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Surgery for Hypertrophic Obstructive Cardiomyopathy

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Hypertrophic obstructive cardiomyopathy (HOCM) is an inherited disorder with variable expressivity, resulting in asymmetric septal hypertrophy and left ventricular outflow tract obstruction (LVOTO). Symptoms are similar to those of aortic stenosis and are due to LV diastolic dysfunction and myocardial ischemia in the absence of epicardial coronary narrowing. Goals of treatment include symptomatic control, resolution of hemodynamic abnormalities and their sequelae, reduction of sudden cardiac death risk and screening of family members. Left ventricular outflow tract obstruction at rest is a predictor of severe symptoms, heart failure, and death. The majority of patients are managed medically, predominantly with b-blockers and calcium channel blockade. Disopyramide, a negative inotrope, can be used in cases with persistently high resting gradients. To prevent sudden cardiac death, implantable cardioverter-defibrillators are used aggressively [1].

Surgical treatment has been limited to patients with refractory symptoms and high resting gradients. Results of surgical intervention are well documented, with 95% of patients being asymptomatic with accompanying dramatic reduction in outflow tract gradient. Transcoronary ablation of septal hypertrophy with alcohol – *TASH procedure* - is a newer percutaneous technique. It is designed to ablate hypertrophied cardiac septal muscle through localized infarction, but its efficacy compared with that of surgical myectomy is uncertain. Surgery is not indicated in the absence of significant left ventricular outflow tract obstruction. Only those patients with symptoms refractory to medication and obstruction either at rest or with provocation are generally referred for surgery [2].

Several techniques for relief of obstruction in hypertrophic obstructive cardiomyopathy (HOCM) have been described ever since Cleland in 1958 started with transaortic myotomy, a procedure known better as *Bigelow technique*. Septal myectomy rather than simple myotomy was introduced by Morrow in 1961 and advanced over the years to the standard operation known as *Morrow procedure*. It was based on the assumption that the asymmetric septal hypertrophy was solely responsible for the left ventricular outflow tract obstruction [3].

Surgery for obstructive HCM has evolved over the past four decades, from ventricular septal myotomy (i.e., without muscular resection), to the classic Morrow myectomy. More recently, an extended and more extensive myectomy (up to about 7 cm long compared with 3 cm for the standard Morrow resection), combined with repair of mitral valve and submitral abnormalities, is practiced by some surgeons. Septal myectomy is performed through an aortotomy. A rectangular trough is created by first making two parallel longitudinal incisions in the basal septum near the nadir of the right coronary aortic cusp (RCC); septum beneath the commissure of the RCC and LCC. Incisions are extended distally and then connected transversely proximally below the aortic valve and distally just beyond the level of mitral-septal contact and

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subaortic obstruction (with standard Morrow myectomy) or to mid-ventricular level at the base of papillary muscles (with extended myectomy), yielding 3 to 12 grams of septal muscle. It has been prudent practice to perform myectomy under intraoperative transesophageal echocardiographic guidance to directly monitor the efficacy of the resection (to identify the level of obstruction and distribution of septal hypertrophy) and allow for possible surgical revision. Mitral valve repair, in addition to myectomy, may be most appropriate for selected patients with severe mitral regurgitation caused by primary valvular disease (e.g., myxomatous, rheumatic or ruptured chordae). Occasionally, if intrinsic mitral valve disease is of sufficient severity to preclude repair, or the proximal septum is only mildly thickened and the risks for either septal perforation (by excessive muscular resection) or residual post-operative obstruction (by inadequate resection) are increased, then replacement with a low-profile mitral prosthesis without myectomy may be prudent. Mitral valve replacement is, however, not routinely recommended as a primary treatment for obstruction, because of the potential postoperative complications related to durability, thromboembolism, and anticoagulation [1]. As an alternative to the transaortic approach, transatrial and transmitral myectomy has been attempted [4].

Occasionally, greatly elongated and flexible mitral leaflets will contribute substantially to the generation of mitral septal contact. In such selected cases, mitral valve plication combined with myectomy has been performed to restrict mitral valve motion and allow for more complete relief of subaortic obstruction and mitral regurgitation. Septal myectomy also offers an opportunity to repair associated major cardiac lesions such as atherosclerotic obstructive coronary artery disease or forms of fixed aortic stenosis, or surgically treat atrial fibrillation with the MAZE procedure.

Either alcohol ablation or myectomy offers substantial clinical improvement for patients with hypertrophic obstructive cardiomyopathy. Hemodynamic resolution of the obstruction and its sequelae is more complete with myectomy. Residual lesions after alcohol ablation might affect longer-term outcomes.

Septal myectomy is a safe, reliable, and durable method of eliminating LVOT obstruction in HOCM [5]. It improves functional status, reduces mitral regurgitation and normalizes life expectancy. It is the benchmark for ablative therapies and is our preferred method for treating this complex and fascinating disease. Earlier reports mentioned a mortality rate of 4%-6% [6,7], more recent studies report mortality rates of less than 2% [5,8]. The surgical results are much improved due to better anesthesia, myocardial protection during cardio-pulmonary bypass, and the use of intra-operative transesophageal echocardiography [9]. The latter permits a thorough evaluation of the surgical result upon the interventricular septum and, if required, the mitral valve, before the patient leaves the operating room. Complications of surgical myectomy include ventricular septal defect due to excessive removal of cardiac muscle, aortic regurgitation due to the transaortic approach and LBBB or complete heart block requiring a permanent pacemaker [2,10,11]. Alcohol ablation creates a strategically placed iatrogenic myocardial infarction (i.e., a scar). Currently published data suggest that procedural success is in the range of 75% to 80%, and among those successes, symptomatic improvement in the short-term and intermediate term is comparable with a myectomy [12]. However, despite the "less invasive" nature of this technique, the procedure-related morbidity and mortality are not lower than standard septal myectomy (SSM), and in some series they are higher. Importantly, there are no data indicating improved late survival after alcohol septal infarction. From a clinical investigation standpoint, it will be difficult for any procedure to show incremental benefit on the robust morbidity, mortality, hemodynamic, symptomatic, and survival benefits provided by SSM. Finally, among centers with substantial focus on HCM in which both the SSM and septal ablation are performed, the SSM myectomy is the preferred and proven therapy, whereas septal ablation is considered an alternative approach if surgical risk or other circumstances render surgery less attractive. A separate, but important consideration is risk of sudden death related to arrhythmia; although postoperative rates of sudden death are very low, risk is not zero, and patients should be evaluated longitudinally regarding the need for medical treatment or ICD therapy, or both [12-19].

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