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Contemporary Surgical Treatment for Hypertrophic Cardiomyopathy

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Abstract

Hypertrophic cardiomyopathy is the most common cause of sudden death in young athletes. Surgical septal myectomy is highly effective for the patients with hypertrophic obstructive cardiomyopathy, which is refractory to medical treatment. The perioperative mortality rate for isolated septal myectomy is less than 1% in high volume centers. The long-term outcomes have been reported to be outstanding with >90% of patients being free of significant symptoms and most being able to return to a normal lifestyle. There is a documented survival benefit after surgical septal myectomy. There is a wide variation of pathophysiology in hypertrophic cardiomyopathy including diffuse midventricular obstruction or subvalvular abnormalities. Several surgical approaches have been applied in accordance with the pathophysiology, such as transaortic, transapical, and transmitial septal myectomy. There is a controversy how to manage concomitant mitral valve regurgitation. The most recent Society of Thoracic Surgeons database showed that operative mortality of concomitant septal myectomy and mitral valve operations was double compared with isolated septal myectomy.

Keywords: hypertrophic cardiomyopathy, septal myectomy, surgical outcomes

1. Introduction

Hypertrophic cardiomyopathy is a genetic disorder of the heart muscle, resulting in a small left ventricular cavity and marked hypertrophy of the myocardium [1, 2]. Although many patients remain asymptomatic throughout life, some patients develop symptoms such as dyspnea,
angina, and syncope. Hypertrophic cardiomyopathy is the most common cause of sudden death in trained athletes [3].

The mainstay of therapy for hypertrophic cardiomyopathy has been the combination of lifestyle changes and medical therapy including beta blockers or calcium channel blockers. However, there remain patients who are refractory to medical treatment. For these patients, surgical septal reduction therapy, called septal myectomy, is indicated with class I indication [4]. When surgery is contraindicated because of serious comorbidities or advanced age, alcohol septal ablation is recommended with class IIa indication [4]. Dual-chamber pacing has also been used for the relief of outflow tract obstruction in patients whom both surgical septal myectomy and alcohol ablation are considered to have unacceptable risk. The treatment algorithm for the management of hypertrophic cardiomyopathy patients is shown in Figure 1.

Given the duration of experience and documented evidence of long-term outcomes, surgical septal myectomy is the preferred treatment for most patients who need invasive therapy. Operator and institutional experience is a key factor of successful surgical outcomes. The guidelines suggested that surgical treatment should be performed only by experienced operators, which were defined as an individual operator with a cumulative case volume of at least 20 procedures or an individual operator who is working in a dedicated hypertrophic cardiomyopathy program with a cumulative total of at least 50 procedures [4].

Compared with surgical septal myectomy in which the hypertrophic muscle is directly visualized and resected, successful alcohol ablation is dependent on the variable septal artery anatomy, which may not supply the targeted area of the septum in up to 20–25% [4].

Figure 1. Treatment algorithm for the management of hypertrophic cardiomyopathy patients. LVOT, left ventricular outflow tract. Figures 2–5 were cited from the study of Price et al, but Figure 1 was not cited from them.
2. Surgical septal myectomy

2.1. Historical perspective of septal myectomy

The attempts of surgical correction for HCM were started in the late 1950s. One of the earliest septal myectomies was performed by Cleland and colleagues in 1958. They reported a series of 12 cases of septal myectomy in 1964, and there were three operative deaths [5].

Septal myectomy is also called Morrow operation, as it was reported by Morrow and Brockenbrough in 1961 [6]. They reported two cases of successful subaortic ventriculomyotomy via a transaortic approach for severe symptomatic hypertrophic subaortic stenosis.

Kirklin and Ellis reported two cases of successful surgical relief of diffuse subvalvular aortic stenosis via a left ventriculotomy in 1961 [7].

2.2. Transaortic septal myectomy

The initial technique reported by Morrow and colleagues involved excising a rectangular segment of the septal myocardium beneath the right coronary cusp with approximately 3–4 cm long, 1 cm wide, and 1.5 cm deep [8].

In recent years, the standard transaortic procedure has evolved into an extended septal myectomy. Following the initiation of cardiopulmonary bypass, exposure of the left ventricle is obtained via an oblique aortotomy. The completed myectomy extends from the subaortic level, about 5 mm below the aortic annulus to the midventricular level, opposite the base of the anterior papillary muscle of the mitral valve, for a total length of about 7 cm (Figure 2).

Ashikhmina et al. reported the importance of direct intraoperative measurement of pressures in the left ventricle and aorta [9]. After cannulation of the aorta, a 2.5 inch, 22-gauge spinal needle is placed into the aorta close to the aortic cannula and another 3.5 inch, 22-gauge spinal needle is placed into the left ventricle through right ventricular free wall and septum.

Nguyen et al. reported that transaortic septal myectomy can be achieved for relatively thin ventricular septum (<18 mm) with low risk of iatrogenic ventricular septal defect (0.3%) [10]. Brown et al. reported that septal thickness or left ventricular mass was not associated with death [11].

2.3. Transapical septal myectomy

Apical hypertrophic cardiomyopathy is a relatively rare form of hypertrophic cardiomyopathy. Eriksson et al. reported that this type of hypertrophic cardiomyopathy is not associated with sudden cardiac death and has a benign prognosis; nevertheless, one of third of these patients experience cardiovascular complications such as myocardial infarction and arrhythmias [12].

For these patients, transaortic septal myectomy is not always effective. Schaff et al. described a technique for resection of hypertrophic apical myocardium via an apical ventriculotomy.
A 6-cm longitudinal incision is made lateral to the left anterior descending artery beginning at the apical dimple. The goal of apical resection is to sufficiently enlarge the left ventricular cavity and shaving of the excess papillary muscle can be carried out if necessary (Figure 3).

Kunkala et al. reviewed 56 patients with midventricular obstruction [15]. Septal myectomy was performed through transaortic approach in 5, transapical approach in 32, and a combination of these in 19. There were no complications unique to the apical incision, and 5-year survival rate was similar to the expected one. Hang et al. also reported the effectiveness of combined transaortic and transapical approach to complex septal hypertrophy [16].

2.4. Transmitral septal myectomy

Transmitral septal myectomy was first described in 1963 by Lillehei and Levy [17]. This approach is indicated for patients with diffuse hypertrophy extending to or below the papillary muscles with mid-ventricular obstruction, which is not amenable to repair via a transaortic myectomy. Gutermann et al. reported 12 cases of transmitral septal myectomy with one mortality [18]. In the transmitral myectomy, the anterior mitral leaflet is widely detached.
from commissure to commissure, but the commissures are left intact. That allows an easy myectomy toward the base of the anterior papillary muscle, with mobility fully restored (Figure 4). The abnormal chordae from the septum to the anterior papillary muscle can be divided. After all intraventricular repairs are complete, the continuity of anterior mitral leaflet was restored either with continuous suture or with augmentation using an autologous pericardial patch.

Transmitral approach may provide excellent exposure to septal hypertrophy in patients with a narrow aorta which limits the transaortic view [19].

There are several case reports of minimally invasive transmitral septal myectomy through right thoracotomy, either with video-assisted methods or robotic platform [20–23].

2.5. Surgical outcomes and complications

Maron et al. reported the operative mortality data from five high-volume centers in North America [24]. Over the 15-year period, 3700 isolated septal myectomy operations were performed, and operative mortality was only 0.4%. In the meantime, it is expected that the surgical expertise of the high-volume centers would be passed down to regional multidisciplinary centers as well [25].
Kotkar et al. reviewed over two decades of surgical experience at Mayo Clinic [26]. More than 3000 patients underwent septal myectomy, and risk of hospital death after isolated septal myectomy was <1%. Postoperative complications such as iatrogenic ventricular septal defect and complete heart block requiring permanent pacemaker occurred infrequently (0.3 and 2%, respectively).

Parry et al. reviewed one surgeon’s experience of septal myectomy for 211 patients at Toronto General Hospital [27]. The in-hospital mortality was 0.5% and 5-year survival rate was 98.1%.

According to the Society of Thoracic Surgeons database, 3452 septal myectomy operations were performed from July 2014 through December 2016, in the United States. Emergency status, endocarditis, aortic stenosis, and planned aortic valve operations were excluded, but concomitant coronary artery bypass was included. In the final cohort of 2038 patients, 1315 (65%) received septal myectomy alone, and 723 (35%) had septal myectomy with concomitant mitral operations. The median number of annual cases per center was 2 (range 1–435). Operative mortality and major morbidity were lower in isolated septal myectomy group than septal myectomy plus mitral operations (1.5% vs. 3.0%, p = 0.03; and 10.6% vs. 21.4%, p < 0.001, respectively). Postoperative iatrogenic ventricular septal defect and complete heart block requiring permanent pacemaker were rare (0.8 and 3.4%, respectively in isolated septal myectomy, and 1.8 and 4.1% in septal myectomy plus mitral operations) [28].

**Figure 4.** (a) View of the atrial surface of the mitral valve, with the dashed line representing the incision line in the anterior mitral valve leaflet. (b) Following incision and release of the anterior mitral valve leaflet, a broad view of the interventricular septum, down to the left ventricular apex, is obtained. Figure was cited from the study of Price et al. with their permission [49].

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2.6. Long-term outcomes of septal myectomy and its impact on cardiac function

Ommen et al. reviewed the experience of Mayo Clinic and studied the impact of surgical septal myectomy on long-term survival [29]. The 1-year, 5-year, and 10-year survival was 98, 96, and 83%, respectively, and they did not differ from those of the general population. They also stated that myectomy significantly improved all-cause mortality, obstructive cardiomyopathy-related mortality, and sudden cardiac death.

Woo et al. reviewed 338 patients who underwent septal myectomy at Toronto General Hospital and reported the excellent long-term survival with 1-year, 5-year, and 10-year survival of 98, 95, and 83%, respectively [30].

Deb et al. studied the impact of septal myectomy to left ventricular remodeling after surgery [31]. They found a significant decrease in the left ventricular mass index which occurred early after surgery and persisted beyond 2 years of follow-up.

Geske et al. reported that septal myectomy is associated with improvement in pulmonary hypertension, and it was most pronounced in patients with moderate or severe pulmonary hypertension [32].

In patients with hypertrophic cardiomyopathy, latent left ventricular outflow obstruction, which is defined as gradient <30 mmHg at rest and that increases to >50 mmHg with provocation, has been recognized important recently. Schaff et al. suggested that surgery should be offered to these patients [33].

2.7. Management of concomitant papillary muscle abnormalities

There is a subset of patients in whom the mitral valve and subvalvular apparatus such as papillary muscles and chordae play a significant role in creating the dynamic obstructive process [34–36]. Klues et al. reported that 66% of the patients with hypertrophic cardiomyopathy had a constellation of structural malformations, including increased leaflet area and elongation of the leaflets or anomalous papillary muscle insertion directly into anterior mitral leaflet, in mitral valve [34].

Minakata et al. reviewed 291 patients who underwent septal myectomy, and 56 (19.2%) had anomalous mitral subvalvular apparatus [37]. These anomalies were successfully treated with resection of anomalous chordae or relief of papillary muscle fusion, and no patients required mitral valve replacement.

Several reports described other surgical techniques to treat subvalvular abnormalities.

Redaelli et al. reported good outcomes of septal myectomy combined with papillary muscle repositioning for patients with abnormal papillary muscle morphology [38].

Ferrazzi et al. reported good outcomes of septal myectomy combined with a secondary chordal cutting for patients with thickened anomalous chordae [39].
2.8. Management of concomitant mitral regurgitation

Mitral valve leaflets play an important role in the pathophysiology of left ventricular outflow tract obstruction. Systolic anterior motion of the mitral apparatus narrows left ventricular outflow tract, and in many patients, leads to mitral regurgitation. There is a controversy how to treat concomitant mitral regurgitation at the time of septal myectomy. Ideally adequate septal myectomy can get rid of not only systolic anterior motion, but also mitral regurgitation. However, some groups have advocated that mitral valve replacement is necessary when mitral regurgitation is severe [40, 41].

Hong et al. reviewed the experience of septal myectomy operations at Mayo Clinic and concluded that mitral regurgitation related to systolic anterior motion of the mitral valve is relieved by septal myectomy alone in most cases, therefore, concomitant mitral operation is not necessary unless intrinsic mitral valve disease is present [42].

Balaram et al. suggested adding anterior mitral leaflet plication to septal myectomy when patients have long anterior leaflet (≥3 cm) and systolic anterior motion [43–45]. They call their method as “resection-plication-release” method (Figure 5). They have reported excellent outcomes using their technique.

Figure 5. (a) The resection-plication-release method identifies areas of left ventricular outflow tract obstruction, including the hypertrophic septum, abnormal papillary muscle attachments, and an elongated anterior mitral valve leaflet. (b) Following the resection-plication-release procedure, the hypertrophic septum is resected, the abnormal papillary muscle attachments are released, and the elongated anterior mitral valve leaflet is plicated. Figure was cited from the study of Price et al. with their permission [49].
As the Society of Thoracic Surgeons database showed, concomitant mitral operations were performed in one-third of the cases in the United States. Operative mortality of concomitant septal myectomy and mitral operations was double compared with isolated septal myectomy (3.0% vs. 1.5%). However, postoperative grade 3–4 mitral regurgitation was found more frequently in isolated septal myectomy than combined septal myectomy and mitral operations (10.6% vs. 5.8%, p < 0.0001). Following risk adjustment, the odds ratio for composite mortality and morbidity was not significant for mitral valve replacement vs. repair at 1.43 [0.9–2.2] (p = 0.0991) [28].

2.9. Recurrent obstruction

The recurrent obstruction after surgical myectomy is reported to be rare.

Minakata et al. reviewed 610 septal myectomies at Mayo Clinic between 1975 and 2003, and 13 patients underwent redo septal myectomy [46]. The interval between initial myectomy and redo myectomy ranged from 13 months to 11 years. The mechanism for recurrent obstruction was limited myectomy at the initial operation in 11, septal hypertrophy at the midventricular level in 8, and anomalous papillary muscle in 3.

Cho et al. also reviewed the surgical series at Mayo Clinic and stated that inadequate length of septal excision was associated with residual and recurrent obstruction [47].

Smedira et al. reviewed 323 patients who underwent isolated septal myectomy at Cleveland Clinic, and there were 10 cardiomyopathy-related reoperations; 4 redo myectomy, and 6 mitral valve procedure [48].

3. Conclusions

Surgical septal myectomy provides a survival benefit to patients with drug-refractory hypertrophic cardiomyopathy. It also has a positive effect on patients’ quality of life and cardiac function. According to the variation of pathology, appropriate surgical approach and technique should be applied. The recent study based on the national database showed surgical mortality doubles when concomitant mitral procedures are done at the time of septal myectomy. Careful decision making should be done in treating concomitant mitral regurgitation.

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