Combined anterior mitral valve leaflet retention plasty and septal myectomy in patients with hypertrophic obstructive cardiomyopathy

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Abstract

Objectives: Septal myectomy is the treatment of choice for patients with hypertrophic obstructive cardiomyopathy (HOCM) with significant left-ventricular outflow tract (LVOT) obstruction. In some HOCM patients, however, systolic anterior motion (SAM) of the anterior mitral leaflet significantly contributes to LVOT obstruction, resulting in mitral regurgitation and insufficient release of the obstruction after myectomy. We, therefore, developed a strategy of combined myectomy and anterior leaflet retention plasty (ALRP), and investigated its results in adult HOCM patients with manifest SAM. Methods: Subaortic septal myectomy and ALRP were performed in 25 adult HOCM patients with significant SAM, as assessed by echocardiography (mean age = 48.5 ± 15 years). All patients received cardiac catheterization and echocardiography evaluation prior to the operation, before discharge, and at follow-up. Follow-up ranged between 0.8 and 14 years (median = 2.5 years). Results: All patients survived the operation, and the Kaplan–Meier estimated survival was 100% at 1 year and 82 ± 6% at 5 years. Freedom from re-operation at 5 years was 83 ± 8%. The mean LVOT pressure gradient decreased from 84 ± 32 to 19 ± 11 mmHg postoperatively (p < 0.001), and only two patients had mild residual or recurrent SAM at follow-up. Mitral regurgitation and New York Heart Association classification were also markedly improved at follow-up. Conclusions: Combined subaortic septal myectomy and ALRP is a safe and effective therapy in HOCM patients with significant SAM. ALRP can help prevent residual or recurrent LVOT obstruction and improves mitral regurgitation.

Keywords: Mitral valve reconstruction; HOCM; SAM; Mitral regurgitation

1. Introduction

1.1. Background

Hypertrophic obstructive cardiomyopathy (HOCM) is usually treated with myectomy if relevant left-ventricular outflow tract (LVOT) obstruction is present [1]. In some patients, however, systolic anterior motion (SAM) of the anterior leaflet of the mitral valve significantly adds to the LVOT obstruction [2], and may also cause mitral regurgitation (MR) [3]. Several hypotheses have been suggested to help explain the development of SAM in HOCM patients. Among those are the Venturi effect due to high-velocity blood flow in the narrowed LVOT, and elongated chordae in the central part of the anterior mitral valve leaflet (A2). In systole, an anterior mitral leaflet (AML) that is elongated in relation to the depth of the posterior leaflet may be pushed toward the LVOT, causing obstruction [4], and, if central co-apotation is hindered, MR. In HOCM, the LV cavity is often extremely small, inflow and outflow tract overlap, and a disarray of muscle architecture may lead to severe hypokinesia of the septum and hyperkinesia of posterior wall. These factors, together with anterior displacement of the papillary muscles, complete the typical picture of the disease [5].

Subaortic septal myectomy widens the LVOT and thus reduces blood-flow velocity, pressure gradient, and — to some extent — also MR [3]. However, mitral valve abnormalities, when they are not addressed, can lead to suboptimal surgical results, especially in those patients with distinct preoperative SAM. Mitral valve replacement can solve these problems [6], but is associated with prosthesis-related problems, such as thrombo-embolism, bleeding, and endocarditis, and is therefore usually reserved for patients in whom mitral valve repair fails. Different approaches to repair the mitral valve in HOCM patients have been introduced, such as AML plication [7], reconstruction of the subannular mitral apparatus [8], and AML extension [9]. Recently, Delmo Walter and colleagues introduced our approach of combined

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myectomy and anterior leaflet retention plasty (ALRP) in children with severe HOCM [10]. Here, we report on the results of this combined surgical approach in adult patients.

2. Patients and methods

2.1. Demographics and clinical presentation

The subjects of this review are the 25 adult patients who presented with HOCM, significant SAM, and at least grade I MR between 1994 and 2008. Eleven patients were female and 14 male, with a mean age of 48.5 ± 15 years. Electrocardiography showed sinus rhythm in 22 patients and atrial fibrillation in three. Despite optimal oral medication with β-blockers, calcium-channel blockers, or both, three patients were in New York Heart Association (NYHA) class 2, 19 in class 3, and three in class 4. LVOT pressure gradient, LV function, MR, and interventricular septal thickness in diastole and systole were evaluated by preoperative transthoracic echocardiography and by transesophageal echocardiography in the operating room. When SAM was found to significantly add to the LVOT pressure gradient and/or produce relevant MR (SAM grade 2 or 3, see later), the decision was made to perform combined myectomy and ALRP. The severity of SAM was graded based on the duration of the systolic AML shift, as assessed by two-dimensional (2D) echocardiographic imaging. Grade 0 indicates no SAM, grade 1 a brief (one-third of systole or less) shift of the AML and its subvalvular apparatus toward septum, grade 2 an AML shift during two-thirds of systole, and grade 3 a pansystolic AML shift. Peak and mean LVOT gradient (P) was estimated from velocity (V) assessment by Doppler echocardiography by the modified Bernoulli equation 

\[ P = 4V^2 \]

All patients had typical marked SAM of the anterior mitral valve leaflet and subvalvular apparatus (mean grade 2.7 ± 0.6). The mean peak LVOT pressure gradient was 84 ± 32 mmHg and reached 150 mmHg in several patients (Fig. 1). The average thickness of the interventricular septum measured 23 ± 7 mm in diastole and 26 ± 7 mm in systole. Two patients had MR grade IV, eight patients had grade III, 11 patients had grade II, and four patients had grade I, determined using transesophageal quantitative Doppler echocardiography with quantification of the regurgitation jet (vena contracta), calculation of regurgitant volume and fraction, and assessment of systolic pulmonary vein flow pattern [11]. Mean LV ejection fraction was 67 ± 11%.

2.2. Surgical technique

Surgery was performed via median sternotomy, aortic and bivacal cannulation, and cardiopulmonary bypass. The LV was vented via the right upper pulmonary vein. After the aorta was clamped, an oblique aortotomy was carried out into the non-coronary sinus and cold crystalloid cardioplegic solution (between 1997 and 2004) or warm-blood cardioplegic solution (since 2004) was directly injected into the right and left coronary ostia.

The technique of subaortic septal myectomy has not changed since its description by Morrow [12]. Next, the mitral valve is inspected via a left atriotomy and the medial and lateral AML segments closest to the trigones are sutured to the corresponding posterior annulus, with polypropylene mattress sutures pledged with autologous pericardium (Fig. 2). In addition, a maze procedure for atrial fibrillation was performed in three patients, coronary artery bypass grafting for coronary artery disease in two, aortic valve replacement for aortic stenosis in two, and atrial septal defect closure in one. Finally, the aortotomy and atriotomy are closed, the heart is de-aired, and the aortic clamp is released. Cardiopulmonary bypass is discontinued and transesophageal echocardiography is performed. We always attach temporary pacing wires to the LV apex and the right atrium, and perform dual-chamber pacing with a short atrioventricular (AV) delay (60–100 ms). If there is a further reduction in the residual gradient across the LVOT by echocardiography and/or a decrease in left atrial pressure (LAP), we implant permanent epicardial DDD pacemaker wires, irrespective of the presence of AV block.

2.3. Data collection

Clinical features, operative procedure, perioperative course, and LV function at the time of discharge were reviewed from clinical records. Between January 2006 and March 2009, 17 of the 25 patients who had undergone ALRP and subaortic septal myectomy received transthoracic echocardiography follow-up studies in our outpatient department. Informed consent was obtained from each patient. Follow-up ranged between 0.84 and 14.4 years (median 2.5 years), representing a total of 96.7 patient-years. Recent information regarding survival and reoperations was available for all patients (100%).
2.4. Statistics

Statistical Package for Social Sciences (SPSS) for Windows version 12.01 was used for statistical analysis. Qualitative data are expressed as numbers (n) and percentages and quantitative data as medians and ranges or as mean and standard deviation, as appropriate. Pre- and postoperative echocardiographic parameters (LVOT gradient and wall thickness) were compared using the Wilcoxon signed rank test. Freedom from re-operation and survival were calculated according to the Kaplan–Meier analysis with 95% confidence intervals (CIs).

3. Results

3.1. Mortality

All patients survived the operation and were discharged from the hospital. The Kaplan–Meier analysis showed 30 day, 1-year, and 5-year survival of 100%, 100%, and 82.2 ± 9.4%, respectively (Fig. 3). Three patients died late after the operation. One patient (aged 53 years) died from multi-organ failure 2.1 years after the initial operation after receiving heart transplantation for restrictive cardiomyopathy. Two other patients died late after operation. Another patient suffered sudden cardiac death 3.1 years after the operation. Preoperatively, her LVOT pressure gradient was 50 mmHg, associated with SAM grade 3 and MI I. She had $\Delta P_{\text{max}}$ of 19 mmHg and no MR at discharge, but declined to undergo follow-up diagnostics at our hospital. Her cardiologist was contacted and reported that she had no severe MR several weeks before she died. The third patient had sudden cardiac death 4.6 years after the operation. With a preoperative $\Delta P_{\text{max}}$ of 70 mmHg, SAM grade 3, and MR grade III, he had $\Delta P_{\text{max}}$ of 12 mmHg and MR grade I at discharge and at 1-year follow-up. His cardiologist reported MR grade I 9 weeks before he suddenly died. Autopsies were not performed.

3.2. Freedom from re-operation

A total of six patients required re-operation. One day after septal myectomy and ALRP, two patients showed hemodynamic instability, and recurrent mitral insufficiency grade III was diagnosed by transesophageal echocardiography. Both patients immediately underwent mitral valve replacement and had an uneventful further postoperative course. Intraoperatively, the ALRP mattress sutures were found to be ruptured. During follow-up, two further patients required mitral valve replacement for recurrent MR grade III (0.9 and 5.2 years after myectomy and ALRP, respectively), despite low LVOT gradients and no evidence of SAM. The ALRP mattress sutures were found to be intact, but there was significant annulus dilation. Mitral valve replacement was again performed and was tolerated well by both patients. Finally, two other patients underwent heart transplantation 2.1 and 11.3 years after the operation for end-stage restrictive heart failure, despite adequate relief of the LVOT obstruction and appropriate mitral valve function. One of these patients died postoperatively from septic multi-organ failure. The other is alive and well 2 years after transplanta-
tion. Five of the six patients receiving re-operation are alive and doing well. Taken together, the Kaplan–Meier-estimated 30-day, 1-year, and 5-year freedom from re-operation was 92 ± 5.4%, 87.8 ± 6.6%, and 83.2 ± 7.7%, respectively (Fig. 4).

3.3. Hemodynamics and mitral valve function

The mean preoperative LVOT pressure gradient dropped significantly from 84 ± 32 to 19 ± 11 mmHg after subaortic septal myectomy and ALRP (p < 0.001; Fig. 1). On echocardiographic follow-up evaluation after a median of 2.5 years (range 0.8–14.4 years), no significant change in LVOT pressure gradient from that at the time of discharge was observed. Mean systolic and diastolic interventricular thickness at follow-up had decreased significantly from 26 ± 6.7 and 23 ± 6.6 mm to 17 ± 4.9 (p = 0.001) and 14 ± 4.3 mm (p = 0.001), respectively.

At the time of discharge, MR had improved in 23 patients (92%) to grade I or 0 (Fig. 5). Two patients developed recurrent mitral valve insufficiency (grade III) 24 h after myectomy and ALRP (see later). Follow-up evaluation of 17 patients, who were alive and had no re-operation, showed a further decrease of MR in three patients (18%), no further change in 10 (59%), and a moderate increase in four (23%). However, no patient had MR > grade II at follow-up. Two patients developed MR grade III after 0.9 and 5.2 years, postoperatively.

Significant SAM was present prior to the operation in all patients, and was shown to have improved postoperatively by intra-operative transeosophageal echocardiography in all. At the time of discharge, transthoracic echocardiography identified four patients (17%) with mild residual SAM and LVOT obstruction with a gradient of <35 mmHg and mild or no MR. At follow-up, only two out of 17 evaluated patients showed SAM: one patient with a maximal LVOT pressure gradient of 50 mmHg and the other with 14 mmHg. However, both patients were in NYHA class 1.

No patient developed postoperative AV block. However, intra-operative analysis of hemodynamics showed 17 patients to have a further improvement of the LVOT gradient and reduction of LA pressure during dual-chamber pacing via the temporary epicardial pacing wires with a short AV delay. Therefore, these patients also received a permanent epicardial DDD pacemaker.

3.4. NYHA classification

Preoperatively, three patients were in NYHA class 2, 19 in NYHA class 3, and three in NYHA class 4. At follow-up, the majority of patients had improved in NYHA class: 12 of 17 patients, who did not require re-operations, were in NYHA class 1 and five patients were in NYHA class 2 (Fig. 6).

4. Discussion

Once an LVOT pressure gradient of ≥50 mmHg has developed despite medical therapy with β-blockers and verapamil, surgery for HOCM is indicated [13], and subaortic septal myectomy [1] has become the gold standard to relieve the LVOT pressure gradient and reduce associated SAM and
MR [14–17]. Although myectomy is a safe and effective procedure, MR and LVOT obstruction symptoms often recur, especially in those patients with severe preoperative SAM. Mitral valve replacement has been proposed by Cooley and colleagues [6,18,19] to solve this problem, especially in the adult patient; nowadays, however, most surgeons try to avoid primary mitral valve replacement for HOCM.

We advocate a form of mitral valve reconstruction that specifically addresses the excessive mobility of the anterior leaflet, which contributes to the SAM phenomenon. This ALRP differs from the anterior leaflet reconstructions described by other groups [10]. After traditional mitral valve reconstruction by shortening the circumference of the posterior annulus, SAM is caused or exacerbated by ‘over-reconstruction’ when the tight posterior annuloplasty shifts the mural part of the mitral annulus toward the LV outflow tract. By contrast, the technique we advocate prevents the shift of the valvular and subvalvular apparatus toward the septum by stretching the central portion of the AML (A2) and limiting AML motion, relieving SAM and MR [10]. It is important to distinguish ALRP from the modified Kay-Whooler annuloplasty, where bilateral mattress sutures are placed longitudinally through the annulus of the posterior mitral valve leaflet, posterior to the right and left fibrous trigones, leaving the zone of apposition between AML and PML open and not interfering with AML mobility. In ALRP, however, the mattress sutures are placed immediately below the fibrous trigones, attaching about one-half to two-thirds of the lateral AML edge to the corresponding portion of the PML annulus. Thereby, AML mobility is reduced while the geometry and length of the posterior annulus remain unchanged. Of major importance to prevent mitral valve stenosis and to achieve good ALRP results is a mitral valve orifice area of at least 3.5 cm² in adults, and a mean age-dependent orifice area of at least –10% of the normal value in children. Again, ALRP facilitates this because the circumference of the posterior annulus is not markedly shortened.

Other groups have reported good results of surgery for HOCM with SAM obtained by combining subaortic septal myectomy and mitral valve repair by more traditional techniques [7,8,20,21]. Schoendube and colleagues combined subaortic septal myectomy with reconstruction of the subannular mitral apparatus [8], mobilizing and separating the papillary muscles. In their series, relief of LVOT obstruction was seen in all patients. At follow-up, mild SAM was present in 10% and mild MR in 37% of patients studied. However, this procedure assumes that papillary muscle displacement is present, which we did not see in our patients. Kofflard and colleagues, performing combined AML extension plasty and subaortic septal myectomy, presented similar good results [9]. In this technique, a pericardial patch is sutured to the ventricular surface of the AML to increase its width. The patch serves to stiffen the leaflet, making it less lax and less likely to buckle into the LVOT during systole in the presence of Venturi forces. McIntosh combined subaortic septal myectomy with AML plication and achieved similar good results with reduction of LVOT gradient to ≤35 mmHg in 90% of patients and absent or mild MR in 80% [7]. This mitral valve procedure resembles the AML extension technique and also restricts anterior leaflet movement.

In addition, we observed a further improvement of LV function in several patients when dual-chamber pacing with an individually adapted shortened AV delay (60–100 ms) was performed intra-operatively. Therefore, we decided to implant a permanent epicardial DDD pacemaker system, although none of our patients had AV block after septal myectomy. We believe that the combination of septal myectomy, ALRP, and, if indicated, optimization of AV and intraventricular impulse propagation helps achieve a satisfactory long-term outcome for patients with HOCM and associated mitral valve dysfunction. However, as evidenced by our two patients who still required heart transplantation, all conventional surgical approaches may fail, when generalized hypertrophy progresses to restrictive cardiomyopathy.

We conclude that combined subaortic septal myectomy and ALRP is a safe and effective therapy in adult HOCM patients with distinct preoperative SAM. It prevents LVOT obstruction by SAM after myectomy and improves MR and heart-failure symptoms in the majority of patients. As it is a relatively simple procedure, it may be considered whenever SAM and MR are present in HOCM patients.

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References


