Operative Treatment in Idiopathic Hypertrophic Subaortic Stenosis

Techniques, and the Results of Preoperative and Postoperative Clinical and Hemodynamic Assessments

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SUMMARY
Operations designed to relieve obstruction to left ventricular outflow were carried out in 25 patients with idiopathic hypertrophic subaortic stenosis (IHSS). All of the patients were symptomatic and 23 were in functional classes III or IV. Preoperatively, a systolic pressure gradient within the left ventricle was demonstrated in every patient (average, 102 mm Hg), and the average left ventricular end-diastolic pressure was 19 mm Hg. Six patients had pulmonary hypertension.

During cardiopulmonary bypass, the outflow tract of the left ventricle was exposed through an aortotomy. In five early patients only ventriculomyotomy was carried out, but in all subsequent operations myotomy was combined with a limited resection of the hypertrophied muscular tissue. One patient died, apparently of arrhythmia, on the eighth postoperative day, and another 8 months after operation following cerebral thrombosis. In two other patients complete heart block was produced, and they have implanted pacemakers.

Twenty-one of the 23 surviving patients have been followed for periods of 1 to 8 years; 15 are asymptomatic (class I). Six patients have mild residual limitation of activity (class II), but none has had congestive heart failure or syncope postoperatively, and only one experiences significant angina.

Left heart catheterization has been performed on one or more occasions after operation in all 21 patients. In 19, no systolic pressure gradient was evident within the left ventricle under resting conditions, and in the other two patients, gradients of 11 and 8 mm Hg were recorded. The left ventricular end-diastolic pressure was lower in 16 of the 17 patients in whom it was abnormally elevated preoperatively, and in nine of them it was normal. The pulmonary arterial pressure was normal postoperatively in all patients who had severe pulmonary hypertension.

Additional Indexing Words:
Diffuse subvalvular aortic stenosis
Obstructive cardiomyopathy
Valsalva maneuver
Pulmonary hypertension
Cardiac catheterization
Isoproterenol

The diagnosis of idiopathic hypertrophic subaortic stenosis (IHSS) has been established in 126 patients studied at the National Heart Institute. A detailed description of the clinical, hemodynamic, and angiographic manifestations of the disease, based on studies carried out in this and other clinics, was presented in 1964, and at that time the initial experience with operative treatment in 10 patients was presented. A current report summarizes the findings in the entire group of 126 patients and provides additional specific information concerning the natural history of the disease. Operations

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designed to relieve obstructions to left ventricular outflow have now been carried out in 25 of the 126 patients with IHSS. The present report presents the pertinent preoperative clinical and hemodynamic findings in these 25 patients and describes the operative methods employed. The effectiveness of operation was assessed by the extent of symptomatic and hemodynamic improvement determined at postoperative intervals of 1 to 8 years.

Patients Operated Upon

The first patient was operated upon in January 1960, and the most recent, in August 1967; in the text, the patients are referred to by numbers which indicate the chronological order in which they were operated upon.

The 25 patients, at the time of operation, ranged in age from 10 to 61 years; 15 were men, and 10 were women. All described symptoms of limited cardiac reserve, and the incidence of specific symptoms is summarized in table 1. Eight patients were in functional class IV (New York Heart Association), 15 were in class III, and two were in class II.

Each of the patients demonstrated the physical findings characteristic of IHSS, and in all, left ventricular enlargement was evident on fluoroscopic and radiographic examinations. Twenty-three of the 25 patients were in normal sinus rhythm, while two had atrial fibrillation. Electrocardiographic findings indicative of left ventricular hypertrophy were present in all; one patient had left and one right bundle-branch block.

Table 1

<table>
<thead>
<tr>
<th>The Symptoms Described Preoperatively by Twenty-Five Patients with IHSS</th>
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<tbody>
<tr>
<td>Condition</td>
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<tr>
<td>Dyspnea with effort</td>
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<tr>
<td>Angina pectoris†</td>
</tr>
<tr>
<td>Congestive heart failure†</td>
</tr>
<tr>
<td>Syncope†</td>
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<td>Orthopnea</td>
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<td>Paroxysmal nocturnal dyspnea</td>
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<td>Paroxysmal arrhythmia</td>
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*Six patients had angina pectoris, congestive heart failure, and syncope.

Left Heart Catheterization

This was carried out on one or more occasions preoperatively in all patients, and in each a systolic pressure gradient between the left ventricle and the aorta or a peripheral artery was recorded. The magnitude of the gradient often varied from one study to another and also in the course of a given study. Therefore, the pressure gradients shown graphically in figure 1 are the highest ones recorded in the resting basal state at the left heart catheterization which immediately preceded operation. In the entire group of 25 patients, the gradients ranged from 26 to 175 mm Hg, and averaged 102 mm Hg. Abnormal elevation of the left ventricular end-diastolic pressure (>12 mm Hg) was noted in 21 of the 25 patients; the end-diastolic pressures ranged from 5 to 30 mm Hg, and the average value was 19 mm Hg (fig. 2). The cardiac index was measured at rest in 24 patients, and was abnormally reduced (<2.6 L/min/m²) in 10 of them; the indices ranged from 1.78 to 3.32 L/min/m², and the average value was 2.60 L/min/m². The calculated effective areas of the stenotic orifices ranged from 0.33
IDIOPATHIC HYPERTROPHIC SUBAORTIC STENOSIS

Figure 2

Measurements of left ventricular end-diastolic pressure made in 21 patients with IHSS preoperatively and postoperatively. The mean values of each group of values are indicated in parentheses.

to 1.50 cm² and averaged 0.65 cm². Twenty-four of the 25 patients demonstrated absence of normal postextrasystolic augmentation of the arterial pulse pressure.

Right Heart Catheterization

This was also carried out preoperatively in 22 of the 25 patients. Severe pulmonary arterial hypertension was evident in three patients, with systolic pressures of 96, 80, and 62 mm Hg, respectively; moderate elevations of the systolic pulmonary arterial pressure to 45, 43, and 42 mm Hg were recorded in three additional patients. Significant systolic gradients within the right ventricular outflow tract were recorded in four patients; the gradients were 60, 21, 28, and 11 mm Hg.

Selective Biplane or Cineangiocardiograms

These were made, with left ventricular injection, in 19 patients. Each study demonstrated the angiographic features characteristic of IHSS. Mitral regurgitation was also evident in 14 of the 19 studies, but precise definition of the severity of regurgitation was, with exceptions to be noted, usually not possible.

Operative Methods

The operative methods utilized were similar in all of the patients, and have previously been described in detail. The operation is carried out during cardiopulmonary bypass, and mild (30°C) general body hypothermia is utilized. A drainage cannula is introduced into the left ventricle through a stab wound in its apex. After occlusion of the aorta, a vertical incision is made in it and extended obliquely downward into the noncoronary sinus of Valsalva. The left coronary artery is continuously perfused through a cannula supplying oxygenated blood from the arterial return line of the heart-lung machine.

The aortic valve leaflets are retracted, and the upper portion of the mass of muscle occupying the interventricular septum and lateral free wall of the left ventricle is visible. Palpation of the ventricular cavity reveals a mound or ridge of muscle within the outflow tract, usually most prominent anterolaterally, below the commissure between the left and right coronary leaflets of the valve. In five of the first 10 patients operated upon, only ventriculomyotomy was carried out, but in all subsequent operations, ventriculomyotomy has been combined with resection of a portion of the muscle mass.

Parallel incisions are made over the mass, the first oriented to the commissure between the left and right coronary leaflets of the valve, and the second about 1 cm to the right (clockwise), oriented approximately to the center of the right coronary leaflet. The incisions extend only through the endocardium and superficial muscle layers. The deeper muscle fibers are then split by digital pressure to a depth of 2 to 3 cm, and the ridge of tissue between the myotomy incisions is then resected with an angled rongeur. When making the initial incisions, and during the resection, it is often convenient to elevate the heart and introduce the knife or rongeur from the apical stab wound. The instruments are directed with the left index finger passed from the aorta, and the anterior mitral leaflet and papillary muscles are pushed posteriorly as the resection is carried out. At the completion of the procedure, a deep channel or trough through the mass is palpable, the margins of which retract and separate during systole. The ventricle is lavaged to remove any particulate matter, and the aortotomy is closed as the patient's temperature is restored to normal.

Results of Operation

Twenty-three of the 25 patients operated upon are living. The course of one patient (no. 8) who died on the eighth postoperative
day, apparently of arrhythmia, has been described previously; of possible significance are the facts that he had been in atrial fibrillation since the second postoperative day and that he had a large systolic pressure gradient within the right ventricular outflow tract (60 mm Hg). The circumstances of the death of the second patient (no. 22) are not entirely clear. She was a woman, 60 years of age; preoperatively, a systolic pressure gradient of 88 mm Hg was recorded within the left ventricle, and the end-diastolic pressure was 20 mm Hg. The pulmonary arterial pressure was 43/18 mm Hg. Effective relief of obstruction was documented at the conclusion of operation. She evidenced left ventricular failure in the early postoperative period, but thereafter her convalescence was slow but satisfactory. She did well at home for 6 months but was then admitted to her local hospital with cardiac failure, and shortly thereafter developed hemiparesis, apparently the result of cerebral thrombosis. She remained paralyzed and died 8 months after operation. Autopsy was not performed.

Complete heart block was produced at operation in two of the first patients operated upon (nos. 3 and 9), and in both patients the heart rate has been maintained by implanted pacemakers. In one of the patients in whom heart block was produced, a small interventricular septal defect was also created; no attempt was made to close the defect, and subsequent hemodynamic studies have revealed a small left-to-right shunt through the defect, Qr/Qs = 1.48. Patient 11 apparently had a cerebral hemorrhage on the twelfth postoperative day, but recovered fully and has no neurological sequelae; the relation of this complication to the operation is obscure.

Twenty-one of the 23 surviving patients have been followed for a period of at least 1 year after operation. The intervals after operation range from 1 to 8 years (average, 3.9 years) and comprise a total of 68 patient-years of postoperative observation. All of the 21 patients have returned to the Institute on one or more occasions for detailed clinical and hemodynamic reassessment.

**Symptomatic Improvement**

Distinct symptomatic improvement has been described by all patients. The functional classifications of the 21 patients, determined at the time of most recent postoperative evaluation, are shown, in comparison with their preoperative status, in figure 3. Fifteen of the 21 patients are asymptomatic (class I). Nine of these 15 patients are men, eight of whom are employed fulltime; the ninth, a youth of 18, is in preparatory school where he participates in athletics. One of the six women works full time as a research assistant, and the other five (one of whom has a pacemaker) carry on their home and family responsibilities without special assistance.

Six of the 21 patients are considered to be functional class II, and the severity of their residual symptoms is worthy of detailed description. Patient 9 (class IV before operation), who has a pacemaker, was asymptomatic for 2 years after operation, but then

**Figure 3**

Functional classification (New York Heart Association) of 21 patients with IHSS before and after operation. Duration of follow-up was 1 to 8 years; mean follow-up was 3.9 years. The number within each box indicates the number of patients in that class.

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began to describe bizarre symptoms related to her wound, her abdomen, and her head. Evaluations at both the Clinical Center and her referring hospital indicate no change in her cardiovascular status, but she has been found to be psychotic (paranoid schizophrenia). Patient 12 (class III before operation) experiences angina on exertion approximately twice each month. She does all her housework without assistance, however, and takes no medicines. Patient 13 (class III before operation), the oldest patient in the group, was aged 61 at operation, and is now 64. She experiences occasional, mild precordial pain with exertion or excitement, and sometimes has edema of the ankles at the end of her day's work as a cashier. She takes digoxin and, intermittently, an oral diuretic. Patient 15 was totally incapacitated preoperatively (class IV), and he had been hospitalized on many occasions because of episodes of paroxysmal atrial fibrillation which always led to the left and right heart failure. Two years after operation he is in stable atrial fibrillation, experiences dyspnea after walking three blocks, and is conscious of palpitations. He has not returned to work. He has had several attacks of supraventricular tachycardia, but these are now controlled with digoxin and 30 mg of propranolol daily. Patient 18 (class IV before operation) works full time as an airline executive, but about once a week experiences precordial pain of 5 to 10 minutes' duration, usually precipitated by exertion or anxiety. He takes no medicines. Patient 20 (class III before operation) once or twice a week experiences severe chest and arm pain which is probably angina pectoris, although it is atypical in nature. A selective coronary arteriogram has subsequently revealed moderate narrowing of the left circumflex coronary artery at its origin. The relation of this finding to his precordial pain remains conjectural. He is not employed, but cares for his home and four children while his wife works. He takes propranolol, 100 mg daily, and a coronary vasodilator.

In the entire group of 21 patients, none has had syncope or congestive heart failure after operation, and with the possible exception of patient 20 described above, none has angina pectoris which limits physical activity.

**Postoperative Hemodynamic Evaluation**

Postoperative hemodynamic studies have been carried out in each of the 21 patients who has been followed 1 year or more. Pertinent data obtained postoperatively are shown graphically and in comparison to the preoperative values in figures 1 and 2. In the case of patients who have been studied on more than one occasion, the values depicted are those recorded at the most recent catheterization. In 19 of the 21 patients, no systolic pressure gradient within the left ventricle was present under resting conditions; in the remaining two patients, peak systolic gradients of 11 and 8 mm Hg were evident (fig. 1). Seventeen of the 21 patients had abnormally elevated left ventricular end-diastolic pressures preoperatively; in 16 of the 17 the pressure was found to be lower postoperatively, and in nine it was normal (fig. 2). In the entire group the average end-diastolic pressure was 20 mm Hg preoperatively and 11 mm Hg postoperatively. In eight of 19 patients the cardiac index at rest was abnormally low preoperatively, and in five of them it was normal postoperatively. In the group of 19 patients in whom the cardiac index was determined both before and after operation, the average cardiac index preoperatively was 2.61 L/min/m² and 2.87 L/min/m² postoperatively. Abnormal postextrasystolic augmentation of the arterial pulse pressure was noted preoperatively in 20 of the 21 patients; a normal response was recorded in 17 patients after operation.

In 19 patients measurements of left ventricular and systemic arterial pressure were made not only under resting basal conditions, but also during one or more physiological or pharmacological interventions which may provoke an intraventricular pressure gradient or increase the magnitude of an existing one. The results of these studies are shown graphically in figure 4.
The effects of the Valsalva maneuver were determined in 14 patients. In six patients no gradient was evident under basal conditions, and none was provoked. In seven patients without obstruction at rest, gradients of 10 to 50 mm Hg (average, 28 mm Hg) appeared during the Valsalva maneuver; in the one patient with a gradient of 11 mm Hg at rest, it increased to 25 mm Hg.

Isoproterenol infusions were carried out in 10 patients. In four, no gradient was evident before or after the drug was given, while five patients without obstruction initially developed gradients of 21 to 52 mm Hg (average, 29 mm Hg); in the remaining patient a gradient of 8 mm Hg increased to 44 mm Hg.

Serial left heart catheterizations were performed in seven patients in whom the initial postoperative study revealed residual obstruction under basal conditions. The data on these patients are presented graphically in figure 5. In five patients who early after operation had residual gradients of 18 to 65 mm Hg, no gradient was evident at the time of later study 15 to 38 months after the first postoperative catheterization. In the remaining two patients, initial resting gradients of 44 and 30 mm Hg fell to 11 and 8 mm Hg, respectively, after 17 and 12 months.

Right heart catheterization was performed both preoperatively and postoperatively in 17 of the 21 patients. In the four patients in whom the systolic pulmonary arterial pressures were 96, 62, 45, and 80 mm Hg preoperatively, the postoperative values were 28, 28, 27, and 34 mm Hg, respectively. In the fifth patient who had significant pulmonary hypertension, the preoperative and postoperative systolic pressures were 42 and 40 mm Hg, respectively. Preoperatively, three of the 21 patients had gradients within the right ventricular outflow tract of 21, 28, and 11 mm Hg; in each a gradient was again recorded postoperatively, and the values were 10, 12, and 16 mm Hg, respectively.

Selective Left Ventricular Biplane or Cineangiograms
These were obtained both before and after operation in seven patients. In four of them mitral regurgitation was evident preoperatively, but the valve was entirely competent at

**Figure 4**
Peak systolic pressure gradients between the left ventricle and a peripheral artery recorded postoperatively in 19 patients studied at rest and during the Valsalva maneuver (left panel) or during isoproterenol infusion (right panel).
the time of postoperative study. In one patient mild regurgitation was evident both preoperatively and postoperatively, and in the remaining two patients there was no evidence of regurgitation either before or after operation. In patient 25, who has not been studied postoperatively, mitral regurgitation was shown by preoperative angigrams, but the valve was competent when palpated at the conclusion of operation.

Association of IHSS and Mitral Regurgitation

The association of IHSS and mitral regurgitation in patient 23 is worthy of special comment. This woman, aged 32 years, had been severely symptomatic for 3 years; she had experienced progressively severe angina and dyspnea, had been hospitalized on many occasions for treatment of congestive heart failure, and twice had had overt pulmonary edema. Cardiac catheterization performed at her referring hospital in October 1966 revealed pulmonary arterial pressure of 50/20 mm Hg, a gradient of 70 mm Hg within the left ventricle, and an end-diastolic pressure of 30 mm Hg. A left ventricular cineangiogram, subsequently reviewed at the Institute, revealed no mitral regurgitation.

On admission, February 1967, she was emaciated, severely dyspneic, and orthopneic. A grade IV/VI holosystolic murmur was heard at the left sternal border, radiating to both the base of the heart and the axilla. Cardiac catheterization revealed a pulmonary arterial pressure of 80/65 mm Hg, and a gradient of only 26 mm Hg within the left ventricle; the left ventricular end-diastolic pressure was 28 mm Hg. A cineangiogram with left ventricular injection demonstrated severe mitral regurgitation, as well as the deformed and hypertrophied left ventricle characteristic of IHSS.

At operation, palpation of the mitral valve revealed it to be severely incompetent, and an intense jet of regurgitant blood appeared to originate near the center of the valve orifice. The mean left atrial pressure was 38 mm Hg, and the v wave was 60 mm Hg. After institution of bypass the left ventricular outflow tract was exposed, and a typical ridge of muscular tissue was incised and resected as described above. The aortic leaflet of the mitral valve, as viewed through the aortotomy, was somewhat thickened, but otherwise appeared normal. After restoration of the circulation, the mitral valve was again palpated, and was found to be entirely competent. At the conclusion of operation the mean left atrial pressure was 18 mm Hg, and the v wave was 25 mm Hg.

The patient made an uneventful recovery. Six months after operation she was asymptomatic, and had gained 24 pounds. No pressure gradient was recorded within the left ventricle at rest, during the Valsalva maneuver, or during infusion of isoproterenol. The pulmonary arterial pressure had fallen to 34/16 mm Hg. A selective angiogram revealed that the mitral valve was completely competent.

Comment

In this clinic, operative treatment has usually been recommended only to those patients with IHSS who are distinctly symptomatic, and in whom severe obstruction to left ventricular outflow has been documented at cardiac catheterization, preferably on more than one occasion. Within the past 2 years, symptomatic patients, with IHSS, particularly those with severe angina pectoris, have been treated with propranolol. Thus, a number of patients who have been operated upon recently are ones in whom propranolol was ineffective in relieving symptoms. Also, three patients operated upon within the past year had intraventricular pressure gradients at rest of 50 mm Hg or less, and by usual criteria they could not be considered to have had severe obstruction. All were in functional class IV. In two, resting gradients of 50 and 40 mm Hg increased to 85 and 78 mm Hg with provocative interventions. Both patients have described excellent symptomatic improvement. The third patient (no. 23) in whom the gradient at rest was only 26 mm Hg, is the woman with associated severe mitral regurgitation described above. Thus,
at the present time, operative treatment may be considered when a patient with IHSS (1) has disabling symptoms, (2) does not improve significantly with propranolol, when a therapeutic trial with this drug is indicated, and either (3) has severe obstruction to outflow at rest, or (4) has severe obstruction provoked by exercise, the Valsalva maneuver, or the administration of isoproterenol, or all three.

The experience with the patients described in this report indicates that the risk of operation in patients with IHSS is low, and abolition or marked improvement of symptoms can be expected in all patients who survive. A comparison of the preoperative and postoperative hemodynamic assessments also demonstrates that clinical improvement is paralleled by a return of the intracardiac pressures to normal or near-normal levels. Thus 19 of the 21 patients studied postoperatively had no resting pressure gradient in the left ventricle, and in the other two, the gradients were only 11 and 8 mm Hg. In 13 of the 21 patients the left ventricular end-diastolic pressure was normal after operation, and all those with severe pulmonary hypertension before operation had normal pulmonary arterial pressures afterward.

In a previous report from this clinic,2 the various operative methods which have been employed by other surgeons in patients with IHSS were reviewed in considerable detail, and all available data concerning the clinical and hemodynamic results following the various operations were summarized. These, and more recent reports, indicate that a number of operative methods are currently being utilized, ranging from simple transaortic ventriculotomy to extensive resections performed after wide exposure of the ventricular outflow tract through an incision in the left atrium and mitral valve or an apical left ventriculotomy. There is, however, no evidence that resection of a large amount of tissue from the outflow tract is necessary to abolish the intraventricular pressure gradient. The operative method described, transaortic myotomy and limited muscular resection, can be carried out with little risk, even in seriously ill patients, and the postoperative data indicate that it is also effective in relieving outflow obstruction. The procedure would appear, therefore, to be worthy of continued application.

References

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