Surgical Treatment for Hypertrophic Obstructive Cardiomyopathy

Evaristo Castedo,a Rubén A. Cabo,a Iván Núñez,a Emilio Monguío,a Carlos G. Montero,a Raúl Burgos,a Santiago Serrano-Fiz,a Gabriel Téllez,a Magdalena González,b Miguel A. Cavero,b and Juan Ugartea

aDepartamento de Cirugía Cardiovascular, Clínica Puerta de Hierro, Madrid. bDepartamento de Cardiología, Clínica Puerta de Hierro, Madrid, Spain.

Introduction and objectives. Five percent of the patients with hypertrophic obstructive cardiomyopathy (HOCM) have symptoms unresponsive to medical treatment and are candidates for invasive therapy. The objective of this study was to analyze our results with surgical treatment of HOCM during the last 10 years.

Patients and method. Between July 1993 and January 2004 26 patients with HOCM refractory to drug therapy were operated on. An extended septal myectomy was performed, in combination with anterior mitral leaflet plication in 19 cases (73%) and with mitral valve replacement in 5 (19%). Evolution of the grade of dyspnea, left ventricle outflow tract gradient (LVOTG), mitral regurgitation, and systolic anterior motion after surgery was analyzed.

Results. Mean follow-up was 63 (37) months. After surgery, a significant reduction in LVOTG (from 96.5 to 19.5 mmHg; P<.001), grade of mitral regurgitation (from 2.54 to 0.69; P<.001) and systolic anterior motion (from 2.92 to 0.23; P<.001) was achieved, which led to improvement in functional class. Hospital mortality and need for pacemaker implantation due to complete heart block after surgery was 3.8% (n=1). There were no cases of iatrogenic ventricular septal defect or mitro-aortic valve injury. Actuarial survival at 5 years was 96% (4%).

Conclusions. Surgery in patients with HOCM yields great clinical improvements with low morbidity and mortality. Simultaneous intervention for both myocardial and valvular components of the disease allows not only reduction in the LVOTG but also correction of mitral regurgitation and abolition of systolic anterior motion.

Key words: Hypertrophic cardiomyopathy. Surgery. Mitral regurgitation. Valvuloplasty.

Full English text available at: www.revespcardiol.org

Tratamiento quirúrgico de la miocardiopatía hipertrófica obstructiva

Introducción y objetivos. Un 5% de los pacientes con miocardiopatía hipertrófica obstructiva (MHO) tienen síntomas pertinaces al tratamiento médico y son candidatos a una terapia invasiva. El objetivo de este trabajo fue analizar nuestros resultados en la cirugía de la MHO durante los últimos 10 años.

Pacientes y método. Entre julio de 1993 y enero de 2004 hemos intervenido a 26 pacientes con MHO pertinaz a tratamiento farmacológico. El procedimiento realizado fue una miectomía septal extendida, que se asoció a plicatura del velo anterior mitral (PVA) en 19 (73%) casos y a reemplazo valvular mitral en 5 (19%). Se analizaron la evolución posquirúrgica del grado de disnea, el gradiente en el tracto de salida del ventrículo izquierdo (GTSVI), la insuficiencia mitral (IM) y el movimiento sistólico anterior (SAM).

Resultados. El seguimiento medio fue de 63 ± 37 meses. Tras la cirugía se produjo una reducción significativa del GTSVI (de 96,5 a 19,5 mmHg; p < 0,001), del grado de IM (de 2,54 a 0,69; p < 0,001) y del SAM (de 2,92 a 0,23; p < 0,001), que se tradujo en una mejoría en la clase funcional de los pacientes. La mortalidad hospitalaria y la necesidad de marcapasos por bloqueo completo poscirugía fueron del 3,8% (n = 1). No ha habido ningún caso de comunicación interventricular (CIV) ni de daño valvular mitroaórtico iatrogénico. La supervivencia actuarial fue del 96 ± 4% a los 5 años.

Conclusiones. La cirugía en pacientes con MHO produce una gran mejora clínica con una baja morbimortalidad. Al tratar simultáneamente los componentes miocárdico y valvular de la enfermedad, permite no sólo reducir el GTSVI, sino corregir la IM y suprimir el SAM.


INTRODUCTION

Hypertrophic obstructive cardiomyopathy (HOCM) usually responds very well to drug therapy.1,3 The administration of negative inotropic agents improves symptoms, functional capacity and quality of life of
most patients with hemodynamically significant obstruction. However, about 5% of patients present persistent symptoms with medical treatment and are eligible for invasive treatment, whether DDD pacemaker implantation, percutaneous transluminal septal myocardial ablation or surgery.

The purpose of this study was to analyze the outcome of surgical treatment in patients with HOCM refractory to drug therapy who have undergone surgery at our hospital over the last ten years. We studied the influence of myectomy with mitral repair or replacement on functional capacity and on the severity of intraventricular hemodynamic obstruction, assessing the postoperative progress of the left ventricle outflow tract gradient (LVOTG), mitral regurgitation (MR) and systolic anterior motion (SAM).

**PATIENTS AND METHODS**

Between July 1993 and January 2004, 26 patients with HOCM refractory to drug therapy underwent surgery. The mean age was 55±12 years (range, 37-74 years), and 58% (n=15) were women. On echocardiography, all patients presented severe obstruction (LVOTG at rest >60 mm Hg, MR2grade 2/4 and SAM) and were symptomatic despite maximum medical therapy with beta-blockers and calcium channel blockers: 92% had dyspnea, 54% angina, and 8% a history of syncope. Two of the patients had received a dual-chamber pacemaker prior to surgery, but had not improved. Surgery was done urgently in 2 (8%) cases: one 51-year-old woman, who had debuted with acute pulmonary edema secondary to acute mitral valve endocarditis and severe MR, and one 41-year-old man, who was admitted in cardiogenic shock and intubated in the emergency room.

The surgical procedure consisted of myectomy plus anterior mitral leaflet plication in 19 (73%) cases, myectomy plus mitral valve replacement in 5 (19%) and myectomy alone in 2 (8%). In 2 patients, an associated procedure was also performed: coronary revascularization and aortic valve replacement, respectively.

### Extended Septal Myectomy and Anterior Mitral Valve Plication

The surgery was performed by midline sternotomy. After cannulation of the ascending aorta and right atrium, on-pump circulation was started and the patient was cooled to 32°C. The interventricular septum was accessed by oblique or hockey-stick aortotomy toward the noncoronary aortic leaflet. A retractor was inserted through the aortic annulus for traction and protection of the right coronary leaflet. Subsequently, a 15 scalpel was used to resect a 4-cm long, 1-cm wide, 1.5-cm thick fragment of the interventricular septum below the leaflet, preserving the 5 mm immediately below the aortic valve and right coronary-noncoronary commissure. The resection extended distally to the insertion site of the papillary muscles (extended septal myotomy). Through the aortotomy, plication of the A2 segment of the anterior mitral leaflet in its ventricular aspect and along its longitudinal axis was done using four interrupted 5-0 polypropylene sutures. In 2 patients, in whom the surgeon found no structural anomalies in the anterior mitral leaflet, myectomy alone was performed with no associated mitral repair. In 8 cases, in order to optimize visualization of the septum and the mitral subvalvular apparatus, the surgery was performed with the help of a cardioscope and rigid optics of 4 mm diameter, 230 mm length, and 70° angulation (Storz Instruments, Tuttingen, Germany), that was connected to a light source and video monitor. When the patient was taken off-pump, an intraoperative transesophageal echocardiogram was performed to confirm the decrease in LVOTG and correction of the MR and SAM.

### Mitral Valve Replacement

In 5 patients, the native mitral valve was replaced with a mechanical valve. There were several reasons for this approach: a) acute (n=1) or subacute (n=1) endocarditis with massive destruction of the mitral leaflets; b) emergency situation, hemodynamically unstable patient, and echocardiography specialist unavailable for the operating theater (n=1); c) aortic valve replacement required because of associated aortic regurgitation (n=1), and d) surgeon’s preference due to more confidence in this technique (n=1). The valve was replaced by resection of the valve leaflets and the subvalvular apparatus, including a portion of the papillary muscles.
Variables and Statistical Analysis

The patients were followed up clinically and by echocardiography; a final check-up was done of all patients when follow-up was discontinued. After surgery, the changes occurring in several clinical (degree of dyspnea) and echocardiographic variables (interventricular septal thickness, LVOTG, MR, and SAM grade, and left ventricular ejection fraction) were assessed. Mitral regurgitation was classified into four grades based on the regurgitant volume: 1, <30 mL; 2, 30-44 mL; 3, 45-59 mL, and 4, ≥60 mL. Four SAM grades were established based on the septum-anterior leaflet distance during systole on the parasternal long-axis view: 0, no SAM; 1, if >10 mm; 2, <10 mm, and 3, 0 mm. The pre- and post-myectomy quantitative variables were assessed by a Student’s t test for paired data, and the actuarial survival analysis, by Kaplan-Meier.

RESULTS

Mean follow-up of the patients was 63±37 months. Comparison of the pre- and postoperative clinical and echocardiographic data is contained in Table 1. The surgery led to a significant decrease in the interventricular septal thickness and LVOTG, corrected the mitral regurgitation and eliminated the SAM. The patients experienced an evident improvement in functional capacity: at the last follow-up, 77% (n=20) were NYHA (New York Heart Association) functional class I, 19% (n=5) were functional class II, and 96% had no angina.

There was no operative mortality. In 2 (8%) patients, resumption of on-pump circulation was necessary because of persistent SAM and MR after the myectomy, for which a larger resection was completed. One patient (3.8%) was reoperated for bleeding. The patient in preoperative cardiogenic shock required intra-aortic balloon counterpulsation in the immediate postoperative period. There were no cases of ventricular septal defect (VSD) or residual mitral or aortic valve damage, although there was 1 case (3.8%) of complete atroventricular block that required implantation of a dual-chamber pacemaker ten days after the procedure. Ninety percent of the patients developed postoperative left bundle-branch block. A 72-year-old woman with 117 mm Hg LVOTG, SAM, and grade 3 MR before surgery presented heart failure in the immediate postoperative period. Despite the apparently acceptable intraoperative result, a new echocardiogram showed residual LVOTG of 62 mm Hg, with grade 4 MR, and grade 3 SAM; therefore the patient was reoperated three days after the myectomy for implantation of a mitral prosthesis. The patient’s subsequent progress was good, and she maintained NYHA functional class II with a residual LVOTG of 23 mm Hg. In-hospital mortality was 3.8%, as one patient died 7 days after the surgery because of malignant ventricular arrhythmia, believed to be secondary to hyperpotassemia due to postoperative renal failure. Actuarial survival at 5 years was 96%±4%.

DISCUSSION

Hypertrophic cardiomyopathy is a complex disease due to its morphological, functional and clinical heterogeneity and affects approximately 1 out of every 500 individuals. Only 25% of patients present significant hemodynamic obstruction of the left ventricular outflow tract. Of these, most respond adequately to medical treatment. Negative inotropic drugs, such as beta-blockers, disopyramide, or verapamil, tend to improve the symptoms, prolong exercise time and decrease LVOTG. However, in 5% of patients with HOCM, the symptoms are refractory to drug therapy and invasive treatment must be considered.

Pathophysiological Basis for Current Surgical Treatment

Since the first procedures were performed in the mid-20th century, HOCM surgery has developed in accordance with the changing pathophysiological concepts of the disease. At first, it was thought that the obstruction was due to the existence of a subaortic sphincter muscle, as occurs in right ventricular infundibular obstruction, and therefore, surgery consisted of septal myotomy. Residual LVOTG was

### Table 1. Comparison of Pre- and Postoperative Clinical and Echocardiographic Data

<table>
<thead>
<tr>
<th>NYHA grade</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>I-II</td>
<td>9</td>
<td>25</td>
<td>&lt;.010</td>
</tr>
<tr>
<td>III-IV</td>
<td>17</td>
<td>0</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Mean</td>
<td>2.8±0.9</td>
<td>1.2±0.4</td>
<td>&lt;.001</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Septal thickness, mm</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>24±5</td>
<td>13±4</td>
<td>&lt;.001</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>LVOTG, mm Hg</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>96.5±26.0</td>
<td>19.5±6.50</td>
<td>&lt;.001</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>MR grade</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-1, n</td>
<td>0</td>
<td>20</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>2, n</td>
<td>12</td>
<td>5</td>
<td>&lt;.010</td>
</tr>
<tr>
<td>3-4, n</td>
<td>14</td>
<td>0</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Mean</td>
<td>2.54±0.51</td>
<td>0.69±0.79</td>
<td>&lt;.001</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SAM grade</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-1, n</td>
<td>1</td>
<td>25</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>3, n</td>
<td>25</td>
<td>0</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Mean</td>
<td>2.92±0.39</td>
<td>0.23±0.43</td>
<td>&lt;.001</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>LVEF, %</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>67.5±8.90</td>
<td>65.7±8.4</td>
<td>NS</td>
<td></td>
</tr>
</tbody>
</table>

*LVEF indicates left ventricular ejection fraction; LVOTG, left ventricle outflow tract gradient; MR, mitral regurgitation; NYHA, New York Heart Association; SAM, systolic anterior motion.*
significant, and in-hospital mortality high, leading to abandonment of the technique. In 1968, Morrow described the “subaortic myectomy,” reporting good clinical and hemodynamic results.\textsuperscript{6} Resection of the subaortic portion of the septum increased the diameter of the ventricular outflow tract, decreasing the blood flow velocity through the tract and thus the attraction force of the anterior mitral valve leaflet due to a Venturi effect, considered to be the main cause of SAM. It is now known that the Venturi effect plays a much less relevant role in the pathophysiology of the disease than was formerly believed. Anterior leaflet pull caused by flow abnormally aimed at the posterior wall because of septal hypertrophy is currently considered to be the main cause of SAM. The procedure of choice to be extended septal myectomy associated with anterior mitral leaflet pllication.

Extended myectomy\textsuperscript{9-11} involves prolongation of the septal resection to the insertion site of the papillary muscles without affecting the subaortic septum. Resection of this structure, which is relatively unininvolved in the pathophysiology of the disease, has a high risk of complications and does not adequately redirect the flow. In contrast, resection of the mid- to distal portion of the septum causes flow to be directed toward the anterior wall without catching the anterior mitral leaflet, thus eliminating SAM and decreasing the LVOTG. The technique is difficult because of the surgeon’s limited vision of the surgical field. Inadequate resection causes the obstruction to persist, whereas excessive resection has a risk of complications. To facilitate extensive, but safe resection, our group proposed video-assisted myectomy to be performed with a cardioscope, allowing resection of the fibromuscular septal band at the insertion site of the papillary muscles, without damaging the mitral subvalvular apparatus.\textsuperscript{12}

The presence of anatomical alterations in the mitral valve of patients with HOCM has been well documented.\textsuperscript{13,14} The leaflets are usually larger than normal and the papillary muscles take on a more anterior position in the ventricular cavity. Anterior mitral leaflet pllication\textsuperscript{15,16} is intended to decrease the size and slackness of the leaflet. This reduces the surface exposed to the flow and its dragging toward the posterior wall, which leads to SAM. We plicate along the longitudinal axis in the fibrotic area of the valve that comes in contact with the septum, usually corresponding to the middle segment (A2). However, a recent article on 3-dimensional echocardiography has suggested that the anterior field (A1) is the portion most highly implicated in SAM.\textsuperscript{17} Several groups perform horizontal pllication\textsuperscript{9} and combine it with mobilization and partial excision of the papillary muscles.\textsuperscript{14} Generalized use of this technique in all patients may not be advisable, but it could be beneficial in those with SAM, MR and large and redundant leaflets, in whom myectomy alone results in 11% of severe residual MR.\textsuperscript{18}

Valve replacement\textsuperscript{19} is perhaps the most definitive treatment of HOCM, as it obtains the largest decrease in LVOTG and eliminates MR. Unlike other diseases involving the mitral valve, the entire valve and subvalvular apparatus, including the papillary muscles, must be resected.\textsuperscript{20} If the anterior leaflet is eliminated, there is no possibility of SAM. However, it is an option that should be used only in selected cases, as there is risk of complications from the prostheses. This strategy is indicated for the following: presence of structural abnormalities of the mitral valve (prolapse, calcification, endocarditis); recurrence after prior myectomy; mid-ventricular obstruction and rare cases of refractory HOCM with SAM and gradient but with septal bulge less than 18 mm, where myectomy may have a high risk of residual VSD.\textsuperscript{21} In our series, the most frequent reasons for valve replacement were valvular destruction from endocarditis and emergency situations, where an expert echocardiographer was not present to ensure adequate repair.

Clinical-Hemodynamic Benefit and Morbidity and Mortality of the Surgery

In our experience, surgery led to a highly significant reduction in LVOTG, MR and SAM, allowing symptomatic improvement in 96% of the patients and complete resolution of the symptoms in 77%. An 80% reduction in LVOTG was achieved and postoperative LVOTG at rest (19.5 mm Hg) was at the upper limit of the level achieved in the best international series (4.5-16 mm Hg).\textsuperscript{9,11,22} The incidence of complications was low. The fact that there was no VSD or iatrogenic aortic regurgitation, and the prevalence of complete bundle-branch block was less than 4% must be related to preservation of the 5 mm of septum immediately below the aortic ring and the right coronary-noncoronary commissure. Postoperative left bundle-branch block was quite frequent, but did not produce deterioration of ventricular contractility and in fact may be considered a sign of sufficiently extensive septal resection.\textsuperscript{23} The appearance of residual VSD is rare in recent series (0%-2%) and the need for a pacemaker due to atrioventricular block is 0% to 10%. The in-hospital mortality of myectomy is 0%-6%, with 5- to 10-year survival of 85%-93% and 70%-88%, respectively.\textsuperscript{9,11,22} Early and late mortality are highest when myectomy is as-

754 Rev Esp Cardiol 2004;57(8):751-6

Castedo E, et al. Surgical Treatment for Hypertrophic Obstructive Cardiomyopathy
associated with coronary revascularization or valve replacement. In our study, 1-month mortality (3.8%) and 5-year survival (96%) are good, particularly because 8% of the cases were emergencies and another 8%, combined surgery. Although no invasive therapy has been shown to reduce the incidence of sudden death in HOCM, we consider it highly significant that no sudden cardiac death occurred after a mean follow-up of 5 years.

Surgical Candidates

One of the main advantages of surgery over other invasive strategies is the possibility to treat not only the myocardial component (septal hypertrophy) of the disease, but also the valvular (mitral) component, thereby correcting the mitral regurgitation and SAM. In our series, there was little or no postoperative MR in 77% of the cases and SAM was eliminated in 100%. Percutaneous transluminal septal myocardial ablation does not appear to have any corrective effect on MR; therefore, we believe that patients with significant MR and SAM and those with structural abnormalities of the mitral valve or subvalvular apparatus should undergo surgery. Likewise, patients with associated heart disease and younger patients, who tend to have a worse response to ablation due to a larger septal bulge and frequent structural abnormalities of the papillary muscles, may also be better surgical candidates.

CONCLUSIONS AND CLINICAL IMPLICATIONS

Surgery in patients with HOCM produces excellent correction of LVOTG, MR, and SAM and leads to a significant clinical improvement in functional capacity. Whenever possible, extended septal myectomy plus anterior mitral leaflet plication is the technique of choice. If performed at a hospital with experience, the surgery can be done with low morbidity and mortality, although this option should always be weighed against the patient’s symptoms and severity of hemodynamic obstruction.

REFERENCES

24. Siegmull JL, Maron BJ, Permut LC, McIntosh CL, Clark R.