Surgical treatment of hypertrophic obstructive cardiomyopathy

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The rationale for surgical treatment of hypertrophic obstructive cardiomyopathy is based on the assumption that dynamical obstruction is mainly caused by a reduction in the left ventricular outflow tract cross-sectional area due to bulging septal myocardial tissue. In this sense, classical myotomy-myectomy is the 'gold standard' therapy for patients with severely symptomatic hypertrophic obstructive cardiomyopathy. In my own experience with extended myectomy, more than three-quarters of all long-term survivors are in functional class I or II (New York Heart Association) and overall survival after 18 years (mean follow up 8.1 years) was 68%, with a linearized mortality rate of 1.9% per patient-year. Notably, there was no case of sudden cardiac death during follow up, leading to the assumption that relief from dynamical obstruction is most effective in the prevention of sudden cardiac death in these patients.

Key Words: extended myectomy, hypertrophic obstructive cardiomyopathy, myotomy-myectomy, surgical therapy

Introduction

The complexity of the anatomical substrate that produces obstruction in hypertrophic obstructive cardiomyopathy (HOCM) is reflected by the variety of different surgical strategies that have been reported during the past 30 years[1–12]. These include incision (myotomy)[1] and excision (myectomy)[2,3] of bulging myocardial tissue from the basal septum, in some series together with plication[9] or additional enlargement[10] of the anterior leaflet of the mitral valve, and mitral valve replacement[5–7,12]. In most series left ventricular myotomy-myectomy proved to be effective in reducing left ventricular outflow tract (LVOT) gradients and symptoms of patients[13–17], and a transaortic approach is regularly preferred to left or right or combined ventriculotomies[4,8,18]. Left ventricular myotomy-myectomy may thus serve as a 'gold standard' modality, whereas the disadvantages of mitral valve replacement are estimated to outweigh its benefits[19].

However, surgeons still consider classical myotomy-myectomy to be a technically challenging operation, which is often referred to specialized and experienced centres because it carries inherent risks for ventricular septal defect and incomplete relief. We developed a modified technique[20,21] that enables better exposure of the basal septum, simplifies myectomy, and even allows the myectomy to be extended to the lateral free wall of the ventricle. This extended myectomy allows safe access to the deeper parts of the ventricle, where resection of hypertrophied trabeculae and mobilization or partial excision of papillary muscles lead to correction of the anatomically distorted subvalvular mitral apparatus.

Surgical technique (extended myectomy)

Full details regarding surgical methods are described elsewhere[20,21]. In brief, all patients are operated while on cardiopulmonary bypass with cardioplegic arrest and under moderate hypothermia (32°C). In case of aorto-coronary bypass grafting, coronary anastomoses are performed first and cardioplegic solution is reinfused via the bypass veins. All other concomitant surgical interventions are done after completion of extended myectomy and intraventricular repair.

A low transverse aortotomy and 2-0 stay sutures in the commissura of the aortic valve expose the basal ventricular septum. The extent of hypertrophy is then assessed by visual inspection and digital palpation. A sharp triple hook retractor is carefully inserted at the deepest point of the hypertrophied septum, thus defining...
myectomy is fixed and clearly defined. Longitudinal incisions are placed 2–3 mm underneath the aortic annulus in the direction of the prongs of the retractor, the first one at the deepest point of the right coronary cusp directly underneath the right coronary ostium and the second one at the transition to the left ventricular free wall. This excision can even be extended behind the insertion of the mural leaflet of the mitral valve. Both incisions are then joined with a transverse incision, thus completely removing the tissue grasped with the retractor. It is noteworthy that all relevant tissue of the basal septum may thus be removed en bloc, which minimizes the risk for septal perforation.

After creation of a large and deep LVOT trough, access to the deeper structures of the left ventricle is gained. Both papillary muscles are then completely mobilized, and all hypertrophied trabeculae, as well as hypertrophied parts of the papillary muscles, are resected. At this point of the operation, excellent vision of the surgical field is mandatory in order to ensure safe resection. In case of significant right ventricular outflow tract obstruction, this right ventricular tissue is excised through a right ventriculotomy that is relevant for the patients. All patients had significant systolic anterior motion of the mitral valve (SAM) with prolonged mitral valve–septal contact (+++ and ++++).

**Echocardiography**

Transthoracic two-dimensional echocardiography with colour flow imaging and pulsed Doppler tracings can be used to detect and semiquantitatively grade mitral regurgitation. Cardiac dimensions, including the degree of SAM, are easily measured from M-mode echocardiograms.

Three-dimensional echocardiography using a modified multiplane transoesophageal echocardiography probe (5 Mhz; Hewlett Packard, Andover, MA, U.S.A.) allows quantitative analysis of intraventricular structures.[22]

For measurement of the minimal cross-sectional area of the LVOT during systole, first the long axis of the LVOT is defined orthogonal to the aortic annulus. Then, in all short-axis cross-sections, the one with the smallest unobstructed cross-sectional area is identified. The boundaries of the LVOT can be traced manually in order to calculate the corresponding area, and this procedure can be repeated during postoperative analysis. For qualitative comparison with the preoperative situation, the corresponding cross-sections are reconstructed in the preoperative data-set, using anatomical landmarks for orientation to find the same cross-section (i.e. distance to aortic ring and to the left ventricular apex).

At late follow-up in our series, the wall thickness of the ventricular septum was impressively low (13.4 ± 4 mm) and was significantly different from that identified in the preoperative evaluation (25 ± 5 mm; *P* < 0.005). Measurements of left ventricular end-diastolic diameters (46 ± 6 mm) and end-systolic diameters (32 ± 8 mm) revealed near to normal left ventricular cavity dimensions. None of the patients showed relevant SAM, although some patients had a trivial degree of SAM (1+). Doppler studies in all patients revealed no flow acceleration at the mid cavity level or within the LVOT. No significant mitral regurgitation was detectable, and no aortic regurgitation occurred. Quantitative analysis of the three-dimensional data-sets showed that minimal cross-sectional area of the LVOT increased from 1.1 ± 1.0 cm² to 4.4 ± 2.7 cm² (*P* < 0.001).

**Early and long-term results**

The overall 30-day operation mortality rate was 1.2%. Perioperative non-fatal complications included one transient cerebral attack with full recovery during hospital stay. No heparin anticoagulation was given regularly during hospital stay. Anticoagulation was restricted to those patients with coronary artery bypass grafting and one patient with aortic valve replacement. Of the patients 3% required permanent pacemaker therapy because of total atrioventricular block after surgery; two already had right bundle branch block before surgery. No ventricular septal perforation occurred.

**Patient selection criteria**

The present patient cohort (n = 82) was operated on for treatment of HOCM at the Department of Thoracic and Cardiovascular Surgery in Aachen, Germany. All patients were symptomatic despite adequate medical treatment with beta-blocking agents or calcium channel blockers, or both; their mean functional class (New York Heart Association [NYHA]) was 3–2. The most common symptoms in these patients were dyspnoea (84%) and angina (58%); 48% of the patients reported frequent attacks of dizziness and vertigo; and 28% had one or more episodes of syncope. Most of the patients (95%) were in sinus rhythm at the time of operation, two patients had atrial fibrillation, and one patient had had a permanent pacemaker implanted before surgery because of total atrioventricular block. Right or left bundle branch block was present in 15% of the patients. The mean duration of cardiac symptoms was 10.3 ± 6 years, and the mean time interval between diagnosis of HOCM and operation was 3.1 ± 3 years.

Preoperative diagnostic procedures include left and right heart catheterization and transthoracic echocardiographic evaluation. The indication for surgery was restricted to those patients with maximal LVOT gradient greater than 50 mmHg under basal conditions or greater than 80 mmHg with provocative manoeuvres such as amyl nitrate inhalation, isoproterenol infusion, valsalva manoeuvre or following extrasystolic potentiation. In our series invasively measured LVOT peak systolic gradients were 85 ± 36 mmHg at rest and 156 ± 51 mmHg with provocative manoeuvres. Preoperative echocardiographic studies showed moderate mitral regurgitation in 27% and severe mitral regurgitation in 5% of the patients. All patients had significant systolic anterior motion of the mitral valve (SAM) with prolonged mitral valve–septal contact (+++ and ++++).
The overall linearized mortality rate was 1.9%/patient-year, 5-year survival rate was 94% and 10-year rate was 86%. The cumulative survival, calculated using the Kaplan–Meier method, is given in Fig. 1. The cause of death was mainly progressive congestive heart failure (n = 6); three of these six patients were initially in functional class IV (NYHA). One patient, who was initially treated with concomitant coronary artery bypass grafting, died nearly 10 years after surgery as a result of myocardial infarction. It is noteworthy that none of the cardiac- or non-cardiac-related deaths appeared suddenly, with the exception of one patient aged 76 years who died 10.5 years after surgery; the cause of death in that patient is unknown. The other deaths were caused by malignancy, multiorgan failure following abdominal surgery and respiratory insufficiency. Details are provided in Table 1.

Independent predictors of late mortality were age at operation \((P < 0.005)\) and preoperative LVOT gradients under basal conditions and under provocation \((P < 0.005)\). Neither haemodynamic parameters, such as left ventricular end-diastolic pressure and pulmonary artery mean or wedge pressure, nor the sex of the patients were significant predictors of late mortality. The significant influence of age of the patient at the time of surgery on late mortality is demonstrated in the survival ratios of two subgroups of patients aged above or below 55 years (Fig. 2). Both actuarial survival rates are significantly different by log-rank test \((P = 0.005)\), but the linearized mortality rates over the first 10 years exhibited no difference.

Improvement in clinical symptoms, especially in the extent of dyspnoea, was the most prominent finding in all patients, and at initial follow up 93% of the patients were in functional class I or II (NYHA). Of all patients 46% improved by one class, 47% by two classes and 3% improved by three classes. At long-term follow-up 80% of all long-term survivors were in functional class I or II (NYHA). Improvement by one or two classes was reported by 84% of the long-term survivors. Among those patients angina and chest pain were apparent in 43% and symptoms of dizziness and vertigo were reported by 30% of the patients; these symptoms were consistently estimated to be of much lesser severity than before the operation.

Of all patients 77% were in sinus rhythm and 14% had a permanent pacemaker implanted (seven patients during follow up, three patients directly after the operation, and one patient had had his pacemaker implanted preoperatively). Twenty-two per cent of the patients had paroxysmal atrial fibrillation, and only 3.5% had chronic atrial fibrillation. No episodes of ventricular tachycardia were documented early or late, with the exception of one patient with preoperative malignant ventricular tachycardia and implanted defibrillator. During a follow-up interval of 46 months that patient had one episode of ventricular tachycardia with a rate greater than 200 beats \(\text{min}^{-1}\), which was successfully terminated by the device. Clinically, the patient reported no other symptoms and was in functional class II (NYHA).

### Table 1 Characteristics and causes of late deaths

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at death (years)</th>
<th>Sex</th>
<th>Duration of follow-up (months)</th>
<th>Preop. LVOT gradient (mmHg)</th>
<th>Preop. LVEDP (mmHg)</th>
<th>Preop. NYHA (class)</th>
<th>Postop. NYHA (class)</th>
<th>Type of surgery</th>
<th>Cause of death</th>
</tr>
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<tbody>
<tr>
<td>1</td>
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<td>Female</td>
<td>29</td>
<td>185</td>
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<td>II</td>
<td>EM/IVR</td>
<td>CHF</td>
</tr>
<tr>
<td>2</td>
<td>62</td>
<td>Male</td>
<td>128</td>
<td>75</td>
<td>15</td>
<td>IV</td>
<td>II</td>
<td>EM/IVR; MR</td>
<td>CHF</td>
</tr>
<tr>
<td>3</td>
<td>64</td>
<td>Male</td>
<td>69</td>
<td>80</td>
<td>24</td>
<td>IV</td>
<td>II</td>
<td>EM/IVR; CABG</td>
<td>CHF</td>
</tr>
<tr>
<td>4</td>
<td>72</td>
<td>Female</td>
<td>166</td>
<td>79</td>
<td>12</td>
<td>IV</td>
<td>III</td>
<td>EM/IVR</td>
<td>MOF</td>
</tr>
<tr>
<td>5</td>
<td>54</td>
<td>Female</td>
<td>90</td>
<td>76</td>
<td>29</td>
<td>IV</td>
<td>III</td>
<td>EM/IVR</td>
<td>CHF</td>
</tr>
<tr>
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<td>54</td>
<td>Female</td>
<td>2</td>
<td>173</td>
<td>40</td>
<td>III</td>
<td>II</td>
<td>EM/IVR</td>
<td>Respiratory insufficiency</td>
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<td>MI</td>
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<td>II</td>
<td>EM/IVR; CABG</td>
<td>Malignancy</td>
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<tr>
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<td>21</td>
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<td>II</td>
<td>EM/IVR</td>
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</table>

Preop.=preoperative; Postop.=postoperative; CABG=coronary artery bypass grafting; CHF=congestive heart failure; EM/IVR=extended myectomy/intraventricular repair; LVEDP=left ventricular end-diastolic pressure; LVOT=left ventricular outflow tract; MI=myocardial infarction; MOF=multiorgan failure; MR=mitral repair; NYHA=New York Heart Association.
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Figure 2  Plots of actuarial (percentage) survival among patients younger than 55 years (upper line) and among those older than 55 years (lower line). The numbers shown indicate the patients at risk (i.e. the numbers of patients followed up during the preceding time period) for both groups. There is a significant difference between the groups, as measured using the log-rank test (\( P = 0.005 \)), but there is no difference in the first 10 years of follow up.

Conclusion

HOCM is a primary cardiac disease that may involve a localized basal asymmetrical left ventricular hypertrophy, enlargement of the anterior mitral valve leaflet or malformation of the subvalvular mitral apparatus, or a combination of these[23,24]. The pathophysiological mechanisms that lead to dynamic obstruction are complex and still not completely understood, in particular because morphological findings may vary significantly among patients with comparable LVOT gradients. Moreover, in the same patient LVOT obstruction is not constant, and is markedly influenced by changes in pre-load and after-load, contractility, heart rate and rhythm. Hampered left ventricular performance with diastolic dysfunction, reduced cavity dimensions and a hyperdynamic contraction pattern are other typical findings in symptomatic patients with HOCM. However, the exact relationship between morphological changes, obstruction to left ventricular outflow and clinical symptoms is still a matter of debate. It is generally assumed that hypertrophic subaortic myocardial tissue, together with a SAM[25–27], represent the major anatomical obstacles in HOCM.

Surgical therapy, especially myotomy-myectomy with resection of septal tissue, has been shown in a number of large series[14,16,17,28,29] to relieve both LVOT gradients and symptoms of patients with HOCM. Early series using this technique were burdened with a high surgical mortality of 10–15%[13,15]. This rate is often cited by those advocating alternative procedures such as dual chamber pacing with right ventricular pre-excitation[30,31] or, most recently, induced infarction of the basal intraventricular septum[32]. Notably, however, perioperative risks for patients with severe hypertrophy appear to be significantly lower now than they were in the earlier series as a result of improved techniques of myocardial preservation and perioperative management. Indeed, early mortality rates with myotomy-myectomy are now well below 5%, and in specialized and experienced centres they are even less than 2%[16,17].

Thrus far, myotomy-myectomy is still the ‘gold standard’ surgical therapy in HOCM patients. Limitations of the classical technique are well recognized, however; it carries an inherent risk for creating ventricular septal perforation or vice versa resulting in incomplete relief. This risk is attributable to the fact that the tissue identified for resection cannot be exposed completely with a transaortic approach, and is therefore not clearly defined. According to the recommendations of Morrow et al.[33], exposure of resectable tissue is achieved by pushing on the right ventricle, thus revealing the septal tissue to the surgeon. However, only through experience will the surgeon be able to gauge the direction and depth of myectomy. Hence, ventricular septal defect may occur, which is reported to occur in the range 2–6%[13–17,23,28]. Incomplete relief, if it does not result in early mortality, may be associated with late survival, occurrence of sudden cardiac death or a rapid new onset of symptoms during follow up[23]. In our experience with the technique of extended myectomy, sustained relief was achieved in surgically treated HOCM patients without SAM or significant mitral regurgitation at long-term follow up, and no sudden cardiac deaths occurred during follow up. Moreover, no ventricular septal defect was created using this modified technique of septal myectomy.

Several reasons may account for these beneficial results. First, insertion of a sharp triple hook retractor simplifies myectomy, in that the amount of septal tissue that is suitable for resection is fixed and the direction in which incisions are to be performed are clearly defined. Second, extension of myectomy to the insertion of the mural leaflet of the mitral valve creates a trough nearly twice as wide as that with classical myectomy. In most cases, a toric muscle bundle between the lateral free wall and the mitral annulus is apparent, which is also resected by extended myectomy. Third, pulling the sharp triple hook retractor forward exposes septal tissue to the surgeon and extends myectomy down to the mid-portion of the left ventricle. In addition, our technique corrects not only the thickened septum, but also addresses the changes in papillary muscle morphology, which contribute significantly to the pathophysiology of HOCM. The latter warrants close attention because recent experimental[33] and clinical studies[34] have shown that the subvalvular mitral apparatus is involved in the pathological mechanisms of obstruction, especially in the presence of SAM. There is much evidence that malpositioned papillary muscles may interpose the mitral valve into the outflow stream, thus causing drag forces at the anterior leaflet and resulting in narrowing of the LVOT and mitral regurgitation. Onset of SAM at or even before aortic valve opening (i.e. with low outflow velocities) is another finding[35,36] that strongly suggests that factors other than Venturi forces contribute to outflow obstruction[37].

Transaortic reconstruction of subvalvular mitral apparatus following extended myectomy addresses the possibility that malpositioning of the papillary muscles is a significant patho-anatomical factor in HOCM. In our series, resection of hypertrophied trabeculae, and mobilization and partial excision of papillary muscles were performed in all
patients. Although it is not clear to what extent intraventricular repair may contribute to the sustained relief observed with this technique, its importance is demonstrated by the difference in LVOT cross-sectional areas before and after extended myectomy and reconstruction of the subvalvular mitral apparatus, as estimated using three-dimensional echocardiographic reconstruction\[22\].

Transaortic extended myectomy and reconstruction of subvalvular mitral apparatus has proven to be a highly effective therapy for patients with severely symptomatic HOCM\[38\]. However, these favourable results have not prompted extended indications for surgery. We still restrict surgical intervention to severely symptomatic patients with significant LVOT gradients under basal conditions, indications for coronary artery bypass grafting, or both. Continuous follow up will be necessary to confirm favourable results, but application of the surgical approach to obstruction presented here may nevertheless be recommended.

**References**


