Surgery for Hypertrophic Obstructive Cardiomyopathy Alive and Quite Well

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Dynamic obstruction to left ventricular (LV) outflow as a result of mitral valve systolic anterior motion is a potentially deleterious facet of hypertrophic cardiomyopathy (HCM).¹⁻⁴ In many patients, outflow obstruction is largely responsible for disabling symptoms of heart failure such as exertional dyspnea (often with chest pain), fatigue, and orthopnea.^{1,3} Consequently, treatment interventions that alleviate the subaortic gradient are critical therapeutic options for patients with HCM. Since the early 1960s, surgery (ie, ventricular septal myectomy) has been the primary treatment option for drug-refractory, severely symptomatic patients with the obstructive form of HCM.⁵⁻¹⁴

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Surgical Experience

Several thousand patients with HCM have undergone surgical septal myectomy worldwide during the past 45 years. Pioneered by Dr Andrew Morrow at the National Institutes of Health,⁵ septal myectomy and related operations have been performed in a number of largely North American and Western European centers.^{6–14}

In this issue of *Circulation*, Woo et al¹⁵ report one of the most important single-center surgical series encompassing 338 adult patients consecutively assembled over 25 years at Toronto General Hospital, with Dr William G. Williams as the senior operating surgeon.⁶ Septal myectomy is traditionally performed through an aortotomy, creating a rectangular trough (usually 3.5 to 5.0 cm in length) by 2 parallel longitudinal incisions in the basal septum (2 to 3.5 cm apart). These incisions are extended distally and connected just beyond the point of mitral–septal contact and obstruction (Morrow procedure)⁵ or at the bases of papillary muscles (extended myectomy),¹⁴ yielding a residual septal thickness of 8 to 10 mm and 3 to 15 g of septal muscle, and thereby enlarging the outflow tract and abolishing systolic contact between the mitral valve and the septum.^{3,4}

Long and extensive experience and the substantial data assembled from >25 centers worldwide have made septal

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Circulation is available at http://www.circulationaha.org DOI: 10.1161/01.CIR.0000164396.80300.1A myectomy an established and reliable strategy for patients of any age with HCM.3.4 Surgical intervention ameliorates obstruction (and mitral regurgitation) and reverses heart failure, thereby restoring functional capacity and an acceptable quality of life.3,4.6-14 Such salutary clinical benefits have been documented by patient history as well as objectively by increased treadmill time, maximum workload, peak oxygen consumption, and improved myocardial metabolism and coronary flow.3,4 Relief of obstruction with myectomy is immediate (and often necessary in severely symptomatic patients), permanent, and virtually complete. Indeed, Woo et al15 report that 98% of their patients had no significant outflow gradient at rest at the most recent echocardiographic examination (mean 5.5 years and up to 25 years after operation). Furthermore, only the surgical approach affords the flexibility under direct anatomic visualization that is often necessary to achieve complete repair and relief of subaortic obstruction, given the complex LV outflow tract morphology frequently encountered in HCM.14 In contrast, alternative catheter-based techniques such as alcohol septal ablation are anatomically restricted to the size and distribution of the septal perforator coronary artery.16-19

Furthermore, accumulating evidence from nonrandomized studies indicates that myectomy also provides a long-term survival benefit that is indistinguishable from that of the general population and superior to nonoperated patients with obstruction and therefore may alter the natural history of HCM.^{20,21} In this regard, the Toronto group¹⁵ also report high postoperative cardiovascular survival rates of 98%, 96%, and 87% at 1, 5, and 10 years, respectively.

Determinants of Long-Term Postoperative Course

Woo et al15 also expand their survival analysis and identify independent preoperative predictors of late postoperative mortality and cardiovascular morbidity. The disease variables that increased the likelihood of adverse HCM consequences late after myectomy were older age at surgery (\geq 50 years), female gender, concomitant coronary artery bypass grafting, preoperative atrial fibrillation, and transverse left atrial dimension ≥46 mm. Patients with atrial fibrillation before myectomy experienced an almost 50% reduction in this arrhythmia long term after surgery; however, another 21% of patients developed atrial fibrillation for the first time late after myectomy, a complication not uncommonly associated with progressive heart failure and major cardiovascular events. This often adverse impact of atrial fibrillation on clinical course in HCM also occurs independently of surgical intervention.22

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Substantial advances in surgical techniques for myectomy have taken place in the past several years, and these have dramatically reduced operative mortality and morbidity (ie, improved myocardial preservation strategies and postoperative care and generally greater experience), as well as the use of echocardiography in the operating room to monitor anatomic and functional results. Before 1990, operative mortality rates of 5% to 7% were reported from some major centers, disproportionately reflecting the early experience with myectomy from 1960 to 19853; however, these data can no longer be regarded as representative of the contemporary operation. During the past 10 to 15 years, surgical myectomy, when unassociated with coronary bypass grafting or valve replacement, has been performed with much lower mortality rates of 1% to 2% or less, in both children and adults,3,4 a result similar to the overall 1.5% reported by Woo et al.15 Most important, Toronto General Hospital has experienced just 1 operative death in the past decade and none in the most recent 145 consecutive cases. Indeed, several other major HCM surgical centers^{8,11} have also had recent operative mortality rates approaching zero during the past 10 years among almost 1000 cases. This point deserves particular emphasis because it establishes procedural-related risk of surgical myectomy at such centers to be less than alternative percutaneous treatments such as alcohol septal ablation3.16-19 performed in a multitude of practices for which mortality and morbidity data often go unreported. Consequently, it is important for cardiologists serving as gatekeepers for surgical referrals to be cognizant of the low mortality rates for myectomy (and to ignore older rates, which are irrelevant to current patients), as well as the favorable clinical results consistently attributable to surgery, when providing recommendations to patients with obstructive HCM.

Surgery as the Gold Standard

For all of the above reasons, septal myectomy is the most established remedy for obstructive HCM should heart failure symptoms become refractory to maximal medical treatment with negative inotropic agents (eg, β -blockers, verapamil, disopyramide), resulting in substantial lifestyle limitation equivalent to New York Heart Association (NYHA) functional classes III and IV, and in the presence of an LV outflow tract gradient \geq 50 mm Hg under resting (basal) conditions or when physiologically provoked with exercise.^{3,4} Children with obstructive HCM are often considered for surgery with somewhat lesser degrees of limitation. These guidelines governing the selection of patients with HCM for surgical myectomy represent those of the 2003 American College of Cardiology–European Society of Cardiology expert consensus panel on HCM treatment.³

Myectomy and Its Alternatives: Alcohol Septal Ablation

Treatment alternatives to surgery for selected patients who otherwise meet the accepted criteria for myectomy have historically been important to the management strategies available to patients with HCM.³ For example, particularly advanced age, associated medical conditions, or insufficient patient motivation can be significant obstacles to low-risk

surgery; also, some patients may lack reasonable access to a center that has experience with surgical myectomy. In this context, the percutaneous alcohol septal ablation technique that produces a permanent myocardial infarction^{3,4,16-19} has undergone a recent surge in popularity, emerging as an alternative to myectomy for some patients. Although reduction in LV outflow gradient and heart failure symptoms have been reported in many patients, a number of important questions remain about the ablation technique. With only short follow-up periods available in relatively small assembled cohorts (average <2 years), the question of whether ablation-related benefits will prove as sustainable as those documented with surgery during an 8-fold-greater time period has not been resolved. Furthermore, comparative nonrandomized studies show that myectomy and alcohol ablation are similarly associated with subjective improvement in NYHA functional class, but surgery yields more favorable outcomes with fewer early complications, more complete relief of obstruction, and greater exercise capacity and oxygen consumption.3 In addition, the risk for permanent complete heart block requiring pacing is up to 20 times greater with ablation (sometimes in young patients) than with myectomy.3,18 Because of the numerous practical and ethical obstacles surrounding this uniquely heterogeneous disease, it is unlikely that these difficult issues related to surgery versus ablation will ever be resolved by a randomized trial.

In addition, the potential for reentrant ventricular tachyarrhythmias and sudden death emanating from the alcohol-induced myocardial necrosis and scarring is of particular concern in patients with HCM, many of whom harbor preexistent, electrically unstable, and unpredictable arrhythmogenic myocardial substrates.3,4 Recent reports suggest that lethal arrhythmogenic events linked to ablation may not be uncommon.^{23,24} This risk cannot be ignored given the short observation period after ablation and the long duration of potential risk for many decades relevant to young patients with HCM. Therefore, specialized HCM centers have recommended septal myectomy, which does not create a residual intramyocardial scar,3,4 as the preferred treatment for refractory symptoms resulting from obstruction in children and adults through middle age.3.25 Alcohol ablation may be an appropriate option for some older patients with shorter potential risk periods, particularly when comorbidities or other contraindications to surgery are present. This is seemingly affirmed by the Toronto data,15 in which advanced age at operation was in fact a determinant of late mortality after myectomy. Furthermore, despite the long-acknowledged tenet that surgery is only necessary for a small number of carefully selected symptomatic patients with HCM (estimated to be $\leq 5\%$ of the overall HCM population)⁴ and that gradient and symptom criteria are essentially the same for surgery and ablation candidates, the threshold for alcohol ablation has obviously been lowered insidiously.3 Indeed, the number of catheter-based ablation procedures appears to have reached epidemic proportions in HCM—exceeding within only ≈5 years the total number of surgical myectomies performed over 45 years.3

An unappreciated factor promoting this circumstance is the peer-reviewed literature's understandable focus on novel

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observations, particularly the short-term results of innovative treatments. This inclination may have disproportionately skewed recent visibility toward the newly introduced percutaneous alcohol septal ablation, at the expense of the older and more accepted surgical myectomy. Unfortunately, this may have also created the illusion that surgery is no longer a viable treatment option to consider for severely symptomatic patients with obstructive HCM and conversely that alcohol ablation is always preferred. Indeed, evidence of this growing misconception in the cardiology community can be found in a recent highly visible editorial declaring myectomy obsolete by arbitrarily removing it from the HCM treatment algorithm.26 Because of the present unbridled enthusiasm for alcohol ablation, intensely promoted by the interventional community, it is possible that access to the important surgical option could eventually be lost to the HCM patient population unnecessarily.

Conclusions

The important article by Woo et al15 is notable for providing the septal myectomy operation with a welcome measure of visibility, which will go a long way toward placing the salutary benefits of surgery into proper perspective within the treatment armamentarium for HCM. This article is an important reminder that septal myectomy remains the time-honored primary strategy for drug-refractory, severely symptomatic patients with marked outflow obstruction. Because of the proven efficacy and the low procedure-related mortality and morbidity now associated with septal surgery, it is justifiable to promote an expanded access to surgery for patients with HCM. Even in this era of rapidly evolving cardiovascular therapeutics and percutaneous interventions, the older, more established, and familiar treatment strategy (ie, surgical septal myectomy) may nevertheless be preferable to that which is new and fresh, highly visible and accessible, and heavily promoted (ie, alcohol ablation).

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