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Long-Term Follow-up in Hypertrophic Obstructive Cardiomyopathy After Septal Myectomy

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Background. Controversy exists about the choice of treatment for patients with hypertrophic obstructive cardiomyopathy. The purpose of this study was to evaluate clinical and echocardiographic long-term results in patients with hypertrophic obstructive cardiomyopathy after septal myectomy and to determine predictors of event-free survival in these patients.

Methods. Between 1965 and 1995, 110 consecutive patients 2 to 66 years old (mean age, 37 ± 15 years) with an invasively measured left ventricular outflow tract gradient of 86 ± 39 mm Hg (81 ± 42 mm Hg by Doppler echocardiography) underwent either septal myectomy only ($n = 87$) or myectomy combined with additional procedures ($n = 23$). Mean follow-up was 11.7 ± 7.5 years. Predictors of late events were calculated using multivariate Cox regression analysis.

Results. The perioperative mortality rate was 3.6% ($n = 4$). The cumulative survival rate at 5, 10, and 15 years was

93%, 80%, and 72%, respectively, and symptom-free survival, 77%, 50%, and 33%, respectively. Predictors of late death were New York Heart Association class III or IV ($p < 0.05$), congestive heart failure ($p < 0.05$) and additional procedures ($p < 0.05$). The left ventricular outflow tract gradient was nearly eliminated in all patients, the left atrial dimension decreased significantly during the early years, and left ventricular dilatation occurred late in 17 patients.

Conclusions. Septal myectomy is associated with a low perioperative mortality and a high late survival rate (72% at 15 years' follow-up). Septal myectomy is still an excellent modality in the treatment strategy for symptomatic patients with hypertrophic obstructive cardiomyopathy.

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Hypertrophic obstructive cardiomyopathy (HOCM), comprising hypertrophied and abnormally oriented muscle cells (muscle fiber disarray), increased interstitial tissue, and dysplastic arteries, is inherited. Diastolic dysfunction and ischemic events cannot be prevented completely by any of the therapeutic modalities. Patients with no symptoms or only moderate symptoms are usually treated medically. However, for patients who have severe symptoms or a high risk of sudden death, alternative procedures are suggested. Of these procedures, septal myectomy, as precisely described by Morrow [1] in 1978, is the most established and has been performed for more than 30 years. Continuing improvements in the preoperative diagnosis of HOCM, together with modifications of the surgical technique, have led to low perioperative and postoperative mortality, disappearance of left ventricular outflow tract (LVOT) obstruction, and reliable relief of symptoms. In most institutions, septal myectomy remains the treatment of choice for severely symptomatic patients who are resistant to medical therapy and have a high LVOT gradient and for

patients who need additional surgical treatment such as bypass grafting.

In recent years, new therapeutic modalities such as dual-chambered pacing [2-4] and catheter-induced ablation of the interventricular septum by alcohol infusions [5] have been developed. The beneficial effect of apical preexcitation, achieved by right ventricular pacing, on LVOT pressure gradient was shown as early as 1967 by Hassenstein and associates [6]. Twenty years later, the technological advances in DDD pacing made this treatment available for many patients, who responded with a drop in the systolic pressure gradient of more than 30% [2-4]. Permanent apical preexcitation is obtained by shortening the atrioventricular conduction. Occasionally, catheter ablation of the atrioventricular node is necessary to achieve this goal [2]. Nonsurgical myectomy aims at selective destruction of the hypertrophied septum by injection of alcohol in the first or second major septal coronary branch. Currently this procedure is still experimental and cannot be recommended for a large number of patients [5].

With the introduction of these methods, it has become necessary to review the risks and benefits of septal myectomy. In addition, there is a need to define which patients will benefit most from surgical intervention. Therefore, preoperative and postoperative characteristics

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in patients operated on between 1965 and 1995 at this institution were analyzed retrospectively.

Patients and Methods

Between 1965 and 1995, 110 consecutive patients who had medically refractory HOCM or were at high risk for sudden death underwent septal myectomy at University Hospital, Zurich. Almost all patients had an LVOT gradient exceeding 50 mm Hg and a normal or higher than normal ejection fraction. In all patients in whom it was performed, echocardiography showed asymmetrical septal hypertrophy with systolic anterior motion of the anterior mitral valve leaflet. Patients with accompanying aortic valve stenosis, complex congenital heart disease, or congenital fibromuscular subvalvular aortic stenosis were excluded from the study. The annual operation rate remained constant over the study period.

There were 69 male and 41 female patients with a mean age of 37 ± 15 years (range, 2 to 66 years), including 12 children and 7 adults older than 60 years. Twenty-four patients (22%) had a family history of HOCM. Most patients were in New York Heart Association (NYHA) functional class III (average class, 2.54 ± 0.8). The mean duration of symptoms was 8.2 ± 6.8 years (range, 0 to 43 years). Exertional dyspnea was present in 62% of patients, angina in 37%, syncope in 24%, presyncope in 23%, congestive heart failure in 15%, and atrial fibrillation in 9%. Ten percent of the patients had experienced pulmonary edema, and 2 patients had been resuscitated from ventricular fibrillation. All adult patients had been treated with either β -blockers or calcium-channel antagonists; none had received amiodarone hydrochloride.

Preoperative hemodynamics were assessed by heart catheterization in 109 patients and by echocardiography in 64. The LVOT pressure gradient was measured at rest in all patients (mean gradient, 86 ± 39 mm Hg) and in 39 after a premature beat or with drug provocation (mean gradient, 119 ± 38 mm Hg) during heart catheterization. Left ventricular peak systolic pressure was 220 ± 51 mm Hg (maximal value, 380 mm Hg) and LV end-diastolic pressure, 22 ± 8 mm Hg. Mitral regurgitation was present in 50% of patients; it was moderate in 8% and severe in 4%. Structural alterations of the valve were found in 5 patients after aortic ($n = 2$) or mitral ($n = 3$) endocarditis. Two aortic valves were bicuspid, and two were thickened and calcified. Two patients had ruptured chordae tendineae. One patient had an aneurysm of the ascending aorta, and another, a subvalvular muscular pulmonary stenosis. Major coronary artery disease was found in 3 patients.

Surgical Technique

The surgical approach evolved during the study period. Most patients ($n = 66$) had operation through an aortic approach (from 1966 to 1995); 32, through a left ventricular approach (from 1965 to 1979); and 12, through a combined aortic and left ventricular approach (from 1970 to 1981).

Isolated septal myectomy was performed in 87 patients

(79%). Three patients also underwent aortocoronary bypass grafting. Two mitral valve replacements were performed because of severe regurgitation resulting from bacterial endocarditis, and ten mitral valve repairs were done for severe systolic anterior motion ($n = 7$), ruptured chordae tendineae ($n = 2$), and bacterial endocarditis ($n = 1$). Mitral valve repair consisted of a modified Kay-Wooler valvuloplasty, reattachment of chordae tendineae, and mobilization of the papillary muscles down to the apex with resection of all hypertrophied portions and muscular trabeculae [7]. The aortic valve was replaced because of thickened and calcified aortic cusps in 2 patients, severe destruction caused by endocarditis in 1, and annuloaortic ectasia with an aneurysm of the ascending aorta (composite graft) in 1. One iatrogenic aortic cusp lesion and one caused by endocarditis had to be repaired, two commissurotomies were performed, and one muscular subvalvular pulmonary stenosis was resected. Intraoperative transesophageal echocardiography (routinely performed since 1987) revealed a major residual LVOT gradient in 1 patient who had to undergo a more extensive myectomy on a second pump run.

Cardioprotection also improved during the study period. Initially all patients had operation under whole-body hypothermia and electrically induced ventricular fibrillation. Patients operated on between 1977 and 1988 received antegrade crystalloid cardioplegic solution and later, antegrade and retrograde blood cardioplegic solution. Today, isolated septal myectomy is performed under normothermic condition. The mean cardiopulmonary bypass time was 61 ± 19 minutes and the mean aortic cross-clamping time, 33 ± 16 minutes.

Follow-up

Clinical follow-up was obtained by review of patient records or questionnaires sent to the patient or the physician. Median follow-up was 10.6 years (mean follow-up, 11.7 ± 7.5 years; range, 0 to 30 years) with a cumulative total follow-up of 1,283 patient-years. Follow-up was complete on July 31, 1996, for 95 patients (86%).

Echocardiographic Evaluation

Preoperative and postoperative evaluation was performed by transthoracic echocardiography. The following echocardiographic techniques were used: since 1974, M-mode and since 1979, cross-sectional and M-mode. Doppler echocardiography was added in 1981. The systolic pressure gradient across the LVOT was assessed quantitatively by Doppler ultrasound. Currently all patients are examined before operation, early after operation, and late postoperatively, ie, with follow-up intervals of 2 to 3 years. A total of 356 echocardiographic studies were analyzed, preoperative studies from 64 patients and postoperative studies from 94.

Left ventricular dimensional measurements were performed at end-systole and end-diastole. They were carried out according to the conventions of the American Society of Echocardiography [8] and included standard-level septal wall thickness, posterior wall thickness at

end-diastole, and LV and left atrial internal dimensions at end-systole and end-diastole. Maximal end-diastolic thickness of the septal wall in the LVOT was also determined.

Left ventricular chamber dilatation has been defined as an increase of more than 2% per year in end-diastolic ventricular diameter combined with a decrease of 2% per year in midventricular systolic fractional shortening [9]. Because these criteria cannot be applied in the perioperative period, the incidence of left ventricular chamber dilatation was determined only in patients with repeated measurements during the early and late follow-up studies (n = 56).

Statistical Analysis

Actuarial survival and event-free survival were calculated using the Kaplan-Meier method. Predictors of death and event-free survival were calculated using multivariate Cox regression analysis. Echocardiographic results were compared by paired Student's *t* test. A *p* value of less than or equal to 0.05 was considered significant. Calculations were performed using SPSS for Windows 6.0.

Results

Early Mortality

The 30-day mortality rate was 3.6% (n = 4). The causes of death were intrapericardial bleeding resulting in cardiac tamponade in 1974, low-output failure, relative coronary insufficiency, and sudden cardiac death. All of these patients except the 1 with cardiac tamponade had additional surgical procedures, dyspnea (NYHA III or higher), or congestive heart failure.

Complications and Reinterventions

All cardiac reinterventions were related to the HOCM itself or to sequelae of the operation. Two patients underwent reoperation on the same day, 1 for a major LVOT gradient and 1 for an iatrogenic aortic cusp lesion. One small ventricular septal defect that had been noticed 3 days after myectomy had to be closed and a substantial residual LVOT gradient combined with persisting mitral valve regurgitation caused two reoperations, a ventricular septal defect patch closure and a modified Konno operation with replacement of the mitral valve, respectively. One patient with a major residual LVOT gradient and mitral regurgitation and 1 with major aortic stenosis (bicuspid leaflets) were treated by more extensive muscular resection and mitral or aortic valve repair. Only 1 patient had reoperation for late aortic valve regurgitation. One bioprosthesis had to be replaced because of failure. Two patients died as a result of reoperation; a child died after the third intervention and 1 patient had development of an intractable chylothorax. The interval between initial myectomy and reoperation ranged from several days to 17 years (mean interval, 6.0 ± 5.8 years).

Five patients had development of complete heart block

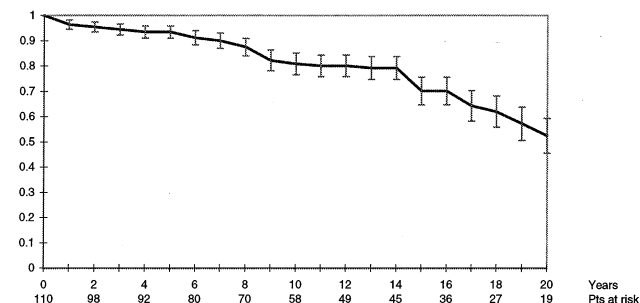


Fig 1. Actuarial survival of all patients. Error bars denote standard error.

as a result of septal myectomy. They were treated with a dual-chambered pacemaker.

Postoperative Medication

Before 1976, patients continued to receive β -blockers after operation. Since 1978, as previously described [10], all patients have been treated medically with verapamil hydrochloride on a long-term basis. Eighteen patients seen with signs of congestive heart failure were treated with diuretics and angiotensin-converting enzyme inhibitors for 10 ± 8 years after operation. Early postoperatively, patients were given warfarin sodium as prophylaxis against thromboemboli originating from the rough surface of the myectomy site. In the absence of other indications, anticoagulation was discontinued after 3 months.

Survival

The causes of late death were either cardiac or probable cardiac in 25 (83%) of 30 patients. Overall actuarial survival rates at 1 year, 5 years, 10 years, 15 years, and 20 years were 95%, 93%, 80%, 72%, and 53%, respectively (Fig 1). Univariate regression analysis detected the following risk factors: NYHA class III or IV, congestive heart failure, need of additional procedures, moderate to severe mitral regurgitation, history of pulmonary edema, duration of symptoms, and persistence of atrial fibrillation. Multivariate analysis identified NYHA class III or IV, congestive heart failure, and need of additional procedures as independent risk factors. Table 1 summarizes the results.

Figure 2 shows the survival curves of patients with none, one or two, and three of the independent risk factors for death. In patients with none of the risk factors, survival was excellent (ie, 98% at 5 years, 95% at 10 years, and 87% at 15 years).

Arrhythmias and Conduction Abnormalities

The incidence of persistent postoperative atrial fibrillation was, as expected, higher in the group with preoperative intermittent atrial fibrillation (5/10) than in the whole group. The onset of persistent atrial fibrillation was noticed in 31 patients after a sinus rhythm interval of 7.0 ± 5.5 years after operation. Ventricular rhythm dis-

Table 1. Preoperative and Operative Predictors of Premature Death After Septal Myectomy^a

Risk Factors for Premature Death	Univariate Cox Regression			Multivariate Cox Regression		
	Odds Ratio	<i>p</i> Value	95% Confidence Interval	Odds Ratio	<i>p</i> Value	95% Confidence Interval
NYHA class III or IV	5.45	0.001	2.19–13.56	3.01	0.05	771.07–8.49
Congestive heart failure	5.04	0.001	2.38–10.67	2.84	0.05	1.09–7.40
Additional procedures	2.93	0.005	1.41–6.10	3.12	0.05	1.26–7.74
Mitral valve regurgitation grade 3 or 4 (moderate to severe)	2.76	0.05	1.19–6.38
Pulmonary edema	3.52	0.01	1.44–8.61
Duration of symptoms	1.82	0.005	1.27–2.93
Atrial fibrillation	3.67	0.01	1.37–9.83

^a Odds ratio for duration of symptoms represents an increased risk for premature death every 10 years.

NYHA = New York Heart Association.

turbances were treated with amiodarone in 13 patients and an automatic cardioverter defibrillator in 1 patient (implanted for life-threatening ventricular tachyarrhythmias 29 years after septal myectomy). Syncope of unclear origin occurred in 8 patients, 3 of whom died. Indications for pacemaker implantation were early postoperative complete heart block (n = 5), late postoperative total atrioventricular block (n = 7), atrial fibrillation with bradycardia (n = 3), and sick sinus syndrome (n = 1). For pacemakers implanted late, the interval between operation and pacemaker implantation was 12 ± 8 years.

More than half of all patients (61/110, 56%) showed left bundle-branch block after septal myectomy.

Functional Classification and Recurrence of Symptoms

Most patients showed significant and long-term improvement in functional classification. One year after operation, 91% of the patients were in NYHA class I or II, and no patient was in class IV. Ten years later, 88% were still in NYHA class I or II and none in class IV (Fig 3).

Symptom-free survival at 1 year, 5 years, 10 years, 15

years, and 20 years was 93%, 77%, 50%, 33%, and 21%, respectively. Several preoperative characteristics were related to symptom-free survival. Of these, angina pectoris, dyspnea (NYHA class III or IV), congestive heart failure, and moderate to severe mitral regurgitation were independent predictors (Table 2). Patients with none of these predictors showed an excellent symptom-free survival (88% at 5 years and 77% at 10 years), whereas those with three or four predictors often showed an early recurrence of symptoms or death (symptom-free survival of 33% at 5 years and 0% at 10 years) (Fig 4).

Doppler Echocardiographic Results

Echocardiographic measurements were divided into preoperative, early postoperative (≤3 years after operation), and late postoperative (≥6 years postoperatively) (Table 3). Soon after operation, maximal instantaneous LVOT pressure gradient at rest decreased from a preoperative value of 81 ± 42 mm Hg to 13 ± 13 mm Hg (*p* < 0.001), maximal septal wall thickness in the LVOT from 21.0 ± 5.3 mm to 17.5 ± 5.7 mm (*p* < 0.001), and septal wall thickness, measured at the standard level, from 20.5 ± 5 mm to 19.1 ± 4 mm (*p* = not significant). Systolic anterior motion of the anterior mitral valve leaflet disappeared during the early postoperative period in 25 (40%) of 63 patients. There was also a significant reduction in

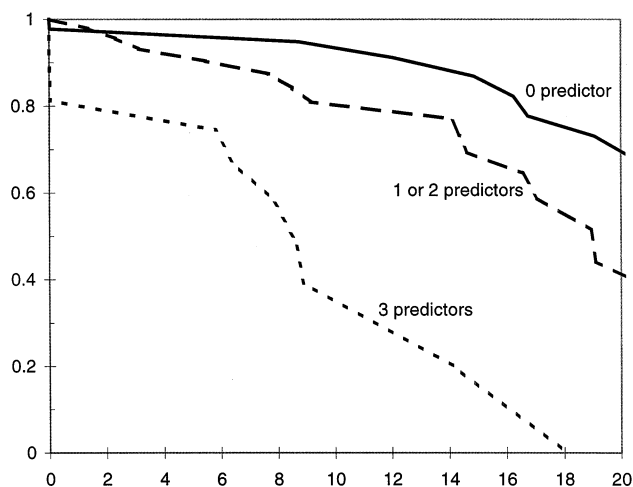


Fig 2. Actuarial survival of patients with no, one or two, and three preoperative risk factors (ie, New York Heart Association class III or IV, congestive heart failure, and additional surgical procedure).

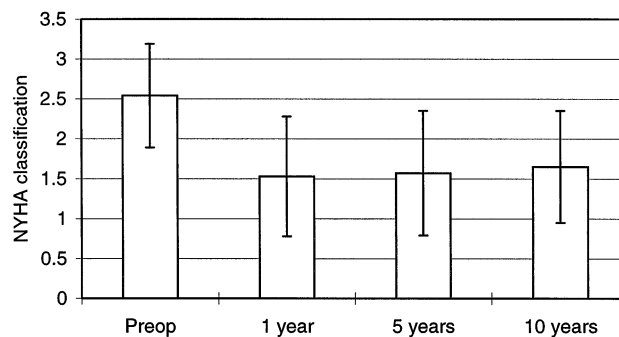


Fig 3. Mean New York Heart Association (NYHA) class before operation and 1 year, 5 years, and 10 years thereafter. Error bars denote standard deviation of the mean.

Table 2. Preoperative and Operative Predictors of Death or Recurrence of Symptoms After Septal Myectomy

Risk Factors for Death and Late Events	Univariate Cox Regression			Multivariate Cox Regression		
	Odds Ratio	<i>p</i> Value	95% Confidence Interval	Odds Ratio	<i>p</i> Value	95% Confidence Interval
NYHA class III or IV	3.13	0.001	1.81-5.41	2.26	0.001	1.22-4.18
Congestive heart failure	3.54	0.001	2.01-6.22	2.57	0.05	1.23-5.36
Pulmonary edema	3.37	0.001	1.71-6.63
Mitral valve regurgitation grade 3 or 4 (moderate to severe)	2.86	0.005	1.51-5.41	2.75	0.01	1.30-5.82
Angina pectoris	1.97	0.01	1.22-3.18	2.81	0.001	1.62-4.87
Atrial fibrillation	2.25	0.05	1.06-4.78

NYHA = New York Heart Association.

the end-systolic left atrial diameter from 45.0 ± 8.3 mm to 41.5 ± 7.3 mm ($p = 0.003$). In addition, a reduction in systolic fractional shortening from 0.45 ± 0.09 to 0.40 ± 0.08 ($p = 0.007$) and a decrease in the posterior wall thickness from 13.1 ± 2.5 mm to 12.1 ± 2.3 mm ($p = 0.008$) were found.

During long-term follow-up, the LVOT gradient was definitively eliminated. However, progressive enlargement of the left atrium and the left ventricle was observed. End-systolic left atrial diameter increased from 39.6 ± 8.0 mm to 48.2 ± 9.8 mm ($p < 0.001$) and end-diastolic ventricular diameter, from 46.5 ± 5.2 mm to 49.2 ± 7.4 mm ($p = 0.015$), probably as a result of diastolic dysfunction with increased filling pressures. Left ventricular chamber dilatation was detected by repeated echocardiographic measurements in 17 (30%) of 56 patients followed long term; 11 of these 17 patients died.

Aortic regurgitation was considered trivial in 6 patients and mild in 2 preoperatively. Postoperatively, 28 patients were seen with trivial, 4 with mild, and 1 with moderate aortic regurgitation. As already mentioned, 2 patients with severe regurgitation underwent reoperation, 1 in the first week after transaortic myectomy and the second 9 years after the first intervention.

Although systolic anterior motion of the mitral valve

disappeared completely during long-term follow-up, mitral regurgitation was present in 43 patients but was considered to be trivial or mild in all. As mentioned, 2 patients underwent late mitral valve reoperation because of persistent regurgitation.

Comment

Surgical treatment of HOCM has evolved in the last decade, and the Morrow septal myectomy with its modifications has become the preferred surgical technique. Better echocardiographic visualization with precise location of the LVOT obstruction before and during operation have helped the surgeon modify the extent of myectomy for various forms of left ventricular obstruction. Despite this progress, the operation remains a demanding procedure.

The present study is compared with others [7, 11-18] in Table 4. The results show that septal myectomy eliminates LVOT gradients and diminishes symptoms and mitral valve incompetence for a long time in most patients. In addition, early postoperative mortality is very low, and long-term follow-up is good. However, some patients may have left ventricular dilatation late after myectomy [9]. Hypertrophic obstructive cardiomyopathy is a rare disorder, and therefore most series report similar long observation periods. Patients are often severely limited (65% in NYHA class III or IV) and have a high LVOT gradient (average gradient, 70 mm Hg). An increasing number of additional cardiac procedures are necessary (28%), with bypass grafting representing half of them. The role of mitral valve operations varies across different institutions, but altogether, mitral replacement represents only 5% of the total of 1,178 operations.

Complications

Complications of the surgical procedure were experienced; the most common was reoperation in 8 patients (7.3%). This incidence is comparable to that of Heric and associates [12]. However, some of these reoperations would have been avoided today with the newer surgical techniques and improvements in preoperative assessment. Complete heart block resulting in pacemaker insertion was observed in 5 patients. The incidence of

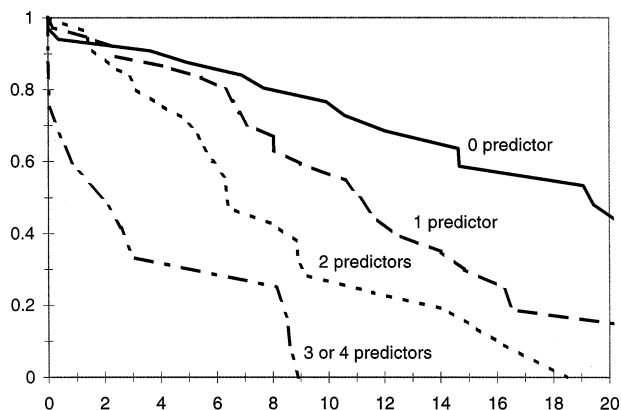


Fig 4. Symptom-free survival with no, one, two, and three or four preoperative risk factors (ie, New York Heart Association class III or IV, congestive heart failure, angina pectoris, and mitral regurgitation grade 3 or 4).

Table 3. Echocardiographic Results After Septal Myectomy^a

Variable at Transthoracic Doppler Echocardiography	Preop (n = 51)	≤3 Years Postop (n = 51)	p Value ^b	≤3 Years Postop (n = 31)	≥6 Years Postop (n = 31)	p Value ^b
Maximal instantaneous LVOT pressure gradient at rest (mm Hg)	81 ± 42	13 ± 14	<0.001	5 ± 12	1 ± 4	NS
Septal wall thickness (mm)	21.0 ^c ± 5.3	17.5 ^c ± 5.7	<0.001	16.7 ^c ± 4.5	17.0 ^c ± 4.6	NS
Posterior wall thickness (mm)	13.1 ^c ± 2.5	12.1 ^c ± 2.3	<0.008	11.4 ± 1.5	11.3 ± 1.7	NS
End-diastolic ventricular diameter (mm)	43.7 ^c ± 7.2	45.0 ± 5.9	NS	46.5 ± 5.2	49.2 ± 7.4	<0.015
LV fractional shortening	0.45 ± 0.09	0.40 ± 0.08	<0.007	0.37 ± 0.08	0.39 ± 0.08	NS
End-systolic left atrial diameter (mm)	45.0 ^c ± 8.3	41.5 ^c ± 7.3	<0.003	39.6 ± 8.0	48.2 ^c ± 9.8	<0.001

^a Data are shown as the mean ± the standard deviation. ^b Significance was determined with the paired Student's *t* test. ^c This is a pathologic value.

LV = left ventricular; LVOT = left ventricular outflow tract; NS = not significant; septal wall thickness = maximal end-diastolic thickness in LVOT.

complete heart block is comparable to the results reported by others [7, 11, 14]. In contrast to the series of Heric and coworkers [12], our patients with complete heart block did not have a worse outcome.

Despite postoperative aortic regurgitation, which is probably due to intraoperative leaflet trauma or resection of ventricular muscle too close to the aortic annulus, the transaortic approach remains our preferred technique, as aortic regurgitation is mostly only trivial or mild [19]. Only 2 patients required reoperation because of major aortic regurgitation.

Progressive dilatation of the left ventricle has been observed repeatedly in medically as well as surgically treated patients with HOCM [9]. It was present in 17 of our patients. It can be argued that septal myectomy provokes dilatation of the left ventricle in long-term follow-up. However, the incidence in moderately to severely symptomatic patients with medical treatment of HOCM is yet unknown. Furthermore, only 18 of the 106 patients surviving the perioperative period had development of clinically overt congestive heart failure during long-term follow-up. Most of these patients were severely symptomatic before operation. Thus, dilatation of the left ventricle can possibly be prevented if patients undergo operation at a relatively early stage of disease.

The expanded cross-sectional area of the LVOT diminishes or eliminates systolic anterior motion of the anterior mitral valve leaflet and may simultaneously alleviate mitral regurgitation. In our patients, mitral regurgitation did not worsen during long-term follow-up, and only patients with moderate to severe mitral regurgitation at baseline needed additional mitral valve procedures, although a diversity of structural mitral valve alterations exists in HOCM [20]. Septal myectomy can be combined with mitral valve repair. However, the role of the latter in these patients must still be determined. It is not possible to evaluate the role of the different repair techniques used depending on the underlying lesion in our small subgroup of 10 patients. Replacement may remain a possibility for select patients with severe intrinsic disease of the mitral valve, but it appears to worsen the prognosis [12].

Long-Term Prognosis and Predictors of Survival

Because of the relative rarity of HOCM, there are currently no long-term results comparing medically and surgically treated patients in a randomized, prospective trial. Therefore, the optimal treatment of these patients is still controversial [2-5, 15, 17, 21]. The survival of our patients compares favorably with natural history studies [22]. In addition, multivariate analysis has allowed us to identify subgroups of patients with relatively low risk during long-term follow-up. On the other hand, patients with advanced disease seen with severe symptoms related to diastolic dysfunction had a worse outcome. In combination with additional surgical procedures, the risk of death was even greater. Thus, it seems advisable to operate early on patients with related disorders that require an additional surgical procedure before they become severely symptomatic. Furthermore, patients with some but not all possible symptoms of HOCM had a much better prognosis after operation. Although our data do not allow a direct comparison with the natural history, patients with progressively symptomatic HOCM should not have operation too late, as survival as well as symptom-free survival is many times higher in mildly to moderately symptomatic patients than severely symptomatic patients.

Age has been repeatedly described as an independent predictor of death in surgically [12, 14, 15] and medically [22] treated patients. However, our data did not show a relation between age and risk of a bad outcome. Although there is no easy explanation for this finding, the relatively small subgroups of patients who underwent surgical therapy either when they were very young or old make statistical comparison difficult. Therefore, differences may be observed by chance and not as a significant finding. In addition, an even slightly higher risk in some age groups does not mean that these patients do not benefit from septal myectomy. In our series, 12 patients who were seen with symptomatic HOCM in childhood had operation and a prognosis as good as that of the other patients.

Table 4. Summary of Literature Comparison

Reference	No. of Patients	Period of Operation	NYHA Classes III and IV (%)	LVOT Pressure Gradient (mm Hg)	SM + Other Procedures (%)	Mitral Valve Replacement	SM + CABG (%)	Early Death (%)	Follow-up (y)	Survival (%)			HOCM Yearly Mortality (%)
										5 Years	10 Years	15 Years	
Cohn et al, 1992 [11]	31	1972-1991	94	96	26	0	19	0.0	6.5	100	86	NA	1.0
Heric et al, 1995 [12]	178	1975-1993	65	93	47	12	23	6.0	3.7	86	70	NA	0.6
McCully et al, 1996 [13]	65	1986-1992	95	66	31	3	7	4.6	2.3	92	NA	NA	NA
Mohr et al, 1989 [14]	115	1972-1987	34	70	30	6	17	5.2	5.1	84	73	NA	1.0
Robbins and Stinson, 1996 [15]	158	1972-1994	72	64	16	3	14	3.2	6.1	85	72	46	1.7
Schoendube et al, 1995 [7]	58	1979-1992	91	79	30	0	17	1.7	7.0	93	86	70	1.4
Schulte et al, 1993 [16]	364	1963-1991	60	54	25	6	6	4.9	8.2	92	88	84	0.6
Ten Berg et al, 1994 [17]	38	1977-1992	84	72	32	11	11	0.0	6.8	100	NA	NA	0.0
Williams et al, 1987 [18]	61	1971-1986	66	79	28	0	8	1.6	3.0	93	93	NA	1.1
Schönbeck et al, 1998 [this study]	110	1965-1995	57	86	21	2	3	3.6	11.7	93	80	72	1.5
Total or average	1,178	1963-1995	65	70	28	5	15	4.2	6.5	n.a.	n.a.	n.a.	1.3

CABG = coronary artery bypass grafting; HOCM = hypertrophic obstructive cardiomyopathy; LVOT = left ventricular outflow tract; NA = not available; NYHA = New York Heart Association; SM = septal myectomy.

Echocardiographic Findings

Our echocardiographic data expand those in previous reports [7, 9, 12, 15]. This study confirms the finding that septal myectomy may lead to a regression in not only the surgically treated septum but also the posterior wall. Further, the left atrial dimension decreased in short-term and medium-term follow-up, thus indicating improvement in diastolic dysfunction after septal myectomy, although it progressed in long-term follow-up. Thus, a high pressure gradient in the LVOT seems to further impair left ventricular myocardium, even in parts without excessive hypertrophy. It is well known that patients with symptoms of aortic stenosis have a bad prognosis and that relief of the pressure gradient over the aortic valve is necessary to prolong survival. Thus, it can be hypothesized that a high pressure gradient in the LVOT is an independent risk factor for a bad outcome and that relief of the gradient improves survival. This may explain a possible benefit of septal myectomy regarding survival in patients with HOCM.

Conclusions

This relatively large series with a mean follow-up exceeding 10 years demonstrates a good outcome for patients with advanced HOCM after surgical septal myectomy. It further indicates that patients should have operation before they become severely symptomatic (eg, NYHA III or IV and overt congestive heart failure), particularly if they need additional surgical procedures. This may be of more importance given the finding that relief of the LVOT pressure gradient favorably influences remodeling of the left ventricle during midterm follow-up.

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DISCUSSION

DR CARY W. AKINS (Boston, MA): I congratulate your group on an excellent presentation on a very difficult group of patients. We see two different kinds of patients with septal thickening: those who have true idiopathic hypertrophic subaortic stenosis and who, when you look at them pathologically, have a disordered arrangement of their muscular fibers, and those who have development of septal hypertrophy that is secondary to aortic valvular stenosis. Have your results been different in these two groups, or did you not include patients with septal thickening secondary to aortic stenosis in this study?

DR TURINA: In this particular group, only those with true idiopathic hypertrophic subaortic stenosis were included. The reactive septal hypertrophy, or asymmetrical septal hypertrophy as it is called by the echocardiographers, is a separate disease. It is secondary to the long-standing pressure gradient and sometimes even occurs without aortic valve disease in patients with hypertension. These patients were not included.

Careful analysis of the resected specimen by our pathologist showed that only in a small proportion of our patients with idiopathic hypertrophic subaortic stenosis can true signs of a cardiomyopathy be found. However, it is not a specific disease. In some patients, it is very difficult to make the distinction between the reactive hypertrophy and the true disease.

DR D. CRAIG MILLER (Stanford, CA): Do I understand you to mean that patients with "old person's septal shoulder," as the echocardiographers call it, seen in the elderly with hypertension very close to the aortic valve, were excluded from this analysis?

DR TURINA: Yes, they were excluded. Today these patients are treated with a calcium antagonist, which causes a reduction in the gradient.

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