

Extended Myectomy for Hypertrophic Obstructive Cardiomyopathy

Changqing Gao, Chonglei Ren, Cangsong Xiao, Yang Wu, Gang Wang,
Guopeng Liu, Yao Wang

Department of Cardiovascular Surgery, General Hospital of PLA, Institute of Cardiac Surgery of PLA,
Beijing, China



Dr. Gao

ABSTRACT

Background: The purpose of this study was to summarize our experience of extended ventricular septal myectomy in patients with hypertrophic obstructive cardiomyopathy (HOCM).

Methods: Thirty-eight patients (26 men, 12 women) with HOCM underwent extended ventricular septal myectomy. The mean age was 36.3 years (range, 18-64 years). Diagnosis was made by echocardiography. The mean (mean \pm SE) systolic gradient between the left ventricle (LV) and the aorta was 89.3 ± 31.1 mm Hg (range, 50-184 mm Hg) according to echocardiographic assessments before the operations. Moderate or severe systolic anterior motion (SAM) of the anterior leaflet of the mitral valve was found in 38 cases, and mitral regurgitation was present in 29 cases. Extended ventricular septal myectomy was performed in all 38 cases. The results of the surgical procedures were evaluated intraoperatively with transesophageal echocardiography (TEE) and with transthoracic echocardiography (TTE) at 1 to 2 weeks after the operation. All patients were followed up with TTE after their operation.

Results: All patients were discharged without complications. The TEE evaluations showed that the mean systolic gradient between the LV and the aorta decreased from 94.8 ± 35.6 mm Hg preoperatively to 13.6 ± 10.8 mm Hg postoperatively ($P = .0000$) and that the mean thickness of the ventricular septum decreased from 28.3 ± 7.9 mm to 11.8 ± 3.2 mm ($P = .0000$). Mitral regurgitation and SAM were significantly reduced or eliminated. During the follow-up, all patients promptly became completely asymptomatic or complained of mild effort dyspnea only, and syncope was abolished. TTE examinations showed that the postoperative pressure gradient either remained the same or diminished.

Conclusions: Extended ventricular septal myectomy is mostly an effective method for patients with HOCM, and good surgical exposure and thorough excision of the hypertrophic septum are of paramount importance for a successful surgery.

Received December 3, 2011; accepted July 14, 2012.

Correspondence: Changqing Gao, MD, Department of Cardiovascular Surgery, PLA General Hospital, 28 Fuxing Rd, Beijing 100853, China; 86-10-88626988; fax: 86-10-88626988 (e-mail: gaochq301@yahoo.com).

INTRODUCTION

Surgical treatment of hypertrophic obstructive cardiomyopathy (HOCM) was initially introduced in the late 1950s. Regarded as revolutionary, it has developed in accordance with the changing pathophysiological concepts of the disease. In 1968, Morrow et al described the "subaortic myectomy" and reported good clinical and hemodynamic results [Morrow 1968]. The classic myectomy operation (also known as the Morrow procedure) has become established as a proven approach for amelioration of outflow obstruction with the standard surgical option. It is the gold standard for patients with HOCM and severe drug-refractory symptoms [Maron 2004, 2007; Brown 2008]. Surgical myectomy is a technically challenging procedure for surgeons and has a steep learning curve. The operative mortality rate and the incidence of complications were relatively high in early surgical series [Smedira 2008]. Although the operative mortality rate has been reduced to 2% in patients undergoing isolated septal myectomy, results for the myectomy operation remain unsatisfactory [Maron 2003a; Ommen 2005; Maron 2006]. The purpose of this study was to summarize the surgical experience of extended septal myectomy by a single surgeon in patients with HOCM refractory to medical therapy.

PATIENTS AND METHODS

Patients

Between June 2003 and March 2011, 38 patients with HOCM refractory to medical therapy underwent surgical treatment. The 26 male patients and 12 female patients ranged in age from 18 to 64 years (mean \pm SE of the mean, 36.3 ± 15.6) and had a mean body weight of 64.5 ± 17.7 kg. Of the patients, 35 (92%) had dyspnea and angina, and 20 patients (53%) had a history of syncope and near syncope. Six of the patients had undergone previous septal alcohol ablation but did not experience symptom improvement. Nine patients (24%) had a family history of hypertrophic cardiomyopathy, 9 patients (24%) had atrial fibrillation, and 6 patients (16%) had left bundle branch block. Sixteen patients were in New York Heart Association (NYHA) class II, 18 patients were in class III, and 2 were in class IV. Chest radiographic examinations showed a mean cardiothoracic ratio of 0.57 ± 0.05 .

(range, 0.52-0.64). The diagnosis of HOCM was established by echocardiography and spiral computed tomography with the characteristic of marked left ventricular (LV) asymmetry hypertrophy. Echocardiographic assessments revealed the mean preoperative peak gradient between the LV and the aorta to be 89.3 ± 31.1 mm Hg (range, 50-184 mm Hg). All patients had moderate or severe systolic anterior motion (SAM) of the anterior leaflet of the mitral valve. Mild or moderate mitral regurgitation was found in 27 cases, and severe mitral regurgitation occurred in 2 cases. One patient had a ventricular septal defect, and 1 patient had infective endocarditis with mitral valve vegetation. Coronary angiography examinations confirmed that 3 patients had a moderate or severe coronary myocardial bridge and that 2 patients had multivessel coronary artery disease.



Figure 1. Excised septum.

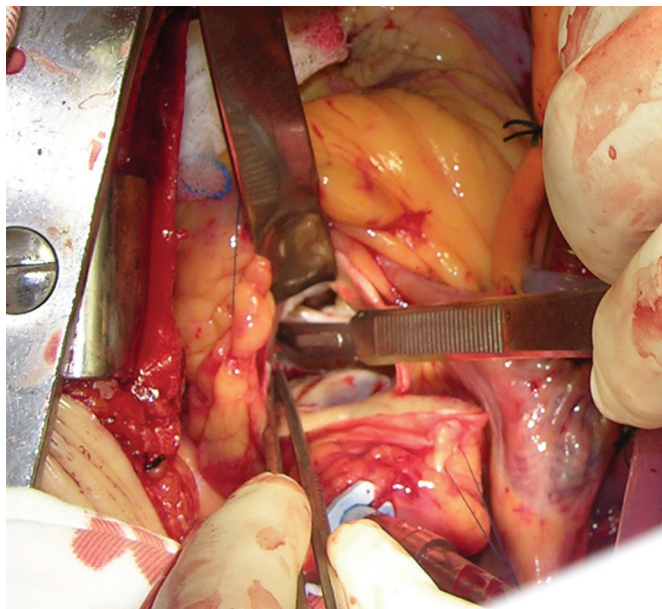


Figure 2. Division of aorta.

Extended Septal Myectomy

Septal myectomy was performed with the patient under general anesthesia; standard cardiopulmonary bypass and mild to moderate systemic hypothermia were used. Myocardial protection was achieved with cold blood cardioplegia through topical cooling after clamping of the aorta. The aorta was opened via a low transverse aortotomy. The right aortic cusp was retracted and depressed anteriorly against the sinus wall by a narrow ribbon retractor placed into the LV outflow tract (LVOT), and the hypertrophied septum could be seen bulging down into the anterior aspect of the LVOT. A no. 15 scalpel blade was used to make an incision deep into the septum, exactly beneath the nadir of the right coronary cusp and parallel to the long axis of the LVOT. A second parallel incision was made in the septum as far leftward to the mitral valve as possible. Pressing a sponge stick against the right ventricular free wall to depress the ventricular septum brought it into better view through the aortotomy. Both incisions were then deepened and carried toward the LV apex as far as possible. The 2 vertical incisions were then connected by a transverse incision, beginning several millimeters below the right coronary cusp. With continuing pressure on the sponge stick, this transverse incision was extended downward into the ventricle until a thick rectangular piece of septum was excised (Figure 1). Septal tissue could be excised thoroughly, if necessary, to further increase the cross-sectional area of the LVOT. The depth of the muscle resection was dependent on the preoperative septal thickness. In 9 cases, the aorta was cut off completely to optimize visualization of the hypertrophic septum and obstructed LVOT and to explore the pathologic anatomy, such as the site and extent of the hypertrophic septum and the mitral valve malformation (Figure 2). All 38 patients with HOCM underwent extended septal myectomy (known as the modified Morrow procedure). After the extended myectomy, a wider groove appeared in the LVOT (Figure 3B). Concomitant procedures included mitral valve replacement (MVR) in 2 patients, coronary artery bypass grafting in 2 cases, ventricular septal defect repair in 1 patient, lyses of the myocardial bridge in 2 patients, and mitral valve repair in 1 patient. A low dosage of β -blockers was continued postoperatively.

Evaluation and Follow-up

Intraoperative transesophageal echocardiography (TEE) was carried out to assess the site and the extent of the proposed myectomy, the structural features of the mitral valve before resection, and the surgical results of the septal myectomy. At 1 to 2 weeks after the operation, transthoracic echocardiography (TTE) was performed to evaluate the effect of the operation. All patients were followed up with TTE after their operation.

Statistical Analysis

Statistical analysis was carried out by the first author with SPSS software (version 13.0; SPSS/IBM, Chicago, IL, USA). All continuous variables are expressed as the mean \pm SE of the mean. Pre- and postmyectomy quantitative variables were assessed with the Student t test or with the t test for paired data. Numeration data were analyzed with the rank sum test. Statistical significance was assumed at a probability level of $<.05$.

Table 1. Comparison of Pre- and Postoperative Echocardiographic Data for 38 Patients Undergoing the Modified Morrow Procedure*

Echocardiography Data	Preoperative	Postoperative	t	P
LA diameter, mm	43.4 ± 8.1	35.4 ± 6.6	4.578	.0000
LVEDD, mm	37.9 ± 5.6	36.1 ± 4.4	1.498	.1387
IVS thickness, mm	28.3 ± 7.9	11.8 ± 3.2	11.656	.0000
PWLV thickness, mm	15.2 ± 4.5	14.7 ± 3.8	0.5130	.6096
LVOTG, mm Hg	89.3 ± 31.1	17.9 ± 12.9	12.729	.0000
LVEF, %	77.3 ± 8.5	67.6 ± 7.4	5.168	.0000

*Data are presented as the mean SE of the mean. LA indicates left atrial; LVEDD, left ventricular end-diastolic diameter; IVS, interventricular septal; PWLV, posterior wall of left ventricle; LVOTG, left ventricular outflow tract gradient; LVEF, left ventricular ejection fraction.

Table 2. Echocardiographic Data Showing Changes in Mitral Regurgitation and Systolic Anterior Motion from Preoperatively to Postoperatively for 38 Patients Undergoing the Modified Morrow Procedure

Grade*	Preoperative	Postoperative	U [†]	P
Mitral regurgitation				
–	9	31		
+	19	7		
++	8	0		
+++	2	0		
Total	38	38	5.213	<.01
Systolic anterior motion				
–	0	32		
+	2	6		
++	20	0		
+++	18	0		
Total	38	38	7.930	<.01

*Grade severity is indicated as follows: negative (–), mild (+), moderate (++), severe (+++).

[†]Mann-Whitney U statistic.

Table 3. Comparison of Postoperative and Follow-up Echocardiographic Data for 38 Patients Undergoing the Modified Morrow Procedure*

Echocardiography Data	Preoperative	Postoperative	t	P
LA diameter, mm	35.4 ± 6.6	35.3 ± 6.5	0.036	.9715
LVEDD, mm	36.1 ± 4.4	35.6 ± 4.8	0.515	.6085
IVS thickness, mm	11.8 ± 3.2	11.5 ± 2.5	0.453	.6522
PWLV thickness, mm	14.7 ± 3.8	14.5 ± 3.9	0.154	.8782
LVOTG, mm Hg	17.9 ± 12.9	15.9 ± 11.7	0.688	.4935
LVEF, %	67.6 ± 7.4	71.9 ± 8.1	2.360	.0211

*Data are presented as the mean SE of the mean. Abbreviations are expanded in the footnote to Table 1.

RESULTS

All patients were discharged without complications. Intraoperative TEE examinations showed that the mean systolic gradient between the LV and the aorta decreased from 94.8 ± 35.6 mm Hg preoperatively to 13.6 ± 10.8 mm Hg ($P = .0000$) after the operation. The mean thickness of the ventricular septum decreased from 28.3 ± 7.9 mm to 11.8 ± 3.2 mm ($P = .0000$). TTE also revealed that the mean systolic gradient between the LV and the aorta decreased from 89.3 ± 31.1 mm Hg preoperatively to 17.9 ± 12.9 mm Hg ($P = .0000$) by 1 to 2 weeks postoperatively. The mean times of cardiopulmonary bypass and aortic occlusion were 83.6 ± 18.3 minutes (range, 63–129 minutes) and 53.3 ± 15.1 minutes (range, 32–86 minutes), respectively. The mean duration of intensive care unit stays was 1.8 ± 0.4 days (range, 1–2 days). Mitral regurgitation and SAM of the anterior leaflet of the mitral valve were significantly reduced or eliminated. During the follow-up, all patients promptly became completely asymptomatic or complained of mild effort dyspnea only, and syncope was abolished. TTE examinations showed that the postoperative pressure gradient either remained the same or was diminished. Tables 1 and 2 summarize the pre- and postoperative echocardiographic data. The surgery led to significant changes in the septal thickness, the LVOT gradient, the size of the left atrium, and the LV ejection fraction (LVEF). Left bundle branch block or left anterior hemiblock were found postoperatively in 17 cases, and atrial fibrillation was found before and after surgery in 9 of the same cases. No iatrogenic ventricular septal perforation or complete AV block was found in any patient postoperatively.

The mean follow-up time was 30 months (range, 6–100 months). There was no long-term operative mortality. The patients experienced an evident improvement in functional capacity: At the last follow-up, 31 cases were in NYHA functional class I, and 7 cases were in functional class II. In 11 cases, use of β -blockers was continued postoperatively. All patients promptly became completely asymptomatic or complained of mild effort dyspnea only, and syncope was abolished. Three patients had mild effort dyspnea only. TTE examinations showed that the pressure gradient either remained the same or diminished and that LVEF values were increased slightly (Table 3). Mild SAM of the anterior leaflet of the mitral valve was found in 6 cases, and mild mitral regurgitation occurred in 7 cases during follow-up, similarly to the early stage after surgery. Left bundle branch block or left anterior hemiblock was found in 17 cases, chronic and paroxysmal atrial fibrillation was found in 1 case each, and no severe AV block occurred in any of the patients.

DISCUSSION

Hypertrophic cardiomyopathy is a complex disease, owing to its morphologic, functional, and clinical heterogeneity, and it affects approximately 1 in 500 individuals (0.2%). Only 25% of patients present significant hemodynamic obstruction of the LVOT and therefore belong to the HOCM subgroup of hypertrophic cardiomyopathy, which is also called

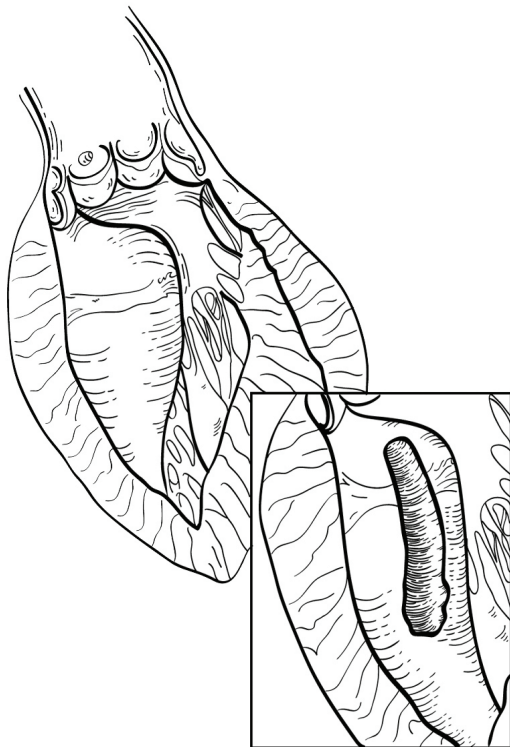
idiopathic hypertrophic subaortic stenosis [Smedira 2008; Wang 2009]. HOCM is characterized by an asymmetrical septal hypertrophy that causes an LVOT gradient. Severe LVOT obstruction can cause dyspnea, chest pain, and syncope, and it predisposes the patient to the development of progressive congestive heart failure, embolic stroke, sudden death, and related high-risk events [Maron 2002, 2004]. The clinical diagnosis of HOCM is established most easily and reliably by significant symptoms and a 2-dimensional echocardiography evaluation, which reveals a hypertrophy of the myocardium that is usually asymmetric, with the septal thickness being greater than that of the free wall.

Pharmacologic therapy is the first-line approach to the relief of symptoms in the management of HOCM. Patients with severe symptoms refractory to pharmacologic treatment and a marked outflow obstruction (outflow gradient 50 mm Hg under baseline conditions) are candidates for surgical intervention [Maron 2003b, 2004; Brown 2008; Smedira 2008]. Young asymptomatic or mildly symptomatic patients with particularly marked outflow obstruction (e.g., 75-100 mm Hg or greater at rest) should undergo surgery to decrease the risk of sudden death [Spirito 2006]. If the patient has atrial fibrillation, surgical intervention may be considered. Patients with severe heart failure-related symptoms, whose outflow gradient may decrease to <50 mm Hg, are still candidates for surgery. Old age and severe symptoms are high-risk factors but are not contraindications for surgery. In our study, the LVEF value of 1 patient with heart failure was only 31% before the operation and recovered to normal postoperatively. Some asymptomatic patients with severe mitral regurgitation

or structural abnormalities of the mitral valve may also be better surgical candidates [Kirklin 1993].

Generally speaking, options for surgical treatment include isolated septal myectomy, isolated MVR, and septal myectomy plus MVR [Maron 2004; Smedira 2008]. We think that ventricular septal myectomy with the modified Morrow procedure is mostly an effective method for patients with HOCM. Our surgical practice is as follows: A low-cut transverse incision is used in the aortotomy. Muscle resection begins below the right coronary cusp at the midpoint to the base of both papillary muscles in the middle and anterior direction without catching the anterior mitral leaflet, thereby eliminating SAM and decreasing the LVOT gradient [Maron 2004]. The width of the muscle resection is extended to the left side of anterior mitral leaflet (as far leftward as possible to the mitral valve; Figure 3B), with the depth of the muscle resection depending on the preoperative septal thickness. The septal myectomy with the modified Morrow procedure is extended far more distally than with the conventional Morrow procedure (Figure 3A). After the extended myectomy, a wider groove appears in the left side of the hypertrophic septum, and the obstruction of the LVOT and SAM are eliminated. Complete heart block (requiring a permanent pacemaker) and iatrogenic ventricular septal perforation are the primary complications that can occur during the operation [Maron 2006, 2007; Smedira 2008] and are important factors that make the technique unpopular. We believe that good surgical exposure of the outflow tract is of paramount importance for avoiding these complications. In our practice, an aortotomy with

A.



B.

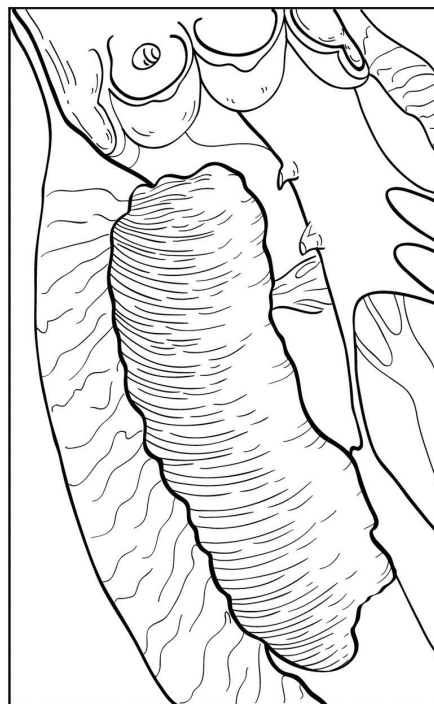


Figure 3. A, Narrow groove in the classic Morrow procedure. B, The septal resection extended, widened, and extended far more distally in the modified Morrow procedure.

a low-cut transverse incision is used. At the same time, the headstock of the operation table is elevated during the operation, and a special slender retractor is inserted into the LVOT through the aortic annulus for traction. The free wall of the right ventricle is then pressed toward the septum by the assistant. If necessary, the aorta is cut off completely to optimize visualization of the hypertrophic septum and the obstructed LVOT and to explore the pathologic anatomy, such as the site and extent of the hypertrophic septum and the mitral valve malformation. In our study, the aorta was cut off completely in 9 cases to optimize visualization of the septum (Figure 2). The depth of muscle resection depends on the preoperative septal thickness, so precise assessment of septal thickness by echocardiography is vital to determine the extent and the site of the myectomy and to avoid iatrogenic perforation of the ventricular septum. Moreover, to prevent complete heart block, the surgeon should not extend the septal myectomy to the right and beyond the midpoint of the right coronary cusp.

For patients without structural abnormalities of the mitral valve (prolapse, calcification, endocarditis), septal myectomy alone is enough to eliminate SAM and to significantly decrease or abolish mitral valve insufficiency. Additional mitral valve management is not imperative. Krajcer et al [1989] found no significant differences in therapeutic effect between septal myectomy and MVR by the 10-year follow-up, so they advocated septal myectomy alone. Myectomy plus MVR does not appear to offer any additional improvement in the LV outflow gradient at rest, the LV outflow gradient with provocation, NYHA functional class, operative mortality, or annual mortality. Therefore, MVR is not routinely recommended as a primary treatment for obstruction because of the potential postoperative complications related to durability, thromboembolism, and anticoagulation. Currently, most surgeons advocate myectomy alone for HOCM in the absence of intrinsic mitral valve disease, even in the presence of a relatively thin ventricular septum. A carefully performed surgical reduction of the septum is the preferred method [Maron 2003a]. We believe that extended septal myectomy with the modified Morrow procedure can markedly decrease the pressure gradient in almost all cases. In addition, MVR is not required for mild or moderate mitral valve regurgitation due to obstruction. MVR is recommended for patients with structural abnormalities of the mitral valve or severe mitral regurgitation.

Intraoperative TEE is very important for managing patients undergoing surgical myectomy, and its use is standard at centers experienced with myectomy [Krajcer 1989; Smedira 2008]. After induction of anesthesia before myectomy, TEE is useful for precisely assessing the site and thickness of the hypertrophic septal muscle and for evaluating the structural features of the mitral valve. Immediately after the procedure, TEE can access the surgical result of the myectomy for the surgeon, including any residual outflow tract gradient due to insufficient resection, any iatrogenic perforation of the ventricular septum due to excessive resection, SAM, and mitral regurgitation. If the results are not satisfactory, a more extensive myectomy should be performed. Thus, intraoperative use of TEE is necessary and is worth advocating for the surgery.

On the basis of the experience of a number of surgical centers throughout the world [Maron 2003a, 2006; Smedira 2008], the ventricular septal myectomy operation has become established as a proven approach for amelioration of outflow obstruction. It is the standard therapeutic option and the gold

standard for HOCM, both in adults and children and in cases of severe drug-refractory symptoms. Septal myectomy is performed to achieve an improved quality of life and an improved functional (exercise) capacity, with permanently eliminated outflow obstruction and mitral regurgitation. The surgical results exceed those achievable with chronic administration of cardioactive drugs [Maron 2004; Smedira 2008]. Some studies [Maron 2003a, 2007; Smedira 2008] have also shown that septal myectomy can reduce the long-term risk of sudden death and have demonstrated that long-term survival after myectomy is equivalent to that of the general population and better than that of patients with HOCM who do not undergo surgery [Maron 2003a]. Two recent treatment options, dual-chamber pacing and percutaneous alcohol septal ablation, have emerged as potential alternatives to surgery for select patients. Two recent meta-analysis studies [Alam 2009; Ashikhmina 2011] have shown that alcohol septal ablation produces results equivalent to septal myectomy at the short-term and mid-term follow-ups, while avoiding surgical trauma and being easily acceptable for the patients. A critical difference is that long-term follow-up data are not yet available for alcohol septal ablation, and there are concerns that the intramyocardial scar could provide a long-term arrhythmogenic substrate and that the extent of the myocardial damage may extend away from the target area and lead to further undesirable remodeling [Watkins 2005]. In our group, 6 patients failed alcohol septal ablation before surgical treatment. At the present time, septal myectomy remains the gold standard treatment for HOCM [Maron 2006, 2007; Brown 2008; Smedira 2008].

In conclusion, extended septal myectomy (modified Morrow procedure) is a safe, reliable, and effective method of eliminating LVOT obstruction in HOCM. Good surgical exposure for the outflow tract and thorough resection of the hypertrophic septum are of paramount importance for a successful surgery.

REFERENCES

- Alam M, Dokainish H, Lakkis NM. 2009. Hypertrophic obstructive cardiomyopathy—alcohol septal ablation vs. myectomy: a meta-analysis. *Eur Heart J* 30:1080-7.
- Ashikhmina EA, Schaff HV, Ommen SR, Dearani JA, Nishimura RA, Abel MD. 2011. Intraoperative direct measurement of left ventricular outflow tract gradients to guide surgical myectomy for hypertrophic cardiomyopathy. *J Thorac Cardiovasc Surg* 142:53-9.
- Brown ML, Schaf HV. 2008. Surgical management of obstructive hypertrophic cardiomyopathy: the gold standard. *Expert Rev Cardiovasc Ther* 6:715-22.
- Kirklin JW, Barratt-Boyes BG. 1993. *Cardiac surgery*. 3rd ed. New York, NY: Churchill Livingstone. p 1717.
- Krajcer Z, Leachman RD, Cooley DA, Coronado R. 1989. Septal myotomy-myomectomy versus mitral valve replacement in hypertrophic cardiomyopathy. Ten-year follow-up in 185 patients. *Circulation* 80(Pt 1):157-64.
- Maron BJ. 2002. Hypertrophic cardiomyopathy: a systematic review. *JAMA* 287:1308-20.
- Maron BJ. 2007. Controversies in cardiovascular medicine. Surgical myectomy remains the primary treatment option for severely symptomatic patients with obstructive hypertrophic cardiomyopathy. *Circulation* 116:196-206.
- Maron BJ, Dearani JA, Ommen SR, et al. 2004. The case for surgery in obstructive hypertrophic cardiomyopathy. *J Am Coll Cardiol* 44:2044-53.
- Maron BJ, McKenna WJ, Danielson GK, et al. 2003. American College of Cardiology/European Society of Cardiology clinical expert consensus

document on hypertrophic cardiomyopathy. A report of the American College of Cardiology Task Force on Clinical Expert Consensus Documents and the European Society of Cardiology Committee for Practice Guidelines. *Eur Heart J* 24:1965-91.

Maron MS, Olivotto I, Betocchi S, et al. 2003. Effect of left ventricular outflow tract obstruction on clinical outcome in hypertrophic cardiomyopathy. *N Engl J Med*;348:295-303.

Maron MS, Olivotto I, Zenovich AG, et al. 2006. Hypertrophic cardiomyopathy is predominantly a disease of left ventricular outflow tract obstruction. *Circulation* 114:2232-9.

Morrow AG, Fogarty TJ, Hannah H 3rd, Braunwald E. 1968. Operative treatment in idiopathic hypertrophic subaortic stenosis. Techniques and the results of preoperative and postoperative clinical and hemodynamic assessments. *Circulation* 37:589-96.

Ommen SR, Maron BJ, Olivotto I, et al. 2005. Long-term effects of surgical septal myectomy on survival in patients with obstructive hypertrophic cardiomyopathy. *J Am Coll Cardiol* 46:470-6.

Smedira NG, Lytle BW, Lever HM, et al. 2008. Current effectiveness and risks of isolated septal myectomy for hypertrophic obstructive cardiomyopathy. *Ann Thorac Surg* 85:127-33.

Spirito P, Autore C. 2006. Management of hypertrophic cardiomyopathy. *BMJ* 332:1251-5.

Wang S, Cui B, Sun H, et al. 2009. Clinical experience of surgical treatment on hypertrophic obstructive cardiomyopathy [in Chinese]. *Natl Med J China*. 89:2776-8.

Watkins H, McKenna WJ. 2005. The prognostic impact of septal myectomy in obstructive hypertrophic cardiomyopathy. *J Am Coll Cardiol* 46:470-6.