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Chapter

Surgical Treatment of Hypertrophic Obstructive Cardiomyopathy

Francesca Nicolo', Antonio Lio, Marina Comisso, Romina Pantanella, Roberto Scrofani and Francesco Musumeci

Abstract

Hypertrophic cardiomyopathy is a genetic disorder of the myocardium, characterized by marked myocardial hypertrophy that may lead to the development of symptoms such as dyspnea, angina pectoris, or stress-induced syncopes, with an increased risk of sudden cardiac death, due to obstruction of the left ventricular outflow tract (hypertrophic obstructive cardiomyopathy). Septal reduction treatment is needed in these patients, in order to relieve the symptoms. In addition, mitral valve apparatus should be assessed in these patients, in order to recognize a dynamic movement of the MV during systole anteriorly toward the LVOT. In this chapter, we will describe the current surgical management of HOCM.

Keywords: hypertrophic obstructive cardiomyopathy, HOCM, myotomy, SAM

1. Introduction

Hypertrophic cardiomyopathy (HCM) is a genetic disorder of the myocardium, characterized by marked myocardial hypertrophy (>15 mm) that cannot be explained by any other diseases that may cause secondary hypertrophy [1]. Its prevalence is 0.2–0.6% [2], with an overall annual mortality rate of 1%.

People with HCM often remain oligosymptomatic or even asymptomatic for many years; anyway, they may develop as cardinal symptoms dyspnea, angina pectoris, or stress-induced syncopes, with an increased risk of sudden cardiac death, particularly in young people and athletes [3], due to obstruction of the left ventricular outflow tract (LVOT). This obstruction is dynamic and largely influenced by changes in left ventricular loading and contractility, with subsequently increased left ventricular systolic pressure and secondary mitral regurgitation (MR), myocardial ischemia, and reduction in cardiac output. Around 70% of patients with HCM display relevant obstruction (HOCM) with a high peak pressure difference (more than 30 mm Hg) [4]; in half of these patients the obstruction is present even at rest, while in the remainder it is latent, so that a gradient can be demonstrated only under stress. Septal reduction treatments can lead to considerable relief of the symptoms even in patients whose obstruction is latent.

Echocardiography is an important and reliable method for diagnosing HOCM and for planning the kind of treatment. Echocardiography enables determination of the extent of the left ventricular hypertrophy and of the site, severity, and
mechanism of any obstruction. In addition, mitral valve (MV) apparatus can be adequately studied, in order to recognize any systolic distortion of the MV associated with systolic anterior motion (SAM) that often leads to secondary MR (Figure 1).

Despite medical therapy (treatment with B-blockers or, sometimes, Verapamil) is the first-line therapy for symptomatic patients with LVOT obstruction [1], a large proportion of patients with dynamic obstruction remain symptomatic so, a septal reduction treatment (SRT) is needed in order to relief of the symptoms. This can take the form of either septal myectomy or percutaneous alcohol ablation of the septum [5]. In this chapter we will describe the current surgical management of HOCM.

2. Anatomical classification of HCM

Morphology is variable, common variants being basal, mid-ventricular, apical, and diffuse types have been described. Maron classification reported 4 variants of HCM [6]:

- Type 1: hypertrophy (10% of patients) confined to the anterior portion of the ventricular septum;
- Type 2: hypertrophy (20% of patients), involves the anterior and posterior portion of the ventricular septum;
- Type 3: hypertrophy (52% of patients), involves the anterior and posterior portion of the ventricular septum as well as the lateral free wall;
- Type 4: hypertrophy (18% of patients), involves left ventricular regions other than the anterior septum and the posterior free wall.

Figure 1.
Transthoracic echocardiography showing LVOT obstruction (A), with a resting LVOT gradient of 77 mmHg (B) and severe MR, due to SAM (C). Figure courtesy of non-invasive cardiology diagnostic laboratory, Cardiology Department, L. Sacco Hospital, Via G.B. Grassi 74, Milano, Italy.
3. Surgical indication

ACC/AHA and ESC guidelines recommend that septal reduction therapy should be performed only by experienced operators, working as part of a multidisciplinary team expert, in the management of HCM [1, 7]. A SRT should be performed in presence of:

1. Significant LVOT obstruction (resting or maximum provoked LVOT gradient of ≥50 mmHg)
2. Angina, Dyspnea (NYHA functional class III-IV), Syncope
3. Persistent symptoms on maximal medical therapy.
4. The current indications have expanded recently to include symptomatic patients with low resting outflow gradients and latent obstruction.

4. Surgical approaches

1. Standard median sternotomy
2. Ministernotomy
3. Right anterolateral minithoracotomy (IV I.S.)

5. Surgical septal myectomy

Surgical septal myectomy is the therapeutic gold standard for the treatment of drug-refractory disabling symptoms in HCM caused by LVOT obstruction. This procedure can relieve hemodynamic disorders and has an acceptable surgical risk when performed on appropriate patients and in experienced centers [7].

It was performed, for the first time, in 1968 by Morrow et al. [8]; initially, the myectomy was limited in scope, but nowadays the resection is much more aggressive in terms of both width and length [5, 9]. Sparing the cardiac conduction system, septal myocardium is resected, depending on the area of hypertrophy and extending if necessary as far as the level of the papillary muscles, to an extent sufficient to eliminate the obstruction, retaining septal thickness of 1 cm. Surgical septal myectomy can be performed as follows:

5.1 Transaortic extended septal myectomy (TAESM)

TAESM through a transverse aortotomy, the aortic valve is totally exposed and the aortic leaflets are retracted so as to inspect the LVOT, the hypertrophic cardiac muscle and anterior MV leaflet. In order to open the LVOT and reduce the gradient to <30 mmHg, resection of the hypertrophic muscle until the thickness of the LV wall and interventricular septum became nearly normal by visual inspection, has to be performed. The myectomy should be initiated about 1.5 cm below the aortic annulus starting at the level of the non-coronary/right-coronary commissure to avoid the membranous septum and avoid creating a secondary ventricular septal defect (VSD). The septum is cut into as much as possible to relieve the obstruction and it is extended toward the left/non-coronary commissure so that the entire
The septum is trimmed off all around (Figure 2). The TAESM is the most commonly used technique and it has been associated with very low mortality, consistent alleviation of outflow gradients and excellent long-term survival [10].

5.2 Transmitral septal myectomy (TMSM)

The MV is approached through the left atrium, in the interatrial groove. Then, the base of the AL is widely detached from commissure to commissure, with the septum that lies just in front of the surgeon. The myectomy is continued further toward the apex with at least 1 cm depth of muscle removed. The anterior MV leaflet is then reconstructed (sometimes using an autologous pericardial patch with the size of the patch that should be as biggest as possible, in order to move posteriorly toward the left ventricle the coaptation plane). This approach is useful for those patients with diffuse hypertrophy extending to or below the papillary muscles with midcavity muscular obstruction: these patients are suboptimal candidates for the Morrow procedure. Benefits of the transmitial exposure include a wide view of the ventricular septum, absent risk of injury to the aortic valve cusps, and the opportunity to address concomitant abnormalities of the MV and subvalvular apparatus.

5.3 Complete septal myectomy by a double approach (aortic and mitral)

In patients with simultaneous MR due to SAM, the insufficiency is almost always reduced or eliminated by myectomy alone. Sometimes, there may be associated a lesion of the aortic valve and/or fibrous subaortic stenosis; these also require surgical correction, depending on their severity. Intraoperative echocardiography helps the surgeon to determine the individual extent of the disease and decide on the necessary scope of resection.
More difficult to treat are patients with an apical variant of HCM; they have obliteration of the left ventricular chamber with severe diastolic dysfunction. The results of myectomy depend essentially on the experience and competence of the surgeon. Only in specialized centers can myectomy be carried out with a risk of perioperative complications under 1%. The hemodynamic results are usually excellent with a postoperative gradient <10 mm Hg [11]. VSD, atrioventricular block and residual obstruction may complicate a septal myotomy; anyway, incidence is lower for experienced surgeons with a risk <3% in expert hands [12]. Long-term results following surgery are good, with more than 90% of patients being asymptomatic [13]. Septal myectomy is the gold standard in SRT, because of fewer complications and better freedom from redo procedures, with a better long term outcome rather than septal ablation.

6. Adjuncts to septal myectomy

6.1 SAM correction

SAM describes the dynamic movement of the MV during systole anteriorly toward the LVOT. It occurs in 31–61% of HCM patients, and it is associated with resting LVOT obstruction in 25–50% of them [14]. SAM-mediated MR typically resolves with extended myectomy alone, anyway, a well comprehension of all the mechanisms that can predispose to or precipitate SAM, is important, in order to treat this mechanism. Factors predisposing to SAM are:

a. structural anomalies such as:

- small left ventricular chamber
- bulging septum
- chordal anomaly
- papillary muscle displacement
- redundant anterior or posterior leaflets

b. geometric factors such as:

- annular undersizing
- anterior displacement of MV (any anatomical or surgical translocation of the MV anteriorly will increase the forces acting to draw the MV anteriorly that may precipitate SAM)
- low anterior—posterior (A-P) length ratio (A-P leaflet length ratio <1.3 is a risk factor for SAM)
- reduced mitral-aortic angle (<15°)
- distance reduction between MV coaptation point and septum
c. kinetic factors such as

- hyperdynamic left ventricle (LV)

Surgical techniques proposed to correct concomitant SAM in HOCM are:

- Changes to posterior leaflet height, in order to avoid anterior shifting of the coaptation point closer to the base of the anterior leaflet, which predispose to SAM [15]. The resulting posterior mitral leaflet (PML) should have a reduced height of 1 cm:
  - triangular resection of PML and ventricularization [16];
  - sliding posterior leaflet plasty technique (moves the coaptation point posteriorly) [17] (Figures 3–5);
  - modified sliding leaflet technique (middle scallop of PML is resected, differs from Carpentier in eliminating triangular resection) [18] (Figure 6);
PML folding plasty [19];

- Loop technique (height adjustment of the mitral leaflets using artificial chordae: after artificial chordae and annuloplasty ring placement in the normal way, a polyester reduction suture with a spaghetti loop supporting it is placed on the posterior leaflet surface, and tied down until its height is <1.5 cm), [20] (Figure 7);

- chordal translocation (PML secondary chordae are transected and moved to the AML) [21];

- edge-to-edge, or Alfieri stitch technique (A1-P1 stitch instead of midline) [22].

- Changes to anterior leaflet height, in order to reduce the redundant leaflet and, subsequently, the A-P length ratio (risk factor for SAM development):
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- Elliptical excision of the anterior mitral leaflet (excision site is then closed with double-layer prolene stitches) [23];
- Elliptical excision with repositioning of major basal chords [24];
- Removal of accessory chordae that may tether the AML to the septum, resulting in SAM [25].

- Annuloplasty ring sizes: any reduction in annular circumference reduces the mitral—aortic angle, which increases the risk of SAM, so an annular undersizing should not be performed, in order to maintain the coaptation plane as much as possible away from the septum:
  - Annular enlargement (post-MV repair, especially in Barlow’s disease): 36–40 mm annular rings are used in presence of excessive mitral tissue [26];
  - Annular plication: plication sutures are used to tie down the PML to the annulus [27];

  - “Resection—plication—release”: in HCM, the concomitant use of myectomy, plication of the AML and release of the papillary muscles remodels the LV in order to reduce SAM [28].

  - Mitral valve replacement: if SAM is severe and persistent despite exhaustive medical and surgical attempts at eliminating SAM, MV replacement can be the only option. In this setting, care must be taken to properly resect any remaining mitral tissue, to prevent SAM recurring.

6.2 Arrhythmia surgery

Patients with HCM are at increased risk of development of atrial fibrillation due to long-standing elevation of the left ventricular end-diastolic and left atrial pressures with subsequent left atrial chamber enlargement. Anyway, there is little evidence to support the addition of the Cox-maze procedure to septal myectomy.
6.3 Management of papillary muscle abnormalities

Sometimes, LVOT obstruction is present after septal myectomy because of anomalies of the (MV) valve apparatus: an accessory papillary muscle that arises from the ventricular septum and that is attached to the side of the anterior leaflet, may be present. In these patients, the accessory muscle has to be excised in its entirety. Other anomalies of the mitral apparatus may be the presence of accessory papillary muscles, or fusion of the anterior papillary muscle with the ventricular septum or left-ventricular free wall, or the presence of abnormal chordae tendineae that attach to the ventricular septum or to the free wall; all of this abnormalities may contribute to a persistent LVOT gradient and should be corrected in order to perform an adequate SRT.

In HCM, the anterior and inward displacement of papillary muscles is thought to create diastolic downwards vortex forces which pull the MV into the LVOT [29].

6.4 Management of concomitant MV disease

Degenerative MV disease requires attention at the time of myectomy in contrast to SAM-mediated MR that typically resolves with extended myectomy alone.

In our experience, when a concomitant mitral valve repair (MVR) is needed, we use a flexible posterior band that is slightly upsized, in order to minimize or avoid potential development of SAM postoperatively.

7. Other indications for septal myectomy

Apical and midventricular variants of HCM are difficult entities to diagnose and treat medically, with the only alternative to myectomy being heart transplantation.

8. Septal myectomy versus alcohol septal ablation

Despite an extended surgical septal myectomy is considered the gold standard for managing symptomatic patients [1], percutaneous alcohol ablation of the septum has emerged as an alternative to surgical septal myectomy [30]. In this approach, alcohol is injected into the first septal perforator in order to create a localized myocardial infarction. The advantages of this non surgical procedures are a faster recovery with a subsequent quick return to daily lifestyle; anyway, the literature tends to support better long-term symptom relief in those patients who undergo septal myectomy [31] with a higher procedural success and a lower rate of complications when myectomy is performed in experienced centers compared with alcohol septal ablation. In addition, we have to keep in mind that young patients and patients with severe or relatively thin septal thickness and a very high LVOT gradient are considered poor candidates for the percutaneous approach. Importantly, surgical septal myectomy also facilitates the correction of other abnormalities of the LVOT and repair of associated abnormalities of the MV and anomalous papillary muscles that can also contribute to residual dynamic outflow tract obstruction.

9. Conclusions

Due to the complex ventricular phenotype of septal hypertrophy which increases the drag forces acting on the MV, and also due to primary MV anomalies, a multifaceted approach to repair and abolition of LVOT obstruction is required.
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in HOCM. Septal myectomy is fundamental and represent the first step to any of these. In addition, specific techniques for SAM in the context of HOCM have been described. Each technique proposed for the surgical correction of HOCM has evolved to meet a specific anatomical problem, so it is inappropiate to rate one surgical procedure as superior to another; anyway, repair which resects as little tissue as possible and that does not distort the anatomy significantly should be preferred. Surgeons must understand the anatomical cause of LVOT obstruction, and this should guide them to the choice of the technique to adopt. Ease of repair should be also considered, as this will have beneficial consequences for the total time under cardiopulmonary bypass.

Alcohol septal ablation for HCM has been proposed as a less-invasive alternative to surgical myectomy, although its role in the management of HCM associated with SAM requires further investigations so that, the current evidence, supports the use of septal myectomy in the clinical practice [32].

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Conflict of interest

None.

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