and 26 mm Hg. The Valsalva maneuver provoked a large gradient (average 59 mm Hg) in every patient preoperatively. Postoperatively, no
RESULTS OF OPERATION IN IHSS

Figure 11

Systolic pressure gradients between the left ventricle and a systemic artery recorded pre and postoperatively in 11 patients who had only provocable severe obstruction preoperatively. The effects of the Valsalva maneuver pre and postoperatively are illustrated. Preoperatively the maneuver provoked a large gradient in all 11 patients (center panel) while only one patient evidenced any gradient postoperatively (right panel).

gradient accompanied the maneuver in ten of the 11 patients. Isoproterenol was administered to seven of the 11 patients preoperatively, and a large gradient (average 88 mm Hg) was produced in each (fig. 12). All 11 patients were tested in this fashion postoperatively, and isoproterenol produced no gradient in ten of the 11 patients.

Comparisons of the Operative Result in Patients with and Without Obstruction at Rest

Prior to 1967, operation was only recommended to patients with IHSS when a large gradient within the left ventricle was demonstrated under basal conditions. In that year, however, patient 23 was referred, in whom both symptoms and findings were those of severe mitral regurgitation, and the intraventricular gradient was only 26 mm Hg. At operation, palpation of the mitral valve revealed severe regurgitation, but immediately after myotomy and myectomy the valve was completely competent, and the patient derived excellent symptomatic and hemodynamic benefit. Since that time 12 additional patients who developed severe obstruction only with provocation have been operated upon. There have been no previous reports of the results of operation in such patients, and for this reason an analysis of their courses in comparison to those of the larger obstructed group seems indicated.

Of the 13 patients there were 11 men and two women aged 17 to 59 years (mean 38 years); there were 35 men and 35 women in the group with obstruction at rest, ranging in age from 10 to 67 years (mean 43 years). The symptoms described preoperatively by the 13 patients with provocable obstruction were similar to those of the 70 obstructed patients. The prevalence of dyspnea, angina, syncope, and orthopnea were essentially identical in the two groups, but paroxysmal arrhythmia and the triad of angina, syncope, and failure tended to be more frequent among patients with only provocable obstruction. There was no difference in the distribution of patients in functional Classes III and IV between the two groups, and the preoperative and postoperative hemodynamic findings have been summarized above.

All of the 13 patients with only provocable obstruction are living, and 11 of them have been followed for one year or more. The symptomatic status of these 11 patients is compared to that of the 48 obstructed patients followed one year or more in table 3. It is apparent that excellent symptomatic improvement resulted in both groups of patients, and that the extent and quality of improvement did not differ between obstructed and provocable obstructed ones.

Electrocardiographic Changes as a Result of Operation

The site of septal hypertrophy in IHSS is closely related anatomically to the area of the normal conduction system. This probably accounts for the variety of conduction abnormalities noted preoperatively in these patients, as described above. It also has been recognized that a myotomy and myectomy into the hypertrophic muscle mass may lead to conduction ab-

Figure 12

Systolic pressure gradients between the left ventricle and a systemic artery recorded pre and postoperatively in 11 patients who had only provocable severe obstruction preoperatively. The effects of intravenous isoproterenol administration were determined preoperatively in seven of the 11 patients and all developed very large gradients (center panel). Postoperatively the drug was given to all 11 patients and only a small gradient was provoked in one patient (right panel).

Circulation, Volume 52, July 1975
normalities. To delineate the specific conduction changes, an analysis of the postoperative electrocardiogram recorded immediately prior to discharge (2-4 weeks after operation) and at most recent follow-up examination was made for each patient. The results of this analysis are illustrated in figure 13.

Abnormal conduction was noted postoperatively in 75 of the 77 surviving patients. The most common abnormality was LBBB, which was found in 54 patients (70%), of whom four had had LBBB preoperatively. Left anterior fascicular block clearly developed in eight patients, who had an abrupt leftward shift in mean QRS axis at the time of operation, together with the other characteristics of LAFB. Five patients with a LAFB pattern preoperatively continued to demonstrate it postoperatively. One patient developed LPFB after operation. Four patients demonstrated RBBB and LAFB postoperatively; two of these patients had RBBB preoperatively. The patient with RBBB and LAFB alternating with LPFB had permanent atrial and ventricular pacing wires implanted at the time of operation, though he had stable RBBB and LAFB since. As mentioned above, three patients developed complete heart block at the time of operation and all have required permanent pacing.

Comparison of electrocardiograms at discharge and at latest follow-up (mean 5 years), has shown significant changes in conduction in two patients. One patient (23) had LBBB preoperatively and had the same abnormality postoperatively for 70 months, at which time she developed Mobitz type II heart block, which subsequently led to complete heart block. She was given a permanent transvenous pacemaker, and has continued to demonstrate this block as well as periods of sinus rhythm with LBBB since that time. A second patient (20) had LAFB preoperatively, and developed complete LBBB at operation. Eighty-eight months later he also developed Mobitz type II heart block, leading to complete heart block, and underwent placement of a permanent transvenous pacemaker.

A comparison of conduction abnormalities between the obstructed and the provable obstructed groups, both pre and postoperatively, did not reveal any consistent relationships. In the preoperative description of symptoms, the presence of paroxysmal arrhythmias (usually atrial fibrillation) was found in 11% of the obstructed and 27% of provable groups (table 3). This difference is significant at the $P < 0.05$ level. At late follow-up the prevalence of paroxysmal arrhythmia was similar (table 3). Among all 59 surviving patients, three had stable atrial fibrillation preoperatively and each has remained in this rhythm postoperatively; three additional patients are in this rhythm at late follow-up; five of the six patients in atrial fibrillation postoperatively had obstruction at rest preoperatively.

### Echocardiographic Assessments

Echocardiographic estimation of the severity of left ventricular outflow obstruction has been made in 16 patients both preoperatively and six months postoperatively. In 14 patients, the preoperative obstruction index indicated obstruction under basal conditions. At catheterization all 14 patients were found to be in the obstructed group, and left ventricular outflow pressure gradients were recorded under basal conditions. The other two patients had obstruction indices that did not indicate a basal gradient, but both had abnormal anterior systolic motion of the mitral leaflet. In our experience, this mitral valve motion would suggest that no left ventricular outflow pressure gradient would be measured.

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### Table 3

**Comparison of Symptoms Described Pre and Postoperatively by 48 Patients with Severe Obstruction at Rest and 11 Patients Who Had Only Provocable Severe Obstruction. Only Patients Surviving Operation and Followed at Least One Year Are Listed**

<table>
<thead>
<tr>
<th></th>
<th>Severe obstruction at rest (48 pts)</th>
<th>Provocable obstruction only (11 pts)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean follow-up (yr)</td>
<td>Preop. (%)</td>
</tr>
<tr>
<td>Dyspnea with effort</td>
<td>94</td>
<td>34</td>
</tr>
<tr>
<td>Angina pectoris</td>
<td>81</td>
<td>18</td>
</tr>
<tr>
<td>Syncope</td>
<td>71</td>
<td>0</td>
</tr>
<tr>
<td>Orthopnea or PND</td>
<td>59</td>
<td>4</td>
</tr>
<tr>
<td>Paroxysmal arrhythmia</td>
<td>11</td>
<td>10</td>
</tr>
<tr>
<td>Angina &amp; syncope &amp; failure</td>
<td>29</td>
<td>0</td>
</tr>
</tbody>
</table>

---

### Figure 13

Conduction abnormalities present pre and postoperatively in all 83 patients operated upon. The postoperative evaluation is based on each patient's most recent ECG (LBBB = complete left bundle branch block; LAFB = left anterior fascicular block; LPFB = left posterior fascicular block; RBBB = complete right bundle branch block; CHB = complete heart block). The number of each block indicates the number of patients with that pattern of conduction.
RESULTS OF OPERATION IN IHSS

under basal conditions at catheterization, but that a large gradient would occur with provocative maneuvers. This was the case with both patients.

Persistent abnormal anterior systolic motion of the mitral leaflet was noted six months postoperatively in every patient, but in 15 of the 16 patients the extent of abnormal systolic motion was reduced compared to the preoperative state. In six patients, intraoperative echocardiograms demonstrated that the change in the systolic motion of the mitral leaflet was present within minutes after myotomy-myectomy. The absolute extent of anterior mitral leaflet excursion increased postoperatively in one patient; however, because the outflow tract was markedly widened postoperatively, the anterior mitral leaflet did not approximate the septum, and obstruction did not occur. The mechanism by which abnormal anterior systolic motion of the mitral leaflets is reduced by left ventriculomyotomy and myectomy is presently under investigation. The working hypothesis is that operation increases the cross-sectional area of the left ventricular outflow tract, thereby decreasing the velocity of blood being ejected, and thus reducing the Venturi forces acting on the anterior mitral leaflet.66 However, we also have noted some paradoxical motion of the ventricular septum in several patients, which may contribute to systolic widening of the outflow tract. Whether this alteration in septal motion is an important factor in the operative relief of outflow obstruction remains to be determined.

Echocardiography was also used in an attempt to determine whether the hypertrophy of the left ventricular free wall that is present in patients with IHSS can be reversed by operative relief of obstruction.60 Posterobasal left ventricular free wall thickness was measured in 16 patients in whom left ventricular outflow obstruction, as determined by catheterization, had been abolished by operations carried out at least two years previously (range 2.5 to 12.3 yr, average 6.5 yr). These measurements of free wall thickness were compared to measurements made in 13 similar patients who had been operated upon within the previous year. Comparison of the preoperative hemodynamic data in the two groups revealed no significant differences in cardiac index, left ventricular end-diastolic pressure, left ventricular outflow pressure gradient, or cardiac symptoms. The patients in both groups had been catheterized six months postoperatively, and in every instance the preoperative basal gradient had been abolished by operation. Echocardiographic study revealed that the postoperative free wall thickness in the patients operated upon several years previously was significantly less than the postoperative wall thickness in those who had undergone myectomy only recently.

These data suggested that free wall hypertrophy in patients with obstruction to left ventricular outflow could be reversed at least partially by operation, and are consistent with the hypothesis that free wall thickening in this group of patients is secondary to the left ventricular outflow obstruction. These conclusions can be regarded as only tentative, however, until changes in free wall thickness can be studied in the same patients pre and postoperatively. To date, echocardiograms suitable for measurement of wall thickness have been obtained in 13 patients preoperatively and again six months postoperatively. In two of the 13 the posterobasal free wall of the left ventricle had decreased in thickness by 2 mm or more. In two of the 11 patients in whom free wall thickness was unchanged at six months, echocardiograms were also recorded at 18 to 24 months. In one patient significant (> 2 mm) thinning of the posterobasal free wall was demonstrated, while in the other the wall thickness remained unchanged at this time.

IHSS and Mitral Regurgitation

It is recognized that more or less severe mitral regurgitation is evident angiographically in many patients with IHSS, both those with obstruction at rest as well as those with only provokable obstruction. An attempt was made to assess objectively the presence and severity of mitral regurgitation before and after operation in the group of patients described in this report. The effort proved fruitless. Many of the selective left ventricular contrast studies were biplane angiograms, and in the majority, both those made at the Clinical Center and those submitted by referring physicians, the X-ray exposures were not recorded with a simultaneous electrocardiogram. Thus, when left atrial opacification was observed on the films it could not be determined whether it was the result of premature ventricular contractions. In the relatively small number of patients in whom both pre and postoperative cineangiograms were available, the interpretation of one study or the other was almost invariably negated by the occurrence of ventricular arrhythmia. It can thus only be stated that, on the basis of physical findings and clinical evaluations, no patient has significant mitral regurgitation after myotomy and myectomy.

In this general context it is probably necessary to broach once more, and perhaps finally, the subject of mitral valve replacement in patients with IHSS. Our reasons for insisting that valve replacement should not be utilized as the primary mode of operative therapy have been presented and discussed in some detail previously.52, 54, 63 Mitral valve replacement has, however, been carried out in two patients in this series. In patient 39, palpably severe mitral regurgita-
tion persisted after myotomy and myectomy, and a possibly congenitally deformed mitral valve was excised and replaced with a discoid prosthesis. The patient died after a course characterized by progressively low cardiac output. In patient 76 numerous ruptured chordae tendineae to the anterior mitral leaflet were observed when the mitral valve was visualized via the aortotomy. After myotomy and myectomy the valve was replaced with a stented porcine heterograft; the patient is living and has symmetric and hemodynamic improvement comparable to that of the remaining patients.

In summary, the results of operation reported herein prove that effective relief of outflow obstruction in a patient with IHSS can be achieved without subjecting the patient to the inherent dangers attendant upon the presence of a prosthetic valve. Rarely, here in two of 83 patients, a patient with IHSS may also have mitral regurgitation because of an anatomically diseased mitral valve; in this case valve replacement with a low profile or central flow prosthesis is obviously necessary.

Comment

It is now recognized that IHSS is not a clinical or physiologic entity, but a form of cardiomyopathy principally characterized by asymmetric septal hypertrophy, symmetric hypertrophy, and idiopathic hypertrophic subaortic stenosis. Classic IHSS (i.e., obstruction to left ventricular outflow present at rest) occurs considerably less frequently than the nonobstructive form of the disease. The management of the nonobstructed symptomatic patient constitutes a difficult therapeutic dilemma. The results of operation presented in this study, however, indicate that gratifying clinical improvement uniformly follows successful left ventriculomyotomy and myectomy in severely symptomatic patients who have large gradients present at rest, or in whom large gradients develop in response to provocative interventions. The efficacy of myotomy-myectomy is also further supported by the demonstration that outflow obstruction has not recurred in any patient during postoperative observation periods up to 14 years. The symptomatic improvement provided by operation appears to be similarly long-lasting. Whether a role exists for prophylactic operation in patients with obstruction to left ventricular outflow, but without severe symptoms, cannot be determined until more information is available concerning the natural history of the disease.

Addendum

Since this report was prepared and submitted for publication, additional patients with the obstructed forms of IHSS have been studied and operated upon. As of this date (April 1975) a total of 102 patients have had left ventriculomyotomy and myectomy. There have been no additional operative deaths. Two additional patients have died late. Patient 35, in whom a ventricular septal defect had been closed, died with progressive congestive heart failure. Patient 24 died after experiencing an attack of severe dyspnea seven and one-half years after operation. Necropsies were not carried out in either patient.

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