Editorial

Hypertrophic Obstructive Cardiomyopathy: Alternative Therapeutic Options

NASSER LAKKIS, M.D., NEIL KLEIMAN, M.D., DONNA KILLIP, R.N., WILLIAM H. SPENCER, III, M.D.

Baylor College of Medicine, Houston, Texas

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Introduction

Hypertrophic obstructive cardiomyopathy (HOCM) is now recognized to be a relatively uncommon form of heart disease, but one that is frequently responsible for progressive symptoms of heart failure, angina, and sudden cardiac death. Symptoms are usually caused by obstruction to the left ventricular outflow tract, and relief of this obstruction is usually followed by clinical and hemodynamic improvement. Choice of treatment for HOCM is confounded by the variable and unpredictable natural history of the disease. The annual mortality rate is approximately 3 to 4% per year.^{1,2} Most of these deaths occur suddenly, especially in patients with a family history of HOCM, a history of syncope, and a diagnosis in childhood.³

Medical Treatment

During the early course of this progressive disease, treatment consists of beta blockers and calcium-channel antagonists, which ameliorate symptoms in many patients.⁴⁻⁶ Antiarrhythmic drugs have also been used in patients with HOCM, and recent clinical trials indicate that amiodarone may reduce the prevalence of sudden death in high-risk patients.⁷ There is a considerable variation in response to drug therapy, many effects have been reported, and the evidence that long-term drug therapy improves survival is tentative.⁸

Surgical Treatment

Surgical treatment in HOCM is usually reserved for patients who cannot tolerate their medical regimens, those who continue to have symptoms despite optimal drug therapy, and patients who have associated cardiac disease requiring surgery. The standard operation is ventricular septal myotomy-myectomy which was initially described by Cleland in 1963.⁹ Major complications of this operation are complete heart block, ventricular septal defect, and severe aortic insufficiency.^{10,11} The reported mortality due to this operation varies from 1% in young patients without underlying disease to 17% in patients older than 65 years, especially in the presence of coronary artery disease, or in those who required concomitant surgical procedures.^{12,13} The hemodynamic change resulting from this operation has been consistently associated with an improved quality of life as manifested by a reduction in congestive symptoms (exertional dyspnea, fatigue, orthopnea, and paroxysmal nocturnal dyspnea), chest pain, and symptoms of near syncope and syncope.^{11,14,15} However, almost 10% of patients had persistent or recurrent functional limitation. Mitral valve replacement is another surgical approach used for treatment of patients with refractory symptoms. It causes symptomatic relief comparable to that achieved by myotomy-myectomy surgery. However, the postoperative course is significantly affected by complications related to the prosthesis itself.^{16,17} There are no conclusive studies demonstrating whether or not any of these surgical approaches prolong life.^{12,14,15}

Dual Chamber Pacing

Dual chamber (DDD) pacing has been proposed as an alternative to surgical treatment in patients with HOCM, supposedly by inducing a more favorable left ventricular excitation pattern and through optimal timing of atrial contraction. Several investigators have independently shown a modest acute decrease in gradient during atrioventricular sequential pacing.¹⁸⁻²¹ McAreavey and Fananapazir²⁰ evaluated long-term outcome (34 ± 10 months) of DDD pacing in 114 adult patients with HOCM and symptoms refractory to beta blockers and verapamil therapy. New York Heart Association class improved significantly in 86% of the patients, and was

Address for reprints:

William H. Spencer III, M.D.
6550 Fannin, Suite 1025
Houston, TX 77030, USA

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associated with a significant decrease in left ventricular outflow tract gradient. The Pacing in Cardiomyopathy Study, a European multicenter study, has been verbally reported as improving the outflow gradient and quality of life but not exercise tolerance (personal communication).

Nonsurgical Ablation of Septal Bulge

Recently, Sigwart described a novel technique based on nonsurgical ablation of the septal bulge in patients with advanced HOCM,²² based on observations that intracavitary pressure gradients diminish greatly when the major first septal artery is temporarily occluded by an angioplasty balloon catheter. Sigwart modified this observation and he injected absolute ethanol through an inflated balloon catheter into the first and/or second septal perforators to produce a localized infarct. Twelve patients have been reported to have undergone the procedure, with a follow-up period of between 3 and 24 months. The hemodynamic improvement continues to be maintained with a mean resting gradient of 20 mmHg compared with one of 55 mmHg before the procedure.²³ A few months ago we started to investigate this procedure in symptomatic HOCM patients for the first time in the United States. Coronary angiography is performed at the time of the procedure in all patients to rule out significant coronary artery disease. Transthoracic echocardiography is performed both before and directly after the procedure to verify the resolution of the intraventricular gradient. We have performed this procedure on 10 markedly symptomatic patients who were resistant to drug therapy. All patients tolerated the procedure well; however, four patients developed complete heart block requiring permanent dual-chamber pacing. At 6 weeks' follow-up, all seven patients were virtually asymptomatic and had complete resolution of the left ventricular outflow tract gradient at rest with a minimal residual gradient during dobutamine stress. The attraction of the procedure resides in its simplicity, minimal morbidity, and in the fact that the outcome of the ablation can be estimated by echocardiography during temporary occlusion of the septal artery.

Note: Medical screening and information about financial arrangements for this procedure may be obtained by calling 1-888-HOCMTEX.

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