Management of Obstructive Cardiomyopathy

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CASE PRESENTATION

A 40-year-old woman presents for management of obstructive, hypertrophic cardiomyopathy. The patient has known of a heart murmur since childhood. There is no history of cardiovascular disease in her family. Six months prior to this presentation, an echocardiogram was interpreted elsewhere as showing concentric left ventricular hypertrophy. Doppler echocardiography disclosed evidence of mild mitral regurgitation, but no mention was made of subaortic obstruction. Cardiac catheterization, performed five months prior to this presentation, demonstrated generalized left ventricular hypertrophy, with additional septal hypertrophy. Midcavitary obliteration was seen with each systole, which was vigorous. Mild mitral regurgitation was recorded. A resting peak systolic gradient of 15 mm Hg was recorded across the left ventricular outflow tract, increasing to 35 mm Hg with the Valsalva maneuver; these gradients were recorded when the patient was in atrial fibrillation. After conversion to sinus rhythm, no gradient was recorded across the left ventricular outflow tract at rest or with the Valsalva maneuver.

Over the previous three months the patient developed exertional dyspnea and exertional light-headedness, with near syncope. She had not experienced chest discomfort or palpitations. Mild dependent edema had also developed. Her effort tolerance has steadily decreased as the result of the exertional dyspnea, fatigue, and exertional light-headedness. Symptoms have progressed despite treatment with a calcium-channel blocker (verapamil) and a beta-blocker (atenolol). On examination, the blood pressure measured 80/52 mm Hg. The upstroke velocity on the carotid pulse was brisk, but only a single peak was palpated. The left ventricle was not palpably enlarged, but a prominent thrill was felt along the middle and upper portions of the left sternal border, accompanied by a grade 4/6 systolic ejection murmur, which was loud, rough, and long-lasting throughout the ejection phase. The murmur did not change with the Valsalva maneuver. The second heart sound increased.

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Figure 1. Left ventricle at end-diastole. Generalized hypertrophy is noted with somewhat greater hypertrophy within the septal region. S = interventricular septum; PW = posterior wall of left ventricle; MV = mitral valve.

Figure 2. Left ventricle at end-systole, demonstrating marked systolic anterior motion of the mitral valve (MV) with virtual obliteration of the left ventricular outflow tract caused by apposition of the anterior leaflet of the mitral valve with the left ventricular septal region. S = interventricular septum; PW = posterior wall of left ventricle.

Figure 3. M = mode echocardiogram through the left ventricle at the level of the mitral valve demonstrates marked systolic anterior motion (SAM) of this valve. S = interventricular septum; PW = posterior wall of left ventricle.
displayed reversed splitting. An S_{2} gallop was heard at the apex with a diastolic long mitral flow rumble. Examination of the chest and abdomen disclosed normal findings.

The electrocardiogram showed tall R-wave voltage and T-wave inversion, consistent with left ventricular hypertrophy; left atrial abnormality was also noted.

Repeat echocardiography (Fig 1) showed concentric left ventricular hypertrophy, with the walls of the chamber measuring at least 2.3 cm thick throughout. In the midportion of the interventricular septum was an area that was slightly thicker than the remainder of the ventricle, measuring 2.5 cm. The left ventricular internal diameter measured 2 cm, with virtually total cavity obliteration with each systole (Fig 2). The anterior leaflet of the mitral valve was apposed to the septum throughout most of the ejection phase (Fig 3). The aortic valve demonstrated prominent notching, reopening in late systole. Doppler examination disclosed a peak instantaneous gradient of 118 mm Hg, with a mean gradient of 45 mm Hg. Moderate mitral regurgitation was recorded as well.

After the current evaluation, a trial withdrawal of atenolol resulted in improvement of exertional dyspnea and dizziness.

**QUESTIONS FOR CONSULTANT:**

1. How would you treat this patient?
2. What is currently the best form of medical therapy? What are the alternative agents for reducing the obstruction and/or improving diastolic function and relaxation? Do beta-blockers commonly increase the severity of symptoms?
3. How likely is it that this patient will respond to drugs?
4. If she fails to improve with medical treatment, is there a suitable surgical treatment? If so, please describe.

**Comments by Barry J. Maron, M.D.**

The case of this 40-year-old woman with hypertrophic cardiomyopathy raises a number of challenging questions regarding treatment strategy in this disease. Although she has a prior history of atrial fibrillation, at present she is not experiencing consistent congestive symptoms or functional limitation; for this reason, the patient would not conform to the generally accepted guidelines for operative intervention in obstructive hypertrophic cardiomyopathy.

Surgery has traditionally been confined to those patients with marked symptoms unresponsive to medical treatment (usually with beta-blockers or verapamil) who also have obstruction to left ventricular outflow due to systolic anterior motion of the mitral valve; to meet the latter criterion, an outflow gradient of ≥50 mm Hg at cardiac catheterization (under basal conditions or with provocative interventions) should be present. Consequently, operation is performed in obstructive hypertrophic cardiomyopathy for the purpose of relieving the subaortic gradient, with the expectation that this will also improve symptoms and the quality of life. Surgical treatment is justified in this regard, predicated on a 30-year experience demonstrating that most patients (about 70 percent) who have operative relief or abolition of their outflow gradient experience long-term symptomatic (functional) benefit. The surgical procedure of choice is ventricular septal myomectomy (Morrow procedure), although mitral valve replacement may be a reasonable alternative for selected patients in whom the basal ventricular septum is relatively thin or congenital malformations of the mitral valve apparatus are present.

Surgical treatment for obstructive hypertrophic cardiomyopathy has, by convention, been withheld from patients with no or only mild symptoms (even if marked obstruction to outflow and elevated left ventricular systolic pressures are present). This is largely because the operative risk of about 5 percent has been judged to be unacceptably high in a clinical setting in which it is not known whether longevity is truly increased by surgical relief of obstruction. Finally, atrial fibrillation (paroxysmal or chronic) has not generally been viewed as an independent indication for surgery in hypertrophic cardiomyopathy since evidence is lacking that relief of subaortic obstruction also abolishes or reduces a patient's predisposition to this arrhythmia.

Of note, the present patient experienced apparent cardiac symptoms (ie, exertional dyspnea and impaired consciousness) over an isolated three-month period. In retrospect, the cessation of these symptoms following withdrawal of her beta-blocker (ie, atenolol) strongly suggested that those symptoms were secondary to medications rather than to her intrinsic cardiac disease process. Such side effects of beta-blockers are well recognized (although not usually to such a degree) and emphasize an important clinical issue relevant to recommendations for surgery in patients with obstructive hypertrophic cardiomyopathy. This particular patient's physicians were wise enough to recognize the possibility that her symptoms were not indicative of true cardiac deterioration, and consequently operation was deferred at that time.

A second important point with regard to this patient concerns the magnitude of left ventricular outflow tract obstruction that was present. A previous cardiac catheterization had reportedly shown absence of a subaortic gradient. However, evidence of marked dynamic obstruction to left ventricular outflow typical of hypertrophic cardiomyopathy was later demonstrated, as evidenced by a loud systolic ejection murmur (at and near the apical impulse), marked systolic anterior motion of the mitral valve, and an outflow gradient of more than 100 mm Hg estimated by continuous-wave Doppler. There are two possible explanations for a discrepancy of this nature. First, it is possible that this patient experienced a true increase in gradient from zero to more than 100 mm Hg over a six-month period. In my experience, however, such a

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marked spontaneous change in magnitude of obstruction is rare in adult patients with hypertrophic cardiomyopathy. Alternatively, it is possible that the outflow gradient had not been identified at cardiac catheterization because of technical reasons or because it had been suppressed by pharmacologic therapy at that time.

REFERENCES
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