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Surgical outcomes and strategy of hypertrophic obstructive cardiomyopathy

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Abstract: Objective: To evaluate the surgical clinical results of hypertrophic obstructive cardiomyopathy. Methods: We retrospectively collected data on 24 patients who underwent surgical management in the past ten years in two hospitals in China and Madras Medical Mission in India. Myomectomy was carried out on all patients. Among them 3 patients underwent mitral valve replacement; 2 patients underwent mitral valve repair (anterior mitral leaflet plication); 2 patients underwent aortic valve replacement; 1 patient underwent aortic valve repair; 2 patients underwent aortic root replacement; 1 patient underwent Bentall's procedure and 1 patient underwent coronary artery bypass grafting because of a breached muscle bridge. Results: One patient died of post-operative heart failure. The mean follow-up time was 4.3 years. There was significant improvement in the symptomatic status. Sixteen patients were asymptomatic with good effort tolerance and only four patients had New York heart association (NYHA) Classes I~II due to associated valvular lesions. Conclusion: Our experience proved that symptomatic hypertrophic obstructive cardiomyopathy or non-symptomatic hypertrophic obstructive cardiomyopathy with combined heart disease is indication for surgery as surgical intervention could get better clinical results in this kind of patients compared with other non-surgical method because it beneficially reduces the systolic anterior motion (SAM) of the mitral valve leaflet, which could not be avoided by other non-surgical treatment.

Key words: Hypertrophic obstructive cardiomyopathy, Systolic anterior motion, Myomectomy
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INTRODUCTION

Hypertrophic obstructive cardiomyopathy (HOCM) is a condition of unknown origin characterized by septal hypertrophy and sudden death. This obstruction is associated with abnormal systolic anterior motion (SAM) of the mitral leaflet. Treatment of this condition includes beta and calcium channel blocker, antiarrhythmics, dual chamber pacing and percutaneous catheter-based ablation (Ivens, 2004). Surgical treatment for HOCM includes extended myectomy, plication of anterior mitral leaflet, mobilization of papillary muscles and mitral valve replacement (MVR). Such procedures should be performed at highly specialized and experienced centers, because they present inherent risks for ventricular septal defect and incomplete relief (Ivens, 2004; Roberts and Sigwart, 2005; Ferrari and Rydén, 2001). We colleted here data on 24 patients who underwent this procedure in Zhong Da and Su Bei hospital in China and Madras Medical Mission in India over the past 10 years period. All patients had been followed up in the post-operative period by echocardiography and clinical examination to determine the effectiveness of this procedure.

MATERIALS AND METHODS

General patients characteristics

From August, 1994 to March, 2004, 24 patients underwent surgical treatment for HOCM. Their age

ranged from 18 to 55 years and there were 17 males and 7 females in the group. Pre-operatively 10 patients had dyspnoea on exertion, 4 patients had palpitations, 3 patients had angina on exertion (Table 1). In all the patients, preoperative septal thickness was ≥20 mm measured by echocardiography. Preoperatively 8 patients showed SAM of mitral valve leaflet, with mitral valve regurgitation Grade I in 1 patient, Grade II in 4 patients and Grade IV in 3 patients. Resting left ventricular outflow tract gradient was 104.7 mmHg (mean). The patients presented the following symptoms.

Table 1 The symptom patients presented

Symptom	Number (%)
Dyspnoea on exertion	10 (41.6%)
Palpitations	4 (16.6%)
Angina on exertion	3 (12.5%)
Tachypnea	3 (12.5%)
Cardiac arrhythmias	13 (50%)
Exertional dyspnea and one or more syncope	6 (25%)
Frequent attacks of dizziness and vertigo	13 (50%)

Operative details

All the patients underwent operation with the use of cardiopulmonary bypass (CPB), antegrade cold blood cardioplegia and vent through right upper pulmonary vein. All the patients underwent trans aortic myomectomy. The first septal incision was kept at the level of the right wall. Myomectomy was carried out on hypertrophied septum with sharp dissection. Papillary muscle mobilization was also performed whenever papillary muscles adhered to the left ventricular wall or septum. In addition to myomectomy, 3 patients underwent mitral valve replacement; 2 patients underwent mitral valve repair (anterior mitral leaflet plication); 2 patients underwent aortic valve replacement; 1 patient underwent aortic valve repair; 2 patients underwent aortic root replacement; 1 patient underwent Bentall's procedure and 1 patient underwent coronary artery bypass grafting (CABG) due to a breached muscle bridge (Table 2). Sixteen patients underwent intra operative trans esophageal echocardiography. Most patients were put on atrium and ventricular pacing wires for atrioventricular sequential pacing if required.

One case underwent the repair of the aortic valve due to damage to its valve cusp during the trans aortic approach. Replacement of the aortic valve was done in patients who presented associated bicuspid aortic valve, stenosis or aortic regurgitation or both. Aortic root replacement with a 21 mm pulmonary homograft was done in a patient who presented dilated aortic root. Bentall's procedure was done in one patient who presented pseudoaneurysm of the ascending aorta. A breached muscle bridge was present in one patient for whom coronary artery bypass grafting was performed. The patients who presented heart block underwent permanent pacemaker implantation.

Table 2 Associated procedure with septal myectomy

Associated procedure	Number of patients
Mitral valve replacement	3
Aortic valve replacement	2
Aortic valve repair	1
Aortic root replacement	2
Bentall's procedure	1
Coronary artery bypass grafting	1
Permanent pacemaker implantation	5

RESULTS

One patient expired in the immediate postoperative period due to low cardiac output. The rest of the patients had an uneventful recovery following surgery. Among them 7 patients had NYHA Class II when they discharged, 4 of the 23 patients had mild systolic murmur. The mean resting left ventricular outflow tract gradient was 21.6 mmHg after operation (P<0.01). Twenty patients came for follow-up after 6 months. The mean follow-up time was 4.3 years. There was significant improvement in the symptomatic status. Sixteen patients were assymptomatic with good effort tolerance and only 4 patients had NYHA Classes I~II symptoms due to associated valvular lesions during follow-up. SAM disappearance after operation in all patients was confirmed by echocardiography.

DISCUSSION

Hypertrophic obstructive cardiomyopathy is a disease with high rate of morbidity and mortality. Our understanding of the pathophysiology of obstruction

in hypertrophic cardiomyopathy has improved since initial descriptions in the late 1950s. In the classic form of HOCM, the hypertrophy is maximal in the cephalad portion of the ventricular septum and lies just apical to the free edge of the anterior mitral leaflet in its open position. The hypertrophied septum bulges into both left and right ventricular to obstruct flow. The diffuse muscular prominence usually tapers off gradually toward the aortic annulus and apex. Occasionally, the hypertrophy is maximal at a site adjacent to the papillary muscles, producing mid-cavity obstruction contributed by papillary muscles and free wall hypertrophy. Obstructed patients have more severe symptoms and their systolic murmurs often bring them to medical attention. Regardless of presence of symptoms, these patients with HOCM are at high risk of sudden death. Five risk markers were identified: family history of sudden death; unexplained syncope; non-sustained tachycardia on Holter monitoring; abnormal exercise blood pressure response; and severe left ventricular hypertrophy (>3 cm). Annual sudden death rates are at least 2% for patients who present two or more of these risk markers (Sigwart, 2001; Ferrari and Rydén, 2001).

Surgical intervention is indicative in patients with severe left ventricular outflow tract gradient and symptoms refractory to medical therapy. Surgery to relieve obstruction in HOCM can be among the more technically challenging cardiac operations for acquired disease (Cleland, 1963) began the surgical treatment of HOCM with myotomy or limited excision myectomy through the trans aortic approach as early as 1958. Though remarkable reductions in gradient were observed in the majority of myotomy patients, in-hospital mortality was high and in some patients obstruction persisted after operation. Morrow's modification of the initial procedures, the wider extensive trough myectomy, consistently decreased obstruction and has remained the standard operation because of its long successful track record (Morrow *et al*., 1968). In publications of results of myectomy from 1987 to 2003, mortality within 1 month of operation ranged from 0% to 6%, our clinical result is similar to the reported results from other heart centers (Yoerger and Weyman, 2003).

The concept of the cause of obstruction, changes from early ideas that a muscular outflow tract sphincter was the cause, through the discovery of systolic anterior motion of the mitral valve leaflet, to current understanding that flow drag, the pushing force of flow, is the dominant hydrodynamic mechanism for SAM. Accordingly, the continuing redesign and modification of surgical procedures to relieve outflow obstruction have corresponded to ideas about the cause of this condition (Yoerger and Weyman 2003; Ommen *et al*., 2005; van der Lee *et al*., 2003).

Anterior mitral leaflet plication

The mitral valve is often enlarged in both area and length in obstructive HOCM, especially in its relationship to the small left ventricular cavity, this disparity is best evaluated by preoperative TEE because it is difficult to completely assess this after cardiopulmonary bypass is initiated. Mitral valve replacement or repair is a consideration in HOCM when septal thickness is less than 18 mm or structural mitral valve abnormality is present. Plication of the native anterior mitral leaflet decreases the size of the leaflet and attendant drag forces and reduces chordal and leaflet slack (Cooley, 1991). In this group, we performed 2 cases with anterior mitral leaflet plication avoiding the burden of prosthetic valve and its life-long risks of valve failure, embolism, infection, and warfarin-induced hemorrhage.

Mobilization and partial excision of the papillary muscles

This approach severs the abnormal connections that bind the papillary muscles to the anterior wall. After the septum is resected, it is easier to visualize structures within the ventricular cavity, abnormal attachments between the papillary muscle and the anterior lateral ventricular wall should be divided and a portion of the junction of the papillary muscle and lateral wall is also resected. The same is often done for the posterior papillary muscle. When this resection is completed, the papillary muscles, with their diameters reduced, are separated from the wall and from each other. This allows the papillary muscles to assume a more posterior position in the left ventricle and does not appear to compromise papillary muscle function in respect to mitral valve closure (Cooley, 1991). Complete mobilization is a most essential step in the relief of SAM. McIntosh *et al*.(1992) reported on 36 selected patients with anterior leaflet plication combined with myectomy because they were judged

morphologically during operation to be at risk for residual SAM and obstruction. In this select group of sicker patients, echocardiographic fellow-up showed that postoperative pressure gradient of left ventricular outflow was reduced to mean of 16 mmHg and overall mitral regurgitation was reduced. Echocardiographic studies revealed that plication appeared to limit anterior leaflet motion and SAM (Marwick *et al*., 1992). Schoendube *et al*.(1995) reported their experience with extended myectomy and mobilization and partial excision of the papillary muscles in 58 patients. There were no peri-operative deaths. No patient had postoperative SAM, gradient, significant mitral regurgitation, or ventricular septal defect.

In recent years, non-surgical interventions to relieve obstruction have been advanced. The main benefit of these interventions over surgery is avoidance of sternotomy and cardiopulmonary bypass. Firstly, DDD pacing with short atrioventricular delay to assure complete ventricular paced activation has been reported to reduce resting pressure gradient of left ventricular outflow by approximately 50% (Megevand *et al*., 2005). However, patients are left with residual pressure gradient of left ventricular outflow from 30 to 48 mmHg on average, generally higher than that found after successful surgery. Secondly, it could not offer improvement in exercise capacity and increase in exercise time or maximal oxygen consumption. Only in the subgroup of patients more than 65 years old did a higher proportion acquire the benefit of both reduction in symptoms and increase in exercise capacity. Thus, pacing cannot be considered a primary treatment for obstructive HOCM. Nevertheless, DDD pacing is still applied in patients refractory to medication who are elderly, have contraindications, or do not want surgery (Chang *et al*., 2003).

Alcohol ablation, a percutaneous catheter-based method to decrease septal thickness by therapeutic infarction, was introduced by Sigwart (2001) and Ivens (2004). After occlusion of a septal perforator by a small balloon to prevent back leakage, 1 to 4 ml, of absolute alcohol is injected into the distal perforator. The balloon is left inflated for 5 to 10 min to prevent back leakage of alcohol. Patients experience chest pain and modest myocardial infarction with CK elevations. In two comparisons of septal ablation and surgical myectomy, pressure gradient of left ventricular outflow reduction and symptom relief were similar in the two treatments, the modalities were nearly the same (van der Lee *et al*., 2005; Ralph-Edwards *et al*., 2005; Yacoub, 2005). In the report of Ralph-Edwards and coworkers, patients were selected for alcohol ablation if they were older or had fewer combined conditions. Follow-up pressure gradients of left ventricular outflow were lower in the surgically treated patients and the need for permanent pacing was again greater in the ablated group. And no mistake when the patients suffer from combined disease such as valve abnormality or coronary artery disease, surgery has more advantage than alcohol ablation. In our group, 14 cases underwent the combined surgical procedure.

There is currently no method to monitor the extent of resection while the patient is on cardiopulmonary bypass. Intra-operative echocardiography is useful to ensure adequate repair after cardiopulmonary bypass and myectomy. Trans esophagus echocardiography (TEE) offers excellent imaging and has the advantage of having the probe out of the operative field and has generally supplanted epicardial imaging. Persistent early SAM, with resting outflow gradient of more than 50 mmHg, or more than moderate-mitral regurgitation should be in prompt immediate investigation. Using these criteria Marwick and coworkers reported that 20% of patients were placed back on heart-lung bypass and TEE was used again to guide the location of additional resection. After the patient is taken off bypass we give intravenous inotropic agents to exclude provocable obstruction. Because patients are already vasodilated due to rewarming and are hypovolemic, at this time we follow the latter approach: patients with resting gradients more than 30 mmHg or with post-peripheral vaso-constriction (PVC) provoked gradients more than 50 mmHg are placed back on heart-lung bypass.

Indications for surgical intervention for symptomatic HOCM in infancy and childhood are not well defined (Minakata *et al*., 2005) because of the rarity of the lesion and the atypical presentation. In HOCM, even more so than in discrete sub-aortic stenosis, extensive endocardial and myocardial resection through a trans aortic approach presents special problems in infants and children because of limited exposure through the aortic valve annulus and the inherent risk of injury of the conduction tissue, the

aortic and mitral valves, the papillary muscles, and the major coronary artery branches as well as potential for creation of an iatrogenic ventricular septal defect. Congestive heart failure with an important gradient across the left ventricular outlet tract (LVOT), or visual evidence of obstruction in echocardiographic or angiographic studies, especially if located in the cephalad portion of the ventricular septum, is probably an indication for surgical intervention for children. Negatively, limited myectomy at the initial myectomy may cause recurrent left ventricular outflow tract obstruction in children, the obstruction may be ascribed to the septal hypertrophy at the midventricular level and anomalous papillary muscles (Minakata *et al*., 2005).

To conclude, the treatment of HOCM is one of the challenges in adult cardiac surgery, even though the surgical method is modified technically and has shown good clinical results. By now, the management of drug-refractory non-symptomatic HOCM is still under dispute. Our experience proved that symptomatic HOCM or non-symptomatic HOCM with combined heart disease is indication for surgery and that surgical intervention beneficially reduces the SAM, which could not be avoided by other non-surgical treatment.

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