The surgical management of hypertrophic obstructive cardiomyopathy with the concomitant mitral valve abnormalities

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Abstract

OBJECTIVES: The purpose of this retrospective study was to analyse the pathogenesis and the treatment strategies of hypertrophic obstructive cardiomyopathy (HOCM) with the concomitant mitral valve abnormalities.

METHODS: Between October 1996 and December 2009, 76 patients with the HOCM underwent the ventricular septal myotomy–myectomy in Beijing Fuwai hospital. There were 51 males and 25 females aged between 6 and 68 years (mean: 37.18 ± 15.85 years) old. All the patients had left ventricular outflow tract (LVOT) obstruction with a resting or physically provoked gradient of ≥50 mmHg and the systolic anterior movement (SAM) of the mitral leaflets, and 64 patients had mitral regurgitation (MR). These patients underwent the ventricular septal myotomy–myectomy under general anaesthesia and cardiopulmonary bypass. The concomitant surgical procedures included mitral valve replacement (MVR, n = 14) and mitral valve plasty (MVP, n = 12).

RESULTS: All the surgical procedures were technically successful. In comparison with the preoperative conditions, the resting LVOT gradient had marked reduction (99.73 ± 38.61–23.55 ± 16.53 mmHg, P < 0.001), the mean septal thickness was decreased from 26.23 ± 5.24 to 17.33 ± 4.74 mm. MR had significant improvement, SAM was resolved completely or only mild. Four patients (5.3%, 4/76) died during the hospital stay. The causes of death included severe ventricular arrhythmias with low cardiac output, severe acute renal failure, septic shock with acute renal dysfunction and the complete AV block with low cardiac output. The others were followed up for 5–18 years: there were no deaths. Moderate MR was noted in two patients at 2 months or 2 years after operation respectively, who had undergone MVP with the edge-to-edge technique stitch procedure, and only had mild or trivial MR at hospital discharge, of whom one received repeat operation with MVR and the other is still in follow-up. All surviving patients were evaluated as New York Heart Association Functional class I or II, and had a significant increase in physical capacity and a significant reduction in disabling symptoms.

CONCLUSIONS: The ventricular septal myotomy–myectomy can be performed successfully for the severe obstructive HOCM and MR with the low morbidity and mortality and excellent survival in the great majority of patients. But for the few patients with the intrinsic mitral valve disease, the concomitant MVP or MVR may be required, and MVR should be performed only as a priority choice for the inherent risks of prosthetic valves and anticoagulation therapy.

Keywords: Hypertrophic obstructive cardiomyopathy • Systolic anterior movement of the mitral valve • Mitral regurgitation • The ventricular septal myotomy–myectomy • Surgical treatment

INTRODUCTION

Hypertrophic obstructive cardiomyopathy (HOCM) is a primary genetic myocardial disease characterized by asymmetric hypertrophy of the interventricular septum and variable degrees of dynamic left ventricular outflow tract obstruction (LVOTO) due to the systolic anterior motion (SAM) of the anterior mitral leaflet. SAM is also responsible for the concomitant mitral regurgitation (MR), which is typically directed posterolaterally into the left atrium. The transaortic left ventricular septal myectomy has been proposed as the gold standard surgical procedure for symptomatic patients with severe HOCM since the late 1960s [1]. Between October 1996 and December 2009, 76 consecutive patients with HOCM and concomitant MR received the transaortic ventricular septal myotomy–myectomy in Beijing Fuwai hospital. In this paper, we analyze the pathogenesis and the treatment strategies on these patients.
MATERIALS AND METHODS

Patient population

Between October 1996 and December 2009, 76 consecutive patients with HOCM received the transaortic ventricular septal myotomy–myectomy. There were 51 males and 25 females. The patients’ ages were from 6 to 68 years (mean: 37.18 ± 15.85 years) old with a body weight range of 27–94 kg (mean: 60.37 ± 13.56 kg). All the patients were suffering from disabling symptoms (e.g. chest discomfort, dyspnoea, syncope or palpitations) and a significant reduction in physical capacity, despite having received the optimal medical therapy with β-blocker, calcium channel blockers or both.

All the patients were diagnosed preoperatively by transthoracic echocardiography and/or magnetic resonance imaging (MRI). Asymmetric hypertrophy of the interventricular septum and SAM of the anterior mitral leaflets resulted in the dynamic LVOT obstruction and the concomitant MR. A resting or physiologically provoked LVOT pressure gradients was greater than or equal to 50 mmHg. Severity of MR level was classified as follows: trivial (n = 7), mild (n = 19), moderate (n = 30) and severe (n = 8). Abnormalities of mitral valve included: mitral valve prolapse (n = 10); mitral leaflet thickening or obvious mitral annular calcification as a result of rheumatic disease (n = 8); leaflet destruction due to endocarditis (n = 4); abnormal papillary muscle directly attached to the anterior mitral leaflet (n = 3); excessive elongation of the leaflet (n = 1). The major associated malformations included: stenotic atherosclerotic coronary artery disease (n = 3); myocardial bridge (n = 9); aortic stenosis (n = 4); patent ductus arteriosus (n = 2); severe arrhythmias (n = 9); tricuspid regurgitation (n = 3). Four patients had undergone the failed percutaneous transluminal alcohol septal ablation and 1 patient had implanted the permanent dual-chamber pacemakers.

Surgical management

The classical ventricular septal myotomy–myectomy, known as the Morrow procedure, was performed under general anaesthesia and cardiopulmonary bypass (CPB) with the moderate systemic temperature and low-volume blood flow. The prebypass transoesophageal echocardiography (TEE) was performed for reassessing the extent and level of LVOTO, abnormalities of the mitral valve and septal thickness. The subaortic septum was exposed by a low transverse aortotomy carried out rightward towards the non-coronary aortic sinus. The classical Morrow procedure was performed to resect the hypertrophic interventricular septal from 2 to 3 mm rightward to the midpoint of the right coronary sinus, extending 10–12 mm towards the left coronary sinus and typically the resection was enlarged to the bottom of the papillary muscle of anterior mitral leaflet in order to open the LVOT thoroughly and eliminate SAM completely. The adequacy of the resection was evaluated by the direct visual inspection through the incision of the aortic root; we could see the papillary muscles’ base after myectomy. Extended septal myectomy, compared with the original Morrow procedure, was a much more extensive resection. For some patients, the extent of resection could be distally beyond the level of the mitral anterior papillary muscles towards the apex, the hypertrophic septal muscle below the membranous septum also could be resected (Figs 1 and 2). The intraoperative TEE evaluation was performed immediately after the patients were weaned from CPB. Fourteen patients had received concomitant mitral valve replacement (MVR) and 12 patients had received mitral valve plasty (MVP). The techniques of mitral valve reconstruction included: edge-to-edge technique (n = 7); radical debridement and repositioning of the papillary muscles (n = 2); plication plasty of the posterior leaflet (n = 1); plication plasty of the anterior leaflet (n = 1); insertion of the annuloplasty ring (n = 1).

Concomitant operative procedures performed in 18 patients included: coronary artery bypass surgery (n = 7); aortic valve replacement (n = 5); tricuspid valve plasty (n = 3); patent ductus arteriosus closure (n = 2); resection of additional discrete membranous subaortic stenosis (n = 1).

Data analysis

Demographic and other patient-related data were obtained from clinical records. The values, unless stated otherwise, are expressed as mean ± standard deviation. Student’s paired t-tests were chosen for the differences between two groups. The level of statistical significance was set at P < 0.05. Early operative mortality was defined as death occurring during the hospitalization, or within 30 days postoperatively.

RESULTS

All the surgical procedures were technically successful. The mean CPB and the aortic blocking time were 90.62 ± 48.53 (25–290) and
was 9.07 ± 3.32 days (6–183 h), and mean hospitalization stay was 9.07 ± 3.32 days (6–23 days).

In comparison with the preoperative conditions, after myectomy, the intraoperative TEE and postoperative transthoracic echocardiography demonstrated marked reduction of the LVOT pressure gradients and the mean septal thickness and significant improvement in MR. In the patients who underwent myectomy alone, 4 needed further resection or extended septal myectomy for a high LVOT gradients and various degrees of SAM by intraoperative TEE after the initial resection and SAM disappeared completely in 67 patients, only 9 still had mild SAM, but had no mitral–septal contact and needed no further operation. In the patients who underwent myectomy with concomitant MVR, 4 underwent second run with MVR after failed MVP, all patients had adequate relief of LVOTO, MR and SAM disappeared completely. And for those who underwent myectomy with concomitant MVP, of whom 1 patient, though adequate relief of LVOTO by adequate myectomy, underwent second run with MVP for moderate MR by intraoperative TEE, LVOTO relieved and SAM disappeared completely in all patients, only 4 still had mild MR, but need no further operation (Tables 1–3).

### Table 1: Changes in haemodynamic parameters pre- and postoperatively

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Preop</th>
<th>Postop</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diameter of LA (mm)</td>
<td>42.40 ± 7.72</td>
<td>35.26 ± 8.45</td>
<td>0.023</td>
</tr>
<tr>
<td>LVED (mm)</td>
<td>42.74 ± 6.75</td>
<td>40.41 ± 5.79</td>
<td>0.728</td>
</tr>
<tr>
<td>LVOT gradients</td>
<td>99.73 ± 38.61</td>
<td>23.55 ± 16.53</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>The septal thickness (mm)</td>
<td>26.23 ± 5.24</td>
<td>17.33 ± 4.74</td>
<td>0.015</td>
</tr>
<tr>
<td>Ejection fraction (%)</td>
<td>70.45 ± 9.31</td>
<td>62.68 ± 8.72</td>
<td>0.032</td>
</tr>
</tbody>
</table>

Values are expressed as mean ± SD, P < 0.05 was considered statistically significant.

LA: left atrial; LVED: left ventricle end-diastolic diameter; LVOT: left ventricular outflow tract; SD: standard deviation.

### Table 3: Changes in SAM level in groups postoperatively (cases)

<table>
<thead>
<tr>
<th>Group</th>
<th>Period</th>
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<th>Trivial</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isolated myectomy</td>
<td>Preop</td>
<td>12</td>
<td>7</td>
<td>15</td>
<td>16</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Postop</td>
<td>31</td>
<td>10</td>
<td>9</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Myectomy + MVR</td>
<td>Preop</td>
<td>0</td>
<td>0</td>
<td>10</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Postop</td>
<td>14</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Myectomy + MVP</td>
<td>Preop</td>
<td>0</td>
<td>0</td>
<td>4</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Postop</td>
<td>7</td>
<td>1</td>
<td>4</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

MR: mitral regurgitation; MVR: mitral valve replacement; MVP: mitral valve plasty.

### Table 2: Changes in MR level in groups pre- and postoperatively (cases)

<table>
<thead>
<tr>
<th>Group</th>
<th>Period</th>
<th>None</th>
<th>Trivial</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
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</thead>
<tbody>
<tr>
<td>Isolated myectomy</td>
<td>Preop</td>
<td>12</td>
<td>7</td>
<td>15</td>
<td>16</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>Postop</td>
<td>31</td>
<td>10</td>
<td>9</td>
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<tr>
<td>Myectomy + MVR</td>
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<td>14</td>
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<tr>
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<td>4</td>
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<td>0</td>
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<tr>
<td></td>
<td>Postop</td>
<td>7</td>
<td>1</td>
<td>4</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

Four patients (5.3%, 4/76) died during the hospital stay. The causes of death included: severe ventricular arrhythmias with low cardiac output (n = 1); severe acute renal failure (n = 1); septic shock with acute renal dysfunction (n = 1); the complete AV block with low cardiac output (n = 1).

The major early complications included: the left bundle branch block (n = 40); intraventricular conduction block (n = 8); complete AV block (n = 7); left anterior hemiblock (n = 7); type I atrioventricular conduction block (n = 5); atrial fibrillation (n = 7); permanent pacemaker implantation for the complete AV block (n = 4).

During the 5–18 years of follow-up (subsequent clinical visits n = 59 and telephone interviews n = 8), 5 patients were lost to follow-up. There was no death or lethal arrhythmia. Moderate MR was noted in 2 patients at 2 months or 2 years after operation respectively, who had undergone MVP with the edge-to-edge technique stitch procedure, and only had mild or trivial MR at hospital discharge, of whom one received repeat operation with MVR and the other is still in follow-up. All surviving patients were evaluated as New York Heart Association Functional class I or II, and had a significant increase in physical capacity and a significant reduction in disabling symptoms.

### DISCUSSION

HOCM, also termed as idiopathic hypertrophic subaortic stenosis, is a primary genetic myocardial disease characterized by the asymmetric left interventricular septum hypertrophy and SAM. SAM is also responsible for concomitant MR, which is due to the diastolic dysfunction of left ventricular. The transaortic left ventricular septal myectomy remains the gold standard therapeutic option to relieve or alleviate dynamic LVOTO. SAM and associated MR, thus improving functional status and survival in symptomatic patients with HOCM [2, 3].

The septal myectomy for HOCM significantly depends on the degree of persistent disabling clinical symptoms despite optimal medical therapy, LVOT pressure gradient greater than or equal to 50 mmHg under resting conditions, or reaching 100 mmHg under provocation. For adolescents or children with HOCM without obvious symptoms, surgical indication requires that LVOT pressure gradients should be greater than or equal to 75 mmHg [4, 5].

The systolic mitral anterior valve anterior displacement (SAM) is caused by a Venturi, or drag forces, which is initiated by rapid LV ejection through a narrow outflow tract caused by the hypertrophic septal muscle and anterior displacement of the mitral valve apparatus. Mismatching of length and mobility variations of
the posterior and anterior mitral leaflet may be the factor that restricts the posterior leaflet to move along with the anterior leaflet effectively, causing dynamic LVOTO and a fissure between the two leaflets, which may explain concomitant MR during middle–late phase of the systole period. For the great majority of these patients, the successful surgical myectomy alone is adequate to eliminate MR and SAM, without requirement for additional mitral valve surgery [6–10]. A minority of patients still have various degrees of MR or SAM after myectomy, which may result from limited resection of the hypertrophic septal muscle at the initial myectomy. Resection of the hypertrophic myocardium at the midventricular part may be too limited and failed to reach the bottom of the papillary muscle of the anterior mitral leaflet, which may cause the residual variable degrees of dynamic LVOTO during systole; these patients require repeated myectomy or extended septal myectomy. Residual traumatic leaflet thickening and minor degrees of mitral annular calcification also can be explained for residual trivial or mild MR after myectomy [11–14].

Few patients, in spite of adequate relief of LVOTO by adequate myectomy, still have variable degrees of residual MR, not due to SAM but due to intrinsic mitral valve disease that includes mitral valve prolapse; chordal rupture; fibrosis of the mitral valve leaflet; abnormal papillary muscle of the anterior mitral leaflet; notable mitral annular calcification; excessive elongation of the leaflet; congenital abnormalities; hurt of mitral leaflet or chordae during myectomy or leaflet destruction (e.g. myxomatous; degenerative; infective endocarditis; rheumatic disease). Under these conditions, successful myectomy has relieved LVOTO completely and resulted in obvious reduction of LVOT pressure gradients and disappearance of SAM, but residual MR is still obvious and usually direct anteriorly or centrally into the left atrium [15–20]. For these patients, concomitant MVP may be attempted. MVP techniques should be individualized and modified according to the mitral valve leaflets and sub-valvular apparatus abnormalities such as commissural annuloplasty; radical debridement and repositioning of the papillary muscles; leaflet resection of the posterior leaflet; insertion of artificial chordate; plication of the anterior leaflet or an edge-to-edge stitch. When an annuloplasty band or ring is used, it is important to select an adequate sized prosthesis to avoid the potential aggravating SAM postoperatively. Only for patients with infective endocarditis, severe rheumatic disease or complex congenital MV abnormalities, unsuitable for MVP or failure of MVP, MVR should be considered, for the disadvantages of the prosthesis, such as durability; endocarditis; bleeding; thromboembolism and anticoagulation. MVP should be performed only as a priority choice for the inherent risks of prosthetic valves and anticoagulation therapy.

Conflict of interest: none declared.

REFERENCES


