REPLACEMENT AND/OR REPAIR OF THE MITRAL VALVE AS TREATMENT OF IDIOPATHIC HYPERTROPHIC SUBAORTIC STENOSIS

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The condition known as idiopathic hypertrophic subaortic stenosis (IHSS) remains a curious pathologic entity which presents many unresolved problems and areas for speculation and conjecture. Etiologic factors, pathogenesis, clinical manifestations, and prognosis are extremely variable. Rather than being a single disease, IHSS may be a spectrum of hypertrophic cardiomyopathies with some similarities.

The first description of the pathologic anatomy of IHSS was made in 1907 by Schmincke,1 who considered the condition to be congenital in origin. He conceived of the pathologic consequences as a vicious cycle in which the muscular hypertrophy of the outflow tract led to further obstruction, causing more hypertrophy and thus further obstruction. In 1910 Bernheim2 added to the knowledge of the condition, and in 1952 Davies3 described a family in which many members had cardiac symptoms ascribable to IHSS. Brock's report4 in 1957 focused more clinical attention on the condition, and since then many reports have appeared in the medical literature. The contributions of Braunwald,5 Morrow,6 Mason7 and others at the National Institutes of Health are well known. Their opinion, based upon experience with medical and surgical management of a large number of patients with IHSS, has been widely accepted. According to this group the logical approach to surgical relief of IHSS was selective incision or partial excision of the hypertrophied septum (hypertroph). Experience with that approach was not entirely satisfactory, and we subsequently devised a technique of myomectomy on the right side of the ventricular septum.8,9 In our experience, results with these techniques were not uniformly satisfactory and other methods of correction were sought.

Several years ago we presented a new approach to surgical relief of the outflow obstruction.10,11 Rather than using the standard attack on the hypertrophied myocardium, we described results in nine patients in whom the mitral valve was replaced with a prosthesis. Since that report, 24 more patients have undergone, as definitive treatment, mitral valve replacement (MVR) without concomitant septal myotomy or myectomy. Recently a patient with IHSS had severe mitral regurgitation and received a Carpentier mitral ring which relieved the valvular incompetence. This technique may provide still another means of relieving the outflow tract obstruction in IHSS. Results of the experience with mitral replacement and annuloplasty are described.

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Mitral Valve Replacement

During the period from June 4, 1970 to October 1, 1976, we operated upon 33 patients with IHSS by using only MVR without surgical excision of myocardial tissue. All patients so treated during this period were included in the series with the exception of two patients: one demonstrated angiographic features of IHSS but had no left ventricular-to-ascending aortic gradient (excellent result); the other died one month postoperatively due to material failure and strut fracture of the prosthetic valve, but the result was not considered to be caused directly by this form of treatment.

Ages of the patients ranged from 14 to 71 years with a mean of 47.6 years (Table I). Twenty-one patients (64%) were men and 12 (36%) were women (Table II).

Congestive heart failure (CHF) was present preoperatively in 27 of the 33 patients. All but three patients demonstrated mitral insufficiency of some degree, although not hemodynamically significant in many. Chest pain was a prominent symptom in 21 of the 33 patients (Table III).

Cardiac catheterization with measurement of pressure gradients between the left ventricle and aorta was performed preoperatively in all patients, but in four patients the only gradients available for this analysis were those measured at the time of operation. Preoperative pressure gradients ranged from 20 to 140 mm Hg with a mean of 74 mm Hg.

Table I. Age range of 27 patients who underwent MVR for IHSS

<table>
<thead>
<tr>
<th>Age</th>
<th>No. of Patients</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 - 19</td>
<td>2</td>
<td>6.0</td>
</tr>
<tr>
<td>20 - 29</td>
<td>2</td>
<td>6.0</td>
</tr>
<tr>
<td>30 - 39</td>
<td>8</td>
<td>24.3</td>
</tr>
<tr>
<td>40 - 49</td>
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<td>24.3</td>
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<tr>
<td>50 - 59</td>
<td>8</td>
<td>24.3</td>
</tr>
<tr>
<td>60 - 71</td>
<td>5</td>
<td>15.1</td>
</tr>
<tr>
<td>TOTAL</td>
<td>33</td>
<td>100.0</td>
</tr>
</tbody>
</table>
Operation was performed under normothermia or moderate hypothermia utilizing disposable bubble oxygenation cardiopulmonary bypass under ischemic cardiac arrest. The Cooley-Cutter\textsuperscript{12} low profile mitral disc prosthesis was used in the 24 most recent cases, and the Bjork-Shiley\textsuperscript{13} mitral prosthesis was used in the nine earlier patients (Table V). The prostheses should be secured firmly by using mattress sutures and felt pledgets to avoid valve dehiscence in the soft mitral annulus.

Early results, evaluated at the time of discharge from the hospital, demonstrated 28 patients (84.9\%) to have experienced an excellent result with complete or significant relief of symptoms and reversal of altered cardiac dynamics (Table IV). Four patients (12.1\%) were considered to have experienced some improvement, and one patient died during the early postoperative period following cardiac arrest and a cerebral infarction (Table IV).

| Table II. Sex of 27 patients who underwent MVR for IHSS |
|---|---|---|
| **Sex** | **No. of Patients** | **Percent** |
| Male | 21 | 64\% |
| Female | 12 | 36\% |
| TOTAL | 33 | 100.0 |

| Table III. Symptoms and clinical findings |
|---|---|---|---|---|---|
| **Symptom** | **None** | **Mild** | **Moderate** | **Severe** | **Total** |
| CHF | 6 | 5 | 10 | 12 | 33 |
| MI | 3 | 6 | 12 | 12 | 33 |
| Chest pain | 12 | 5 | 8 | 8 | 33 |

CHF = congestive heart failure; MI = mitral insufficiency
In the follow-up period, we know of six other deaths. Causes of late death included rupture of dissecting aneurysm of the thoracic aorta (one patient), valve dehiscence (two patients), congestive heart failure (two patients), and myocardial infarction (one patient).

Pressure gradients across the left ventricular outflow tract were obtained either at the time of operation (two patients) or later at cardiac catheterization (26 patients). The mean postoperative pressure gradient was 6.59 mm Hg compared to the mean preoperative gradient of 74 mm Hg (Table V).

CASE REPORT

**Mitral Annuloplasty (Carpentier Ring)**

A 61-year-old retired welder was admitted to the Texas Heart Institute for evaluation of severe chest pains and episodes of dizziness.* His chest pain was disabling and exacerbated by minimal exercise and meals; however, the pain was incompletely relieved by nitroglycerin. Physical examination was normal except for cardiomegaly and a grade III/VI apical holosystolic murmur which was transmitted to the axilla. A routine laboratory examination was normal; however, the EKG revealed left ventricular hypertrophy, and a cardiac echo examination showed asymmetric septal hypertrophy and prolapse of the anterior mitral leaflet into the aortic outflow tract. Cardiac catheterization documented the idiopathic hypertrophic subaortic stenosis with a left ventricular pressure to 240/4 mm Hg and a 115/60 mm Hg aortic pressure. There was also moderate right ven-

Table IV. Early results (in-hospital)

<table>
<thead>
<tr>
<th>Result</th>
<th>No. of Patients</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excellent</td>
<td>28</td>
<td>84.9</td>
</tr>
<tr>
<td>Improved</td>
<td>4</td>
<td>12.1</td>
</tr>
<tr>
<td>Unchanged</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Worse</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Died</td>
<td>1</td>
<td>3.0</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td><strong>33</strong></td>
<td><strong>100.0</strong></td>
</tr>
</tbody>
</table>

*Patient of Dr. Orlando Terneny
<table>
<thead>
<tr>
<th>Valve</th>
<th>No. of Patients with Post-op Cath Data</th>
<th>Mean Postoperative Gradient* (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bjork-Shiley</td>
<td>9</td>
<td>0</td>
</tr>
<tr>
<td>Cooley-Cutter</td>
<td>24</td>
<td>11.15</td>
</tr>
<tr>
<td>TOTAL</td>
<td>33</td>
<td>6.59</td>
</tr>
</tbody>
</table>

*Measured in 28 early survivors
Fig. 1 a&b. Insertion of Carpentier ring in 61-year-old patient with IHSS and mitral regurgitation.

Above (a) interrupted sutures are placed in the mitral annulus and attached to the ring.
tricular outflow tract obstruction with a right ventricular pressure of 68 mm Hg and 36 mm Hg pulmonary artery pressure. Coronary angiography revealed significant occlusive disease (greater than 85% of vessel diameter) of the right, left anterior descending, and circumflex coronary arteries. In addition, moderate mitral regurgitation was present.

On October 8, 1976, he underwent left ventricular septectomy of the subaortic muscular ridge. To relieve the prolapsing anterior mitral leaflet and mitral regurgitation a mitral annuloplasty with a Carpentier ring (Number 26) was accomplished (Fig. 1 a&b). Because of his severe coronary artery disease, a coronary bypass was performed to his right, left anterior descending, and obtuse marginal coronary arteries.

His postoperative recovery was uneventful, and he underwent repeat cardiac catheterization twelve days postoperatively. This revealed no gradient across either the left or right ventricular outflow tracts. His left ventricular pressure was 115/4, and the coronary bypass grafts were all patent; there was no mitral regurgitation or anterior mitral leaflet prolapse. He has continued to have uneventful convalescence and has experienced no chest pain.

Above (b) sutures are tied, securing the ring in the annulus.
DISCUSSION

Introduction of echocardiography into cardiac diagnosis has added another significant diagnostic tool to the pressure measurements and left ventriculography performed at cardiac catheterization. Echocardiography has elucidated the action of the anterior leaflet of the mitral valve in patients with IHSS and suggests that it may be either an etiologic or aggravating factor in pathogenesis of the disease.\textsuperscript{14,15} The outflow tract of the left ventricle is surrounded by two tissue elements, the myocardium

\begin{center}
\begin{tabular}{c c}
\textbf{DIASTOLE} & \textbf{SYSTOLE} \\
\includegraphics[width=0.4\textwidth]{dia_stole.png} & \includegraphics[width=0.4\textwidth]{sys_stole.png}
\end{tabular}
\end{center}

\begin{center}
\begin{tabular}{c c}
\textbf{NORMAL} & \textbf{IHSS} \\
\includegraphics[width=0.4\textwidth]{norm.png} & \includegraphics[width=0.4\textwidth]{ihss.png}
\end{tabular}
\end{center}

* Interventricular septum  
** Left ventricular cavity  
*** Right ventricular cavity

Fig. 2. Drawing showing the relationship of the anterior leaflet of the mitral valve to the interventricular septum during diastole and systole in normal and IHSS patients. Arrows indicate the motion of the leaflet.
and the anterior leaflet of the mitral valve (Fig. 2). Echocardiography has allowed demonstration of a characteristic feature in patients with IHSS. During systolic contraction of the left ventricle the anterior mitral leaflet moves forward toward the hypertrophied interventricular septum, while in normal patients the mitral leaflet moves posteriorly. The cause for systolic anterior motion of the mitral valve (SAM) has been conjectured. The leaflet could be sucked forward into the outflow tract as a Venturi effect or may fold forward due to the abnormal muscle mass posteriorly or abnormal traction by the papillary muscles. Regardless of the cause for SAM one cannot disregard the probability that the mitral valve leaflet plays a significant role in the pathophysiology of IHSS. The frequency of clinical mitral valve regurgitation in patients with IHSS further incriminates the valve as a causative or aggravating factor.

**SURGICAL APPROACH TO IHSS**

Fig. 3. Drawing showing three methods of relieving left ventricular outflow obstruction in IHSS. Right ventricular septectomy merely weakens the septum, allowing it to move away from the mitral leaflet.
Surgical treatment of IHSS must be directed toward relief of the obstruction to the outflow. This may be accomplished by either an attack on the hypertrophied myocardium in the interventricular septum or by changing the mechanism of the mitral valve. The septum may be excised on the left side providing a channel for the blood to follow or from the right side permitting the septum to move forward and away from the mitral valve (Fig. 3).

In 1970 we had our first opportunity to assess the effect on left ventricular outflow obstruction by doing nothing more than replacing the mitral valve. A 53-year-old patient with IHSS and severe mitral regurgitation underwent MVR with a Bjork-Shiley low profile disc prosthesis without incision or excision of the hypertrophied muscle mass. Not only was the mitral regurgitation relieved but also the pressure gradient across the left ventricular outflow tract was eliminated. The clinical result was excellent, and six years later he remains in excellent condition. Since that time, 32 other patients have undergone MVR as the sole method of relief of outflow obstruction. In all surviving patients the pressure gradient has been eliminated and most have had symptomatic relief. Although the rigid ring of the prosthesis remains fixed during systole, it does not advance on the hypertrophied septum and apparently no outflow obstruction remains.

The use of the annular ring described by Carpentier should prevent systolic anterior motion of the mitral valve (Fig. 4). Obviously, the experience with one patient reported herein is too recent to draw conclusions.
about this technique in the physiologic relief of IHSS. Moreover, the excellent result, as evidenced by the early postoperative catheterization, could be either entirely or partly due to the outflow myotomy and myectomy which was done. Thus, this case is different from the patients with MVR reported in whom no hypertrophied myocardium was incised or excised. In the future we plan to attempt relief of the IHSS by Carpentier ring annuloplasty alone to ascertain whether it would provide relief of the outflow pressure gradient. An advantage of this technique over MVR would be that the patient would not require postoperative anticoagulation, and that complications of prosthetic valves would be avoided.

COMMENT

This experience has led to a controversy regarding the selection of MVR or septal myectomy as definitive treatment of IHSS.

Advantages of MVR

1. Complete and predictable relief of outflow obstruction even in the most advanced stages of the disease.
2. Elimination of the mitral regurgitation.
3. Avoidance of postoperative conduction disturbance and electrocardiographic consequences of septal myotomy or myectomy. May be applied with safety in patients with right bundle-branch block or serious damage to the left bundle or the fascicles.
4. Prevention of septal infarction or perforation due to division of coronary tributaries.
5. May be applied successfully to patients who have failed to respond to myotomy or myectomy.

Disadvantages of MVR

1. Complications may result from the valve prosthesis (thromboembolism, dehiscence, infection, mechanical obstruction of the prosthesis, etc.).
2. Relative difficulty in valve replacement in patients with extreme cardiomegaly and left ventricular hypertrophy.

Indication for MVR

As previously stated, selection of the appropriate method of treatment (medical or surgical) depends upon the clinical findings, severity of the symptoms, age of the patient and other factors. Surgical treatment of IHSS should be reserved for those patients who have incapacitating symptoms, CHF, severe left ventricular hypertension, significant electrocardiographic changes of myocardial ischemia and strain patterns, and the presence of symptoms of angina pectoris. Most young patients less than 20 years of age may be managed without medication or with a moderate medical regimen. Many older patients with mild-to-moderate symptoms may be managed with Propranolol, Valium and other medications. Failure
of nonsurgical treatment, therefore, provides the only generally acceptable indication for operation. The choices of operation now include myotomy or myectomy, usually approaching the left side of the septum, but occasionally the right, and replacement of the mitral valve.

We believe that MVR should be the first choice in patients with IHSS who have the following manifestations:

1. Severe mitral valve dysfunction including involvement by rheumatic or bacterial endocarditis, and in patients with ruptured chordae tendinae.
2. Elderly patients with advanced stages of cardiomyopathy complicated by ischemic myocardial heart disease.
3. Patients with complicating electrocardiographic findings including right bundle-branch block, intermittent A-V dissociation or incomplete left bundle-branch block.
4. Patients who have failed to respond to septal myotomy or myectomy.
5. Patients who are unable to tolerate Propranolol even in small doses.

SUMMARY

Treatment of idiopathic hypertrophic subaortic stenosis (IHSS) remains a controversial problem, and medical or surgical treatment may be elected depending upon many factors. When medical therapy fails and surgery is recommended, choice of an appropriate surgical technique may be difficult.

An analysis is given of 33 patients who have undergone only mitral valve replacement as definitive treatment. Thirty-two patients were dismissed from the hospital with good or excellent results, and one died (3.0% mortality). Pressure gradients across the left ventricular outflow tract after operation were eliminated in every instance. The mean preoperative gradient was 74 mm Hg and postoperatively was 6.9 mm Hg.

A case is reported in which the Carpentier mitral ring was used rather than mitral valve replacement. Annuloplasty may be sufficient to eliminate the systolic anterior motion (SAM) of the mitral valve, which plays a role in the pathogenesis of IHSS.

REFERENCES