

Hypertrophic Cardiomyopathy Characterized by Marked Hypertrophy of the Posterior Left Ventricular Free Wall: Significance and Clinical Implications

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This report describes a subgroup of 17 patients with hypertrophic cardiomyopathy and an unusual and distinctive pattern of left ventricular hypertrophy characterized on echocardiography by marked thickening of the posterior left ventricular free wall and virtually normal or only modestly increased ventricular septal thickness. This distribution of hypertrophy often created a distinctive pattern of "inverted" asymmetry of the posterior wall relative to the septum. The thickness of the posterior wall was 20 to 42 mm (mean 25), while that of the basal ventricular septum was only 12 to 24 mm (mean 17). The left ventricular outflow tract was narrowed because of anterior displacement of the mitral valve within the small left ventricular cavity. Systolic anterior motion of the mitral valve was present in 16 of the 17 patients.

The patients ranged in age from 13 to 54 years (mean 31) at most recent evaluation; most (11 of 17, 65%) were severely symptomatic and had experienced important symptoms early in life (before age 40). The condition of only 4 of these 11 patients

improved with medical therapy over an average follow-up period of 9 years; however, 6 of the 7 patients who had unsuccessful medical treatment and underwent operation with mitral valve replacement (5 patients) or ventricular septal myotomy-myectomy (1 patient) experienced symptomatic benefit from surgery.

The subgroup of patients described in this report underscores the morphologic and clinical diversity that exists within the overall disease spectrum of hypertrophic cardiomyopathy. Characteristically, the patients were young, severely symptomatic and demonstrated evidence of outflow obstruction and an "inverted" asymmetric pattern of posterior free wall left ventricular hypertrophy. Because of their relatively modest ventricular septal hypertrophy, mitral valve replacement (rather than myotomy-myectomy) may be the operative procedure of choice in such patients with obstructive hypertrophic cardiomyopathy.

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Hypertrophic cardiomyopathy is a primary myocardial disease with a diverse clinical and morphologic spectrum (1-15). Although a variety of patterns of left ventricular hypertrophy have been described, the vast majority of patients show asymmetric and predominant thickening of the ventricular septum (7-18), whereas the posterior free wall is usually the least thickened portion of the ventricle (7,16). This morphologic feature of hypertrophic cardiomyopathy is the basis of the characteristic finding of asymmetric hypertrophy of the ventricular septum on the M-mode echocardiogram (7,8,16). In contrast, we have recently observed a number of other patients with atypical morphologic forms of hypertrophic cardiomyopathy who have a striking and often asymmetric thickening of the posterior free wall relative to

the ventricular septum. Consequently, we undertook the present analysis to characterize in detail the clinical and morphologic profile and significance of this unique morphologic subgroup of patients with hypertrophic cardiomyopathy.

Methods

Selection of patients. The case records of the Echocardiography Laboratory of the National Heart, Lung, and Blood Institute were reviewed for the period from January 1984 to June 1989. During that time, 17 patients with hypertrophic cardiomyopathy met the following criteria for inclusion in the present study: 1) asymmetrically hypertrophied and nondilated left ventricle in the absence of associated cardiovascular or systemic disease capable of producing left ventricular hypertrophy (17); and 2) increased posterior free wall thickness (≥ 20 mm), similar to or greater than that of the basal ventricular septum. All patients gave informed consent for participation in the study. At the most recent evaluation, the study patients ranged in age from 13 to 54 years (mean 31); only three were >40 years of age. Nine patients were female and eight were male. Eight of the 17 study patients had a family history of hypertrophic cardio-

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myopathy or premature sudden death; however, systematic echocardiographic studies were not performed in the pedigrees of the other 9 patients.

Echocardiography. Two-dimensional echocardiograms were performed with use of an Advanced Technology Laboratory mechanical sector scanner or a Hewlett-Packard phased arrayed imaging system with a 2.25 or 2.5 MHz transducer. Two-dimensional images were obtained in multiple cross-sectional planes with use of standard transducer positions (19). Magnitude and distribution of left ventricular hypertrophy were assessed from the two-dimensional cross-sectional images as previously described (7). Wall thickness was measured directly from the television monitor utilizing the calibration scale produced by the instrument. Basal ventricular septal and posterior free wall thicknesses were derived from an integrated analysis of the M-mode and two-dimensional echocardiograms.

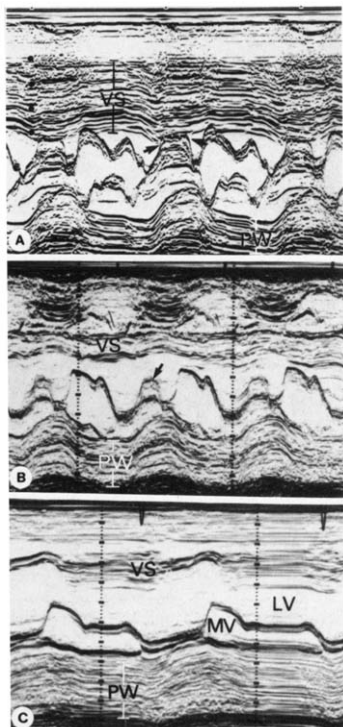
M-mode echocardiograms were derived under direct anatomical visualization from the two-dimensional images. Measurements of left ventricular wall thickness and chamber dimensions were made according to the recommendations of the American Society of Echocardiography (20). The position of the mitral valve in the left ventricular cavity was assessed by using the mitral valve position index, which was calculated by dividing the distance between the mitral valve (at the time of leaflet closure) and the posterior left ventricular free wall endocardium by the distance between the mitral valve and ventricular septal endocardium (21). Systolic anterior motion of the mitral valve was defined as mild, moderate or severe according to the classification of Gilbert et al. (22).

Results

Left ventricular morphology (Fig. 1 to 4). *Posterior wall thickness.* By selection, in each of the 17 study patients, left ventricular posterior free wall thickness was markedly increased, ranging from 20 to 42 mm (mean 25), and was particularly striking (>30 mm) in 3 of these patients. In 7 of the 17 patients, the posterior wall was diffusely thickened in both the proximal (basal) and distal portions (Fig. 2 and 3A); in the other 10 patients, the posterior wall was thickened only in the basal segment, that is, the region between the mitral annulus and the papillary muscle (Fig. 3, B to D). When particularly localized, this area of posterior wall hypertrophy appeared as a prominent "bump" or bulge as a result of the sharp and abrupt change in thickness between contiguous portions of the wall (Fig. 3, C and D).

Septal thickness. Thickness of the basal ventricular septum ranged from 12 to 24 mm (mean 17) and was nearly normal (12 and 13 mm) in 5 patients and only mildly increased (14 to 15 mm) in 4. Consequently, in each of the 17 patients, the posterobasal free wall was at least as thick as the basal septum; the conventional septal/free wall thickness

Figure 1. M-mode echocardiograms at the mitral valve level from three patients with hypertrophic cardiomyopathy. A, From an 11-year old patient (not in the present study) with obstruction to left ventricular outflow and typical asymmetric hypertrophy of the ventricular septum. Ventricular septal (VS) thickness is 25 mm and posterior free wall (PW) thickness is 13 mm; the septal/free wall thickness ratio is almost 2.0. Marked systolic anterior motion of the mitral valve and prolonged mitral-septal contact are also present (arrows). B and C, From two study patients (aged 36 and 13 years) with predominant and asymmetric hypertrophy of the posterior left ventricular free wall. B, Patient 14, Table 1. The ventricular septum at this level is of near normal thickness (12 mm) whereas the posterior free wall is hypertrophied (22 mm), creating an "inverted" pattern of left ventricular asymmetry (compare with panel A); mitral systolic anterior motion without mitral-septal contact (arrow) is present. C, Patient 1, Table 1. The posterior wall is strikingly thickened (>35 mm) whereas the septum is particularly thin at this level; mitral systolic anterior motion and left ventricular outflow obstruction are absent. Calibration dots are 10 mm apart. LV = left ventricle; MV = mitral valve.



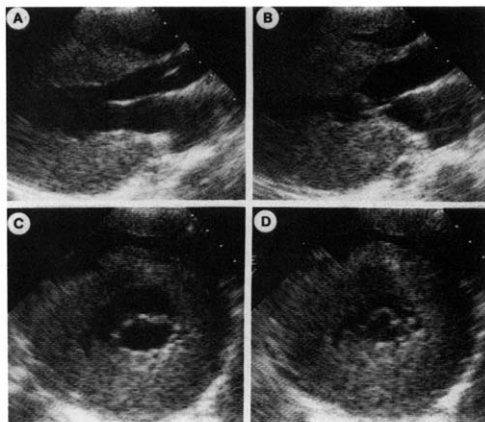
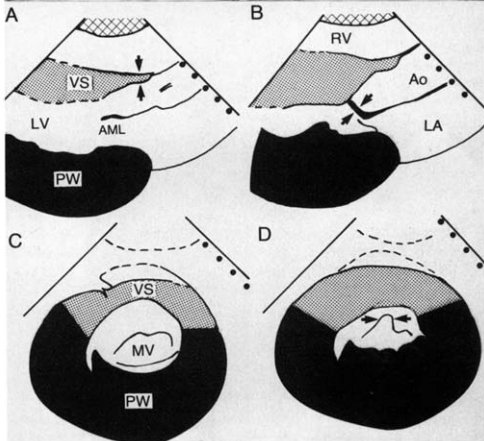


Figure 2. Stop-frame two-dimensional echocardiographic images from a 14-year old girl with hypertrophic cardiomyopathy (Patient 2, Table 1). Schematic drawings are shown below. A and C, Diastole. B and D, Systole. A, As viewed in the parasternal long-axis plane, the thickness of the posterior free wall (PW) is strikingly increased in both proximal and distal regions and substantially exceeds the thickness of the basal ventricular septum (VS). The most proximal portion of the septum just below the aortic valve is particularly thin (arrows), while the distal septum is markedly thickened. B, As viewed in the long-axis plane, the anterior mitral leaflet (AML) bends sharply during systole and the distal tip makes contact with the ventricular septum near the point of maximal thickness (arrows), producing a subaortic gradient of 40 mm Hg. C, In the short-axis plane, the ventricular septum appears to be of normal thickness whereas the posterior free wall and contiguous portions of the posterior septum and lateral free wall are substantially thickened, resulting in the appearance of inverted asymmetry of the posterior wall relative to the anterior septum. D, A short-axis plane that is slightly more distal than the plane shown in C. During mid-systole, the central third of the mitral valve has moved anteriorly toward the septum (arrows), further narrowing the left ventricular outflow tract. Calibration dots are 10 mm apart. Ao = aorta; LA = left atrium; LV = left ventricle; MV = mitral valve; RV = right ventricle.



ratio on M-mode echocardiogram was ≤ 1.0 in 16 patients and 1.1 in 1. Indeed, the left ventricle in 13 of the 17 patients had the distinctive appearance of "inverted" asymmetric hypertrophy of the posterior wall with respect to the septum; that is, the thickness ratio of the basal ventricular septum to the posterobasal free wall was ≤ 0.8 for this subgroup (Fig. 1 to 4).

Patterns of hypertrophy. Assessment of overall distribution of left ventricular hypertrophy in the 17 study patients revealed a variety of patterns of wall thickening. In six patients, marked hypertrophy was largely confined to the posterior free wall, while other left ventricular segments were virtually spared from the hypertrophic process with wall thickening of 12 to 15 mm (Fig. 3, B to D). Five patients

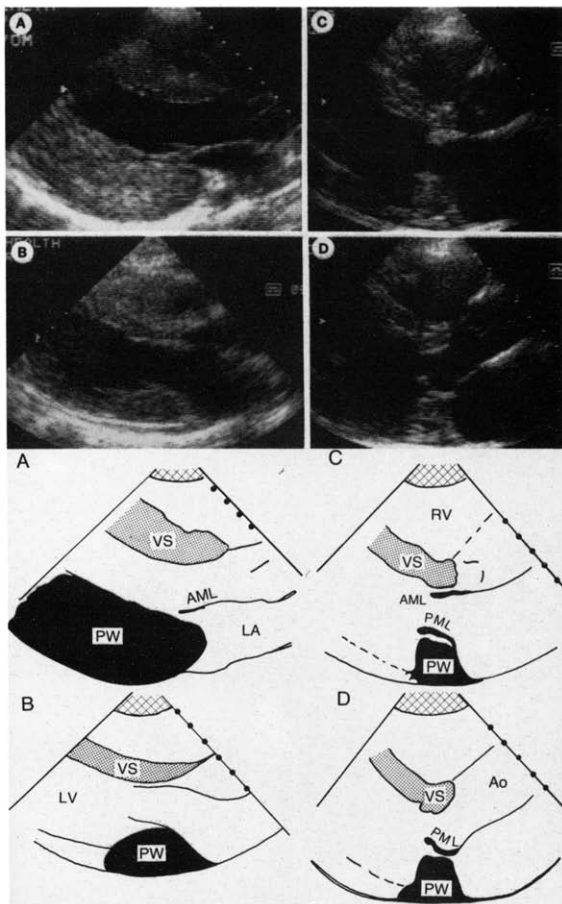


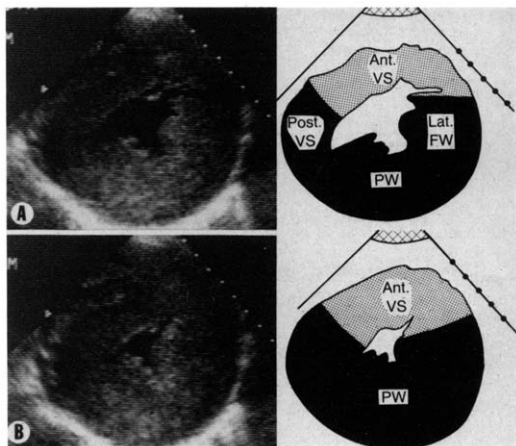
Figure 3. Echocardiographic images in the parasternal long-axis plane from three patients demonstrating variability in the extent of posterior free wall hypertrophy. Schematic drawings are shown below. A to C, Diastole. D, Systole. In each patient, the thickness of the posterior free wall (PW) exceeds that of the ventricular septum (VS). A, Patient 1, Table 1. The posterior wall (PW) shows a particularly striking increase in the thickness of both proximal and distal portions of the left ventricle (of up to 42 mm). B, Patient 9, Table 1. Increased left ventricular thickness confined to the most proximal portion of posterior wall behind the mitral valve (20 mm) creates the appearance of a bulge or "bump." C, Patient 17, Table 1. There is relatively localized thickening of the posterobasal left ventricular free wall; an abrupt decrease in wall thickness is evident distal to the thickened segment. D, During systole (from the same patient shown in C), the anterior mitral leaflet (AML) bends anteriorly and the tip approaches the ventricular septum. Calibration dots are 10 mm apart. Ao = aorta; LA = left atrium; LV = left ventricle; PML = posterior mitral leaflet; RV = right ventricle.

showed more extensive hypertrophy involving the anterolateral free wall in addition to the posterior wall, but also frequently extending into portions of the anterior or posterior ventricular septum. The remaining six patients showed an asymmetric pattern of diffuse hypertrophy with marked

thickening (>20 mm) involving each left ventricular segment, including the posterior wall (Fig. 2, 3A and 4).

Outflow tract. Despite the relatively modest septal thickness present in most patients, the left ventricular outflow tract was markedly narrowed, as evidenced by substantially

Figure 4. Patient 1, Table 1. Two-dimensional echocardiogram in the short-axis plane at the papillary muscle level from the same patient shown in Figure 3A. At end-diastole, the posterior wall (PW) shows the most marked thickening (about 35 mm) and the anterior portion of the ventricular septum (Ant. VS) the least thickening (22 mm); the left ventricular cavity is small. B. At end-systole, the left ventricular cavity is virtually obliterated. Calibration dots are 10 mm apart. Lat. FW = lateral free wall; Post. VS = posterior ventricular septum.



reduced mitral valve to ventricular septal transverse distance at end-diastole (range 15 to 31 mm [mean 22]) and anterior displacement of the mitral valve within the left ventricular cavity (mitral valve position index 0.2 to 1.1 [mean 0.7]; normal <0.4) (21). These values for mitral valve position are comparable to those previously reported (23) in patients having more typical morphologic forms of obstructive hypertrophic cardiomyopathy with marked asymmetric ventricular septal hypertrophy.

Systolic anterior motion of the mitral valve. This was present in 16 of the 17 study patients (Fig. 1B, 2B, 2D and 3D) and was judged to be marked in 9 (with prolonged mitral-septal apposition) and moderate in 7 (with brief or near contact) (23). In 15 patients, both the anterior and the posterior leaflets appeared to participate in mitral systolic anterior motion, although the anterior leaflet made contact with (or most closely approached) the septum during systole because of its anterior anatomic position; in the remaining patient, the posterior mitral leaflet was preferentially responsible for systolic anterior motion (24).

By selection, the left ventricular chamber was nondilated in each patient (end-diastolic dimension 28 to 52 mm [mean 38]). Left atrial size ranged from 36 to 75 mm (mean 50) and was dilated (>40 mm) in 15 of the 17 patients.

Hemodynamic findings. Ten of the 17 study patients underwent complete cardiac catheterization. Nine of these 10 patients had a left ventricular outflow tract gradient ≥ 30 mm Hg under basal conditions (range 50 to 160 mm Hg [average 94]); the remaining patient had no basal outflow

gradient, but developed a gradient of 50 mm Hg after provocation with amyl nitrite.

Of the seven patients without cardiac catheterization, three had a basal outflow gradient of 35 to 75 mm Hg estimated with continuous wave Doppler or M-mode echocardiography (25,26). Three of the four remaining patients without cardiac catheterization showed moderate mitral valve systolic anterior motion with brief mitral-septal contact (or near contact) under basal conditions, consistent with the propensity to develop a subaortic gradient, primarily with provocation (21,22). Hence, on the basis of the integrated hemodynamic and echocardiographic data, 16 of our 17 patients showed evidence of basal or provokable obstruction to left ventricular outflow. Eleven of the 17 study patients had mild to moderate mitral regurgitation demonstrated by left ventriculography or Doppler echocardiography; no patient had a transmural diastolic gradient.

Electrocardiographic findings. The 12-lead electrocardiogram (ECG) obtained at the most recent clinical evaluation was abnormal in each patient, although no pattern was characteristic of the overall study group. Electrocardiographic abnormalities included left ventricular hypertrophy (Romhilt-Estes score ≥ 5 points [nine patients]) (27), left atrial enlargement (seven patients) and conduction abnormalities such as nonspecific intraventricular conduction delay (two patients), first-degree atrioventricular block (two patients), right bundle branch block (one patient) and left bundle branch block (one patient). In addition, two patients showed particularly marked T wave inversion in the lateral precor-

Table 1. Clinical, Echocardiographic, Hemodynamic and Electrocardiographic Findings in 17 Patients With Hypertrophic Cardiomyopathy and Marked Hypertrophy of the Posterior Free Wall

Patient No.	Age (yr)/Gender	NYHA Class	Symptoms	Basal Septum (mm)	Posterior Wall (mm)	Distribution of LVH	LVOT Obstruction (mm Hg)		ECG			Operation
							Basal	Prov	LVH	ST-T	Other	
1	13/M	I	None	24	42	Intermediate*	0 [†]	-	+	LBBB	0	
2	14/F	I	None	24	40	Diffuse†	40 [‡]	-	+	LAE	0	
3	15/F	III	CP,Doc	13	20	Intermediate	85	100	+	LAE	MVR	
4	16/M	II	Doc,CP	20	20	Diffuse	0 [‡]	-	+	Q waves, LAE	0	
5	21/M	III	Syn	24	39	Diffuse	0	50	0	RBBB-LAFB	0	
6	23/M	III	CP,Doc,Syn	24	24	Diffuse	130	-	+	0	0	
7	25/M	III	Doc,Syn	14	29	Posterior wall‡	0 [‡]	-	0	0	0	
8	26/F	I	None	13	23	Posterior wall	75‡	-	0	+ IVCD	0	
9	28/F	III	CP,Doc,Syn,Palp	12	20	Posterior wall	80	88	0	0	Q waves, LAE	MVR
10	31/M	III	Doc	14	23	Intermediate	90	120	+	0	1° AVB, LAE	MVR
11	31/M	IV	CP,Doc,Syn,Palp	15	21	Posterior wall	50	85	+	+	LAE	MVR
12	34/F	III	Syn,Palp	13	22	Intermediate	169	-	+	+	0	MVR
13	36/F	III	CP,Doc,Palp	22	20	Diffuse	70	100	+	+	0	0
14	36/M	I	None	14	22	Posterior wall	0 [‡]	-	+	+	0	0
15	43/F	III	CP,Palp	22	22	Diffuse	110	-	+	0	LAE	M-M
16	46/F	IV	Doc,Syn	18	25	Intermediate	70	-	0	0	AFib, IVCD	0
17	54/F	I	None	13	20	Posterior wall	36‡	-	0	0	1° AVB	0

*Hypertrophy involving the posterior and anterolateral free walls, but frequently extending into portions of the anterior or posterior septum. †Asymmetric pattern of hypertrophy with marked thickening involving each left ventricular segment, including the posterior wall. ‡Patients who did not undergo cardiac catheterization; left ventricular outflow gradient estimated with continuous wave Doppler study (Patients 2 and 17) or M-mode echocardiogram (Patients 1, 4, 7, 8 and 14). §Marked hypertrophy confined to the posterior left ventricular free wall; Romblit-Estes score = 5. AFib = atrial fibrillation; AVB = atrioventricular block; CP = chest pain; Doc = exertional dyspnea; ECG = electrocardiogram; 1° = first degree; IVCD = nonspecific intraventricular conduction delay; LAE = left atrial enlargement; LBBB = left bundle branch block; LVH = left ventricular hypertrophy; LVOT = left ventricular outflow tract; M-M = ventricular septal myotomy-myectomy; MVR = mitral valve replacement; NYHA = New York Heart Association functional class; Palp = palpitations; Prov = provocation; RBBB = right bundle branch block; ST-T = ST-T wave abnormalities; Syn = syncope. 0 = absent; + = present; - = data not available or applicable.

dial leads (28). Most patients (15 of 17) had normal sinus rhythm; the other 2 had atrial fibrillation.

Clinical findings. Eleven of the 17 patients experienced severe, incapacitating cardiac symptoms (New York Heart Association functional class III or IV); the other 6 had only mild symptoms. Onset of symptoms occurred before age 40 in each patient (range 9 to 39 years [mean 25]). The most common symptoms were exertional dyspnea (nine patients), syncope or near-syncope (seven patients), chest pain (seven patients) and palpitation (five patients).

Medical treatment. Only 4 of the 11 severely symptomatic patients showed sustained improvement on medical treatment with beta-adrenergic or calcium channel blocking agents over a follow-up period of 1 to 28 years (mean 9). The other seven symptomatic patients proved refractory to these pharmacologic agents; six underwent operation (mitral valve replacement in five and ventricular septal myotomy-myectomy in one). Surgery was deferred in the remaining study patient because of marked obesity.

Operation. Mitral valve replacement was performed in five patients in whom the risk of producing a ventricular septal defect with myotomy-myectomy was judged to be unacceptably high because the basal septal thickness was only 12 to 15 mm (29); in the patient who underwent myotomy-myectomy, septal thickness was 22 mm. Cardiac

catheterization was performed postoperatively in five of the six surgically treated patients; complete relief of left ventricular outflow obstruction (i.e., zero gradient under basal conditions) was demonstrated in four patients and the remaining patient had a postoperative basal outflow gradient of 80 mm Hg. Each of the six surgically treated patients showed initial postoperative improvement by at least one functional class that has persisted in five patients over a follow-up period of 1 to 5 years (mean 1.3); the remaining patient developed symptomatic deterioration 3 years after operation. There were no deaths in the study group during the follow-up period.

Discussion

Although the distribution of ventricular hypertrophy is characteristically asymmetric in patients with hypertrophic cardiomyopathy, there is marked diversity with regard to both the precise magnitude and the pattern of left ventricular wall thickening (5,7-11,14,15). Hypertrophy is commonly diffuse, involving substantial portions of both the ventricular septum and the anterolateral free wall; usually the ventricular septum is the thickest portion of the left ventricle, and the posterior free wall is largely spared the hypertrophic process. Indeed, comparison of the markedly increased thick-

ness of the septum with the normal or only mildly increased posterior wall thickness is the basis of the characteristic asymmetric ventricular septal hypertrophy and abnormal septal/free wall thickness ratio identified with M-mode echocardiography in the vast majority of patients with hypertrophic cardiomyopathy (8,16,18).

Left ventricular morphology. The 17 patients described in this study constitute an uncommon but distinctive variant within the diverse morphologic spectrum of hypertrophic cardiomyopathy (7). Unlike the vast majority of patients with this disease (7,10,11,14), our patients demonstrated substantial and often predominant hypertrophy of the posterior left ventricular free wall. Indeed, most showed an inverted pattern of asymmetric left ventricular hypertrophy in which the thickness ratio of basal ventricular septum to posterobasal free wall was substantially <1.0 . This observation and the pattern of left ventricular hypertrophy shown by these patients underscore the recognized limitation (16) of using asymmetric hypertrophy of the ventricular septum relative to the posterior free wall as the sole morphologic marker of hypertrophic cardiomyopathy.

Whereas ventricular septal thickening was relatively mild (≤ 15 mm) in most of our patients, the marked thickening of the posterobasal left ventricular free wall appeared to contribute to the anterior position of the mitral valve within the cavity and consequently to narrowing of the outflow tract. Therefore, even though the ventricular septum is not as thick as the posterior wall in these patients, the mitral valve is nevertheless positioned close to the septum. Presumably, this distortion of left ventricular geometry produces a circumstance similar to that in patients with more typical asymmetric hypertrophy of the ventricular septum, in which the high velocity of blood flow through the narrowed outflow tract pulls (30) (or possibly drags [31]) the mitral leaflets anteriorly toward the septum during systole. This mitral systolic anterior motion and contact between the mitral leaflets and ventricular septum was apparently responsible for the dynamic left ventricular outflow gradients recorded under basal conditions or with provocative maneuvers in the catheterization laboratory in the vast majority of patients (30,32,33).

Clinical course. The distinctive morphology of the subgroup of patients described in this report was associated with a particular clinical profile. Patients were relatively young (mean age about 30), the majority had severe symptoms that usually proved refractory to medical therapy and most of these patients showed symptomatic benefit from operation. Development of important limiting symptoms at a relatively young age suggests that this morphologic variant may represent a clinically severe form of hypertrophic cardiomyopathy. However, because the clinical course in hypertrophic cardiomyopathy is typically influenced by multiple factors (14), we do not wish to suggest a causal relation between the severity of symptoms and the unusual distribution of left ventricular hypertrophy observed in the relatively small group of patients studied. Moreover, we cannot exclude the

potential influence of selection bias in the referral patterns to our institution on the clinical course of the study patients.

Implications for surgery. The unusual left ventricular morphology present in our study patients had important clinical implications for operative strategy. Ventricular septal myotomy-myectomy is the preferred operative procedure in most patients with obstructive hypertrophic cardiomyopathy having severe, intractable symptoms unresponsive to medical treatment (29,34-37); however, in certain patients in whom basal ventricular septal thickness is <18 mm, the risk of either incurring an iatrogenic ventricular septal defect as a result of resection of septal muscle or failing to achieve an optimal hemodynamic result may be unacceptably high (29,30,37). In such patients, mitral valve replacement constitutes an alternative operation for relief of left ventricular outflow obstruction and cardiac symptoms (29). Indeed, these considerations were pertinent for several of the present study patients. Despite a striking increase in overall left ventricular mass, 5 of our 6 surgically treated patients underwent mitral valve replacement (rather than myotomy-myectomy) because only relatively mild thickening (≤ 15 mm) of the basal ventricular septum was identified.

Relation of study patients to the overall spectrum of hypertrophic cardiomyopathy. The unusual left ventricular morphology present in our study patients, which differs from that of other patients with hypertrophic cardiomyopathy, raises the consideration of whether these patients truly constitute part of the broad spectrum of this disease. In support of the diagnosis of hypertrophic cardiomyopathy is the clinical evidence of genetic transmission of this disease in first-degree relatives of about half of our patients (38). Furthermore, 16 of the 17 study patients showed evidence of dynamic obstruction to left ventricular outflow characteristic of hypertrophic cardiomyopathy with a typical pattern of systolic anterior motion of the mitral valve and a dynamic subaortic gradient documented in the cardiac catheterization laboratory under basal conditions or with provocative maneuvers (11,18,22,30). Finally, none of our patients had an associated cardiac disease (such as systemic hypertension or aortic valve stenosis) that itself could have produced or contributed to the marked asymmetric left ventricular hypertrophy.

Conclusions. We have described a subgroup of patients with hypertrophic cardiomyopathy characterized by an unusual morphologic pattern in which there is marked and often asymmetric thickening of the posterior left ventricular free wall. The clinical profile of these patients included outflow obstruction and severe symptoms early in life that usually proved refractory to medical therapy; however, each patient undergoing operation experienced symptomatic benefit. The pattern of left ventricular hypertrophy present in many patients had important potential implications for operative strategy. Despite a marked increase in overall left ventricular mass, most surgically treated patients underwent mitral valve replacement rather than ventricular septal myotomy-myectomy because the relatively modest degree

of basal ventricular septal thickening constituted an unacceptable risk for iatrogenic ventricular septal defect.

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