Hypertrophic cardiomyopathy is a disease that was first described clinically by Brock in 1957 as functional obstruction of the left ventricle. Most information about the pathophysiology and natural history of hypertrophic cardiomyopathy has been obtained from young and early middle-aged patients. Although hypertrophic cardiomyopathy is known to occur in the elderly, most natural history studies include very few patients over the age of 60. A few recent studies have focused on the elderly with hypertrophic cardiomyopathy and have suggested that the condition takes a different form in this age group. Wadieha et al studied a group of 58 patients (mean age, 51 years) and described a relatively benign course in terms of morbidity and mortality at 6 years follow-up compared with previous reports in younger patients. Cohen et al have shown that the natural history of this disease is considerably more benign in older than in young patients and may in fact represent an entirely different form of disease. McKenna et al showed that the combination of young age, syncope at diagnosis, severe dyspnea at last follow-up, and a family history of hypertrophic cardiomyopathy with sudden death was associated with a worse prognosis, including an annual mortality of 5.9% in those less than 14 years of age, compared with a mortality of 2.6% in the group between 45 and 60 years. Studies have also indicated that this condition may be more common in the elderly than generally recognized.

Whether or not hypertrophic disease of the elderly is the same as that in the young and early middle-aged adult is not clear. If it is the same, several questions arise: how can it have such a benign and silent course for years, and does it have the same etiology, pathophysiology, and genetic basis as that in the young? A first step in addressing these ques-

Hypertrophic Cardiomyopathy in the Elderly
Distinctions From the Young Based on Cardiac Shape

Harry M. Lever, MD, Roger F. Karam, MD, Philip J. Currie, MBBS, and Bernadine P. Healy, MD

The striking clinical and pathologic features of hypertrophic cardiomyopathy have been defined almost exclusively in the young. Little is known about this condition in the elderly, although it is assumed to be part of a single disease. Accordingly, we studied 28 patients who were 65 years of age and older (mean age, 72±6.4 years) who were diagnosed as having hypertrophic cardiomyopathy by M-mode, two-dimensional, and Doppler echocardiography and compared them with a group of 28 consecutive patients with this disease who were younger than 40 years of age (mean age, 26±9.5 years). No clinically detectable differences existed between the two groups, except for an increased incidence of mild hypertension in the elderly. Echocardiography in both groups showed hypertrophy with a small left ventricular cavity, and Doppler outflow tract velocity or cardiac catheterization in most patients showed systolic anterior motion of the mitral valve or a systolic outflow tract gradient or both. Significant differences existed between both groups, however, concerning left ventricular size and shape. The elderly group had a predominantly ovoid cavity contour with normal septal curvature. In contrast, in the young group, a markedly abnormal cardiac shape predominated (p<0.0001) with a crescent-shaped left ventricular cavity and a reversed curvature of the interventricular septum. The right ventricular free wall was prominent by echocardiography in the young compared with the elderly group (p<0.001). Apart from a more frequent history of mild hypertension in the elderly, likely related to age, these findings show that hypertrophic cardiomyopathy, when present in the elderly, has similar clinical features to that in the young. However, the elderly had a strikingly different and generally undetected cardiac shape by echocardiography, supporting the notion that hypertrophic cardiomyopathy in the elderly may be a disease distinctively different from that which predominates in the young. (Circulation 1989;79:580–589)
tions is to determine whether or not the structural and functional characteristics of the heart in hypertrophic cardiomyopathy in the elderly are similar to those in the young. We therefore studied a consecutive group of elderly patients with hypertrophic cardiomyopathy and compared their clinical and echocardiographic features with those of a consecutive group of patients with the same diagnosis but at the other end of the age spectrum, selected only on the basis of younger age (<40 years). Although hypertrophic cardiomyopathy was similar clinically in the elderly and the young, a significantly different pattern of cardiac shape was identified between these two groups. These different shapes may have implications for investigating the etiology and natural history of this disease and may provide a framework for studying its heterogeneity.

Methods

Patients

We reviewed the records of the echocardiographic laboratory at The Cleveland Clinic Foundation between January 1986 and January 1987 for the diagnosis of hypertrophic cardiomyopathy in the elderly aged 65 years and older, without any primary valvular heart disease. We then identified an equal number of consecutive patients less than 40 years of age with hypertrophic cardiomyopathy. It was necessary to review our records back to 1982 to find an equal number of younger patients that could be compared with the older age group. For the two selected groups, the only criterion for subsequent exclusion was a poor quality echocardiogram.

The diagnosis of hypertrophic cardiomyopathy was based upon a combination of the following accepted criteria, drawn from two-dimensional, M-mode, and Doppler echocardiographic and cardiac catheterization data: left ventricular hypertrophy (septum wall ≥15 mm for adults) with a normal to small-sized cavity (in the absence of valvular stenosis); systolic anterior motion of mitral valve at rest or with provocation; presence of a gradient in the left ventricular outflow tract by Doppler or cardiac catheterization, at rest or with provocation; and asymmetry of the interventricular septum. The clinical charts and echocardiographic studies were reviewed separately and at different times for all patients.

Clinical Findings

Data were collected by one reviewer from clinical records. Functional class was based upon the New York Heart Association (NYHA) classification system (I-IV). Hypertension in the young was defined as blood pressure greater than 140/90 mm Hg and in the elderly as blood pressure greater than 160/95 mm Hg, as recommended by the Framingham Study. The electrocardiograms were reviewed for 1) cardiac rhythm; 2) left ventricular hypertrophy, as defined by $V_{15} + V_{s5}$ and/or $V_{25} + V_{s5} ≥ 35$ mm and/or R in aVL ≥ 11 mm; 3) presence of a Q wave in the anterior septal or lateral precordial leads; and 4) intraventricular conduction abnormalities.

Echocardiographic Data

Echocardiographic studies were performed with either a Hewlett-Packard instrument with 2.5/1.9-MHz transducer (Andover, Massachusetts), an Aloka Model 880 instrument with 3.5/2.0-MHz transducer (Tokyo, Japan), or an Irex Meridian instrument with 3.5/2.0-MHz transducer (Ramsey, New Jersey). All images were recorded on 1/2-in. VHS video tape recorders. Tapes were reviewed on a Panasonic editing controller system (Osaka, Japan) with a Panasonic AG-6500 video recorder. Each echocardiographic study was reviewed by three investigators; two were unaware of patient age and clinical data. Based on an initial blinded review, patients with two-dimensional echocardiograms that were technically inadequate for detailed analysis were discarded from the study, and this was the only exclusion criterion. After an initial review of the echocardiograms, the following morphologic characteristics were identified as features for analysis during formal prospective evaluation of the echocardiograms.

Septal curvature and cavity contour. Septal curvature was evaluated in the apical four-chamber view. An abnormal convexity of the septum toward the left ventricular cavity was identified as reversed septal curvature, which produced a crescent-shaped left ventricular cavity; slight concavity toward the left ventricular cavity or a more straight septal contour was considered as normal septal curvature. The latter produced an ovoid left ventricular cavity. Cavity contour was assessed in systole and diastole.

Proximal septal bulge. Proximal septal bulge was defined as a subaortic protrubance of the septum toward the left ventricular outflow tract with the middle and distal septum having normal shape. This was assessed in the parasternal long-axis and apical four-chamber views.

Symmetry or asymmetry. Symmetry or asymmetry was determined by M-mode and two-dimensional echocardiography. Measurements of wall thickness that led to the final determination were obtained from M-mode echocardiography. Wall thickness measurements were made according to the American Society of Echocardiography recommendations for measurements of the septum and posterior wall of the left ventricle. The right ventricular free wall prominence. Right ventricular free wall prominence was evaluated in the apical four-chamber view. It was described when the right ventricular free wall appeared increased in thickness and when muscular obliteration of the moderator band and apex occurred, leading to loss of the normal crescent shape of the cavity. To help substantiate right ventricular free wall prominence, the parasternal short- and long-axis views were used also whenever they were well visualized.
Left ventricular outflow gradient. Left ventricular outflow gradient was measured either by continuous-wave Doppler echocardiography or at the time of cardiac catheterization. The Doppler measurements were made in meters per second and then converted to millimeters of mercury with the simplified Bernoulli equation $p = 4V^2$.

Mitral anular calcification. Mitral anular calcification was described as an increased echocardiographic density of the mitral anulus in the parasternal long-axis, short-axis, or four-chamber views.

Systolic anterior motion of the mitral valve. Systolic anterior motion of the mitral valve was determined with M-mode and two-dimensional echocardiography when a displacement of the anterior or posterior leaflets or both toward the septum in systole was observed, with or without amyl nitrite provocation.

Interobserver and Intraobserver Variability of Cardiac Shape

Thirty-six randomly identified echocardiograms were reviewed at a separate sitting, without knowledge of patient identity or prior readings, and a consensus of 97% was achieved in determining left ventricular shape. Another echocardiographer who was not part of the original reading (P.J.C.) reviewed independently the same 36 echocardiograms to assess interobserver variability and agreed with the consensus assessment of reversed septal curvature in 94% of the echocardiograms. Intraobserver and interobserver variability for the proximal septal bulge was lower; the two reviewers agreed 84% of the time. Intraobserver and interobserver variability for right ventricular prominence was 92% and 78%, respectively.

Data Analysis

Student’s $t$ test was used to compare mean values between groups. Associations for contingency table data were tested with the $\chi^2$ statistic when appropriate. For tables with small expected cell frequencies, exact tests (Fisher’s exact or an exact $\chi^2$) were used. $p$ values of 0.01 or less were considered statistically significant.

Results

Between January 1986 and January 1987, 34 patients 65 years of age and older were identified. Six of these 34 patients were excluded from the study because their two-dimensional echocardiograms were technically unsatisfactory for analysis; the remaining 28 patients form the basis of this study. No patients had systemic disease or hypertension severe enough to warrant exclusion. To establish the comparison group of 28 young patients, we had to review our records consecutively back to 1982. Thirty-three patients less than age 40 were identified, and five were excluded because of poor quality echocardiograms. Thus, a total of 56 patients make up the study groups.

<table>
<thead>
<tr>
<th>Findings</th>
<th>Elderly (Age ≥65 yr, $n = 28$)</th>
<th>Young (Age &lt;40 yr, $n = 28$)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>72±6.4 yr</td>
<td>26±9.5 yr</td>
</tr>
<tr>
<td>Range</td>
<td>65-89 yr</td>
<td>9-39 yr</td>
</tr>
<tr>
<td>Sex</td>
<td>male 15 (53)</td>
<td>female 22 (79) $p = 0.05$</td>
</tr>
<tr>
<td>Symptoms</td>
<td>dyspnea 24 (86)</td>
<td>palpitations 17 (61)</td>
</tr>
<tr>
<td></td>
<td>angina 13 (46)</td>
<td>syncope 6 (21)</td>
</tr>
<tr>
<td>Murmur grade</td>
<td>0 2 2</td>
<td>1 1 1</td>
</tr>
<tr>
<td>New York Heart Association classification</td>
<td>I 4 (14)</td>
<td>II 13 (46)</td>
</tr>
<tr>
<td></td>
<td>III 10 (36)</td>
<td>IV 1 (4)</td>
</tr>
<tr>
<td></td>
<td>Left ventricular hypertrophy 18 (64)</td>
<td>23 (82)</td>
</tr>
<tr>
<td></td>
<td>atrial fibrillation 7 (25)</td>
<td>1 (4) $p = 0.05$</td>
</tr>
<tr>
<td></td>
<td>right bundle branch block 4 (14)</td>
<td>3 (11)</td>
</tr>
<tr>
<td></td>
<td>left bundle branch block 2 (7)</td>
<td>5 (18)</td>
</tr>
<tr>
<td>Q waves: anteroseptal or lateral or both</td>
<td>8 (29)</td>
<td>14 (50)</td>
</tr>
<tr>
<td></td>
<td>normal 1 (4)</td>
<td>1 (4)</td>
</tr>
<tr>
<td>Systemic hypertension</td>
<td>13 (46)</td>
<td>5 (18) $p = 0.02$</td>
</tr>
</tbody>
</table>

Data in parentheses are percentages for that patient group.

Clinical Findings

Elderly patients. The elderly patients ranged in age from 65 to 89 years, mean 72±6.4 years, $n = 28$ (Table 1). In the elderly group, four patients (14%) were in NYHA functional Class I, 13 (46%) in Class II, 10 (36%) in Class III, and one (4%) in Class IV. The most frequent symptoms were dyspnea in 24 (86%), palpitations in 17 (61%), angina in 13 (46%), and syncope in six (21%) patients. Of the 13 patients with anginal pain, eight underwent coronary angiography. In six of them (75%), no coronary obstructions greater than 40% occurred; in the other two, narrowings of 60% and 50%, respectively, occurred in the left anterior descending coronary arteries. Hypertension, defined by either a systolic blood pressure of more than 160 mm Hg or a diastolic blood pressure of more than 95 mm Hg, was present in 13 elderly patients (46%). Of the 28 patients, 17
(61%) had either a systolic pressure greater than 140 mm Hg or a diastolic pressure greater than 90 mm Hg or both. The duration of hypertension was documented in 11 (85%) of 13 patients and ranged from 1 to 20 years (average, 10 years). Left ventricular hypertrophy by echocardiography was detected in 18 (64%) patients, and atrial fibrillation was present in seven (25%). No clinical differences existed in the elderly group between hypertensive and nonhypertensive patients.

Young patients. The comparison group of younger patients ranged in age from 9 to 39 years, mean 26±9.5 years, n=28 (Table 1). Five patients (18%) were in NYHA functional Class I, 15 (54%) in Class II, eight (28%) in Class III, and none in Class IV. The most frequent symptoms were palpitations in 23 patients (82%), dyspnea in 20 (71%), angina in 12 (43%), and syncope in six (21%). One patient had atrial fibrillation. Left ventricular hypertrophy by echocardiography was detected in 23 patients (82%). Of the 12 young patients who had angina, six had coronary angiograms, all of which were normal. Five patients (18%) were hypertensive with a blood pressure greater than 140/90 mm Hg. The duration of hypertension was documented in three patients and ranged from 2 to 11 years (average, 6 years).

Comparison of the clinical variables between the two age groups showed three clinical factors that were barely significant: a higher incidence of atrial fibrillation (p=0.05) and of systemic hypertension (p=0.02) in the elderly and a greater predominance of male sex in the younger group (p=0.05). No differences existed between the two groups concerning clinical symptoms, the presence of cardiac murmurs, or echocardiographic findings of left ventricular hypertrophy.

Echocardiographic and Hemodynamic Findings

Elderly patients. All 28 patients 65 years and older had septal hypertrophy ≥15 mm (average, 19 mm) (Table 2). The left ventricular posterior wall was hypertrophied in 18 patients by ≥13 mm (average, 15 mm). Asymmetry, defined as a septal to posterior wall ratio of ≥1.3, was present in 20 (71%) patients. A normal septal curvature with an ovoid left ventricle was present in 24 (86%) elderly patients (Figure 1). A proximal septal bulge was observed by two-dimensional echocardiographic long-axis and apical four-chamber views in 15 (54%) elderly patients, all of them with an ovoid left ventricle (Figures 2 and 3). Reversed septal curvature, which is characterized by a midseptal convexity toward the left ventricular cavity and a crescent shape of the left ventricle, was present in four (14%) elderly patients. Right ventricular free wall prominence was present in seven (25%) elderly patients, but only three (13%) of the 24 patients with normal left ventricular shape had a prominent right ventricular free wall by qualitative assessment. Systolic anterior motion of the mitral valve was present in 20 (71%) patients. Fourteen of these patients had a gradient over 20 mm Hg. Cardiac catheterization in eight patients showed a resting gradient in four and a provokable gradient in three. Measurements by Doppler echocardiography or at cardiac catheterization showed a gradient >20 mm Hg in 17 patients. Eight (29%) were at rest, and nine (32%) were provokable. The mean gradient for the elderly group was 70 mm Hg. Ten of the 15 patients with a proximal septal bulge had a gradient greater than 20 mm Hg. Six had resting, and an additional four had provokable left ventricular outflow obstruction. Mitral anular calcification was present in 15 (54%) patients. When the elderly group was subdivided into those with and without hypertension, no significant differences existed between the groups concerning measures of ventricular shape, systolic anterior motion of mitral valve, left ventricular hypertrophy, proximal septal bulge, or other morphologic features distinguished by echocardiography.

Young patients. Echocardiographic findings in the young were similar to those in the elderly concerning asymmetry, systolic anterior motion of the mitral valve, and a left ventricular outflow tract gradient greater than 20 mm Hg (Table 2). At cardiac catheterization, six of the 11 patients with hemodynamic studies had gradients at rest and five others with provocation. Resting gradients determined by echocardiography or catheterization were

<table>
<thead>
<tr>
<th>Table 2. Echocardiographic and Hemodynamic Data for Elderly and Young Patients With Hypertrophic Cardiomyopathy</th>
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<tbody>
<tr>
<td>Findings</td>
</tr>
<tr>
<td>Age ≥65 yr, n=28</td>
</tr>
<tr>
<td>Dimensions</td>
</tr>
<tr>
<td>Septum (mm)</td>
</tr>
<tr>
<td>15-20</td>
</tr>
<tr>
<td>&gt;20</td>
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<tr>
<td>Mean±SD</td>
</tr>
<tr>
<td>Mean posterior wall thickness (mm)</td>
</tr>
<tr>
<td>Posterior wall ratio &gt;1.3</td>
</tr>
<tr>
<td>M-mode septum/posterior wall ratio &gt;1.3</td>
</tr>
<tr>
<td>Systolic anterior motion of the mitral valve</td>
</tr>
<tr>
<td>Gradient</td>
</tr>
<tr>
<td>At rest</td>
</tr>
<tr>
<td>With provocation</td>
</tr>
<tr>
<td>Ovoid ventricle</td>
</tr>
<tr>
<td>Reversed septal curvature</td>
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<tr>
<td>Proximal septal bulge</td>
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<tr>
<td>Prominent right ventricular free wall</td>
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<tr>
<td>Mitral anular calcification</td>
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Data in parentheses are percentages for that patient group.
present in 13 (46%) patients and were provokable in an additional 9 (32%) patients. Left ventricular hypertrophy was significantly greater in the young than in the elderly for both the interventricular septum ($p=0.006$) and the posterior wall ($p=0.002$). All of the young patients over the age of 10 years had septal wall thickness $\geq 15$ mm. Two young patients, aged 9 and 10, had septal wall thicknesses of 13 mm, which is significantly increased for body surface area. In contrast to the elderly, a proximal septal bulge and mitral anular calcification were not observed in any of the young. Reversed septal curvature, with a distinctive crescent shape of the left ventricular cavity, was prominent in the young and was present in 21 (75%) (Figures 4 and 5). Normal septal curvature and the more normal ovoid ventricular cavity were present in seven (25%) patients. A prominent right ventricular free wall occurred in 18 (72%) of the 25 patients in which the right ventricular wall could be satisfactorily visualized. These shape differences between elderly and young were highly significant ($p<0.0001$).

In five patients, color Doppler flow mapping studies were available to assess the site of outflow obstruction. In the two elderly patients, the color flow acceleration was at the proximal septal bulge. In the three young patients, in whom there was systolic anterior motion, the point of contact between the mitral valve and the septum in systole was the origin of color flow acceleration. A sufficient number of patients with color flow studies was not obtained, however, to conclude whether or not the sites of obstruction or ventricular flow patterns differ between the two groups of patients.

In brief, the overall shape of the heart in hypertrophic cardiomyopathy, as assessed by two-dimensional echocardiography, was strikingly different between the young and the elderly. In the young, who had significantly thicker hearts, a reversal of septal curvature with a prominent right ventricular free wall predominated, whereas in the elderly, a more ovoid ventricular shape and proximal septal bulge occurred, regardless of the presence or absence of hypertension (Figure 6).

**Discussion**

Hypertrophic cardiomyopathy is a disease that afflicts the elderly. The identification of 28 elderly patients with hypertrophic cardiomyopathy during a 1-year period at a single institution indicates that the condition is not a rare occurrence. Moreover, in our referral patient population, hypertrophic cardiomyopathy was present approximately twice as often in the elderly as in the young, supporting the notion that the elderly may be a large and neglected subset of patients with this disease. Whether or not the disease takes the same form in the elderly, has the same genetic basis, carries the same prognosis,
has the same pathophysiology, however, is by no means clear.

In this study, our aim was to determine whether or not hypertrophic cardiomyopathy in the elderly is the same disease that is commonly described in the young. That is, is it the disease that Teare described in young patients who died suddenly and who had a severely hypertrophied interventricular septum, which was generally characterized by asymmetry of the left ventricle, a resting or provokable obstruction to left ventricular outflow, and transmitted as an inherited disease, or is it a distinctly different disease? These questions are important to our understanding of hypertrophic cardiomyopathy and bear on the issue of whether or not hypertrophic cardiomyopathy is a heterogeneous spectrum of disease or a single disease. Our approach was to compare the group of elderly patients having hypertrophic cardiomyopathy with a group of patients diagnosed as having the disease by the same criteria but in the more typical age range of less than 40 years. The two age groups were similar concerning clinical presentations of chest pain, dyspnea, palpitations, and cardiac murmurs and in measures of left ventricular asymmetry and outflow tract gradient.

There were three marginally significant clinical differences between the groups: a greater predominance of males with the disease in the young and more atrial fibrillation and hypertension in the elderly. When we compared patients with and without hypertension, no differences occurred in clinical or echocardiographic findings. Also, the prevalence of hypertension, which was mild and controlled in most patients, mirrored the prevalence of blood pressure elevation in the public at large by age.

The most striking and significant differences between the young and elderly were in the gross structural features of the ventricles as assessed by echocardiographic determination of cardiac size and shape.

**Echocardiographic Distinctions in the Elderly and Young**

Echocardiographic findings of asymmetric hypertrophy of the interventricular septum and of systolic anterior motion of the mitral valve were similar in the young and elderly groups. Both groups also had a similar prevalence and severity of an outflow tract gradient, although the younger group had more severe hypertrophy as determined by free wall and interventricular septal measurements. Of importance, analysis of the hearts by two-dimensional echocardiography showed significant differences in cardiac shape between the two groups, differences that seem to have gone largely unrecognized.

The young group had marked abnormalities in ventricular cavity contours, which is consistent with several clinical studies of the heart in hypertrophic cardiomyopathy. The septum, more hypertrophied in the young group, had a markedly abnormal shape with a reversal of septal curvature in most patients. The reversal of septal curvature (Figures 4 and 5) was associated with a crescent-shaped, rather than an ovoid, left ventricular cav-

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**FIGURE 2.** Two-dimensional echocardiogram in the parasternal long-axis view from a 66-year-old man. Arrows point to the proximal bulge of the septum. A schema is shown above. RV, right ventricle; LV, left ventricle; LA, left atrium.

**FIGURE 3.** Two-dimensional echocardiogram in the apical four-chamber view from a 67-year-old man. The cavity has a normal ovoid shape, and the septum has normal curvature. Arrows point to the bulge in the proximal portion of the septum. LV, left ventricle; RV, right ventricle; RA, right atrium; LA, left atrium.
ity, which was only detectable by two-dimensional echocardiograms. The septum was also diffusely thickened relative to the free wall and had the more typical shape of asymmetric septal hypertrophy reported at autopsy by Teare\(^6\) and others.\(^{17,18,24}\) Although septal asymmetry and microscopic structures have been the subject and emphasis of most studies, some reports have described abnormal septal shape in hypertrophic cardiomyopathy. Roberts and Ferrans\(^25\) described a slitlike cavity with S-shaped configuration. Hutchins and Bulkley,\(^24\) in a detailed autopsy study of the shape of the interventricular septum, described a reversal of normal septal curvature in patients with hypertrophic cardiomyopathy of the morphologic type reported by Teare.\(^6\) That study also described a more globular configuration of the right ventricular cavity in these cardiomyopathic hearts. The reversal of septal curvature was not present in normal and other types of diseased hearts, including those with isolated left ventricular hypertrophy.

The reversal of septal curvature probably reflects a thickened and bizarrely shaped septum, which has a concave contour to the left rather than the normal convex curvature. The crescent-shaped cavity reflects the abnormal septal configuration but not asymmetric septal hypertrophy per se. This abnormality of curvature is not detected by M-mode echocardiography and can be seen only in the apical four-chamber view by two-dimensional echocardiography. Prior methodologic limitations may have led to an emphasis on a one-dimensional septal measurement rather than two-dimensional left ventricular cavity contour.

Abnormalities of the right ventricle were also present in the young group and were not frequent in the elderly. In most of the young hearts, the right ventricular cavity had a more ovoid contour rather than the normal crescent shape (Figures 4 and 5). This change was also likely due to abnormal septal curvature, with the left ventricle taking on a shape more like the normal crescentic right ventricle and with the right ventricle taking on a more ovoid shape like the normal left ventricular cavity (Figure 5). In addition to differences in cavity contour, the right ventricle was prominent by echocardiography in most of the young patients but rarely in the elderly. The presence of right ventricular hypertrophy was described as a universal feature of hearts with the Teare type of morphology studied at autopsy by Roberts and Ferrans\(^25\) and Roberts and colleagues.\(^26\) This abnormality of the right ventricle has largely gone unrecognized, possibly because it too had been difficult to assess in vivo before the advent of two-dimensional echocardiography. We recognize, however, that quantitative methods to assess right ventricular mass are still not available, thus limiting a more detailed analysis of the right ventricle.

The structural features of the hearts of most of our elderly patients with hypertrophic cardiomyop-

**Figure 4.** Two-dimensional echocardiogram in the apical four-chamber view from a 19-year-old man with a hypertrophied left ventricle and reversal of septal curvature. A schema is shown above. RV, right ventricle; LV, left ventricle; RA, right atrium; LA, left atrium.

**Figure 5.** Two-dimensional echocardiogram in the apical four-chamber view from a 31-year-old man, which shows right ventricular prominence and the abnormal curvature of the interventricular septum. RV, right ventricle; LV, left ventricle; RA, right atrium; LA, left atrium.
Aortic bulge in the septum. Using paraffin casts at autopsy, they showed that, in some, the bulging of the proximal septum and the rightward angulation of the aorta narrowed the left ventricular outflow tract. They also found a focal fibrous plaque on the ventricular septum similar to that in hypertrophic cardiomyopathy, which is an anatomic marker of systolic anterior motion of the mitral valve and of an outflow tract obstruction that could only be inferred by an autopsy study. Thus, the development of a proximal bulge—which we identified in half of our elderly patients and in none of the young—may be a shape change acquired with advancing age. Moreover, most of our patients with a bulge similar to that described by Goor et al\(^29\) had an outflow tract gradient, and color Doppler flow mapping studies in two of these patients identified the flow acceleration at the site of the proximal bulge. Others have also described a proximal bulge by two-dimensional echocardiography\(^31\) and have shown specifically that the unusual septal configuration can be associated with left ventricular outflow tract gradient.\(^{22,32,33}\) Recently, Iida et al\(^32\) showed provocable obstruction at the time of cardiac catheterization in a 67-year-old man who had a sigmoid septum by two-dimensional echocardiography. Dalldorf and Willis\(^22\) also showed noninvasively the presence of left ventricular outflow tract obstruction in patients who had a sigmoid septum at autopsy, concluding that these patients had a distinctly different form of hypertrophic cardiomyopathy than generally recognized. If advancing age can cause this alteration in ventricular shape, characterized by a proximal bulge in the interventricular septum, and if the latter causes a subvalvular gradient, then patients may develop the clinical stigmata of hypertrophic cardiomyopathy only when they are elderly.

Hypertrophic Cardiomyopathy: A Heterogeneous Disease Spectrum

Our findings show that at either end of the age spectrum, distinctly different forms of hypertrophic cardiomyopathy predominate, which suggests that different disease processes may be involved. Hypertrophic cardiomyopathy in the young as described by Teare\(^16\) and subsequently by others\(^24\) is associated with marked abnormalities of the septum, including reversal of septal curvature. We observed this septal shape abnormality in most of our young patients. In contrast, however, hypertrophic disease in our elderly patients was usually localized to the left ventricle, and normal myocardial shape was generally preserved, except for the frequently present exaggerated proximal septal bulge. In these elderly patients, acquired factors, such as shrinking of the heart, angulation of the aorta and septum, diminished vascular compliance, and mild systemic hypertension, which are all associated with aging, although not exclusively, may serve as stimuli to hypertrophy. This latter type of hypertrophic car-

Change in Cardiac Shape With Aging

Kitzman et al\(^30\) in an autopsy series showed that in the 7th through 10th decades of life in presumably normal people a significant increase occurs in septal thickness, with the ratio of septum to free wall thickness often exceeding 1.3. Goor et al\(^29\) have described the development of a “sigmoid septum” in elderly patients. In an autopsy study, these investigators showed a progressive change in the shape of the heart with aging, characterized by a rightward shift in the aorta with respect to the ventricular septum, and the development of a sub-

Figure 6. A schema compares the different cardiac shapes observed in the study patients. RV, right ventricle; LV, left ventricle.
diomyopathy seen in most of our elderly patients and described in similar forms by others22,34,35 closely resembles the disease originally described by Brock.1
Indeed, in Brock’s earliest report of hypertrophic cardiomyopathy, he described a subvalvular muscular subaortic “functional” obstruction localized at the left ventricle, occurring in a 63-year-old woman with a history of hypertension. The left ventricular hypertrophy was concentric, and in Brock’s estimation, systemic hypertension was probably a contributing factor. The alternative hypothesis that one disease process is present that is in a milder form in those who live to an advanced age has not been excluded, however, and resolution will require further insight into the molecular mechanisms underlying both forms of the disorder.

In summary, our findings show that there are distinct differences in the hypertrophic cardiomyopathy predominating in the elderly compared with that in young patients. Although clinical features may be similar, the disease states seem heterogeneous based upon cardiac shape. Just as all aortic stenosis is not due to a congenitally malformed bicuspid valve, all hypertrophic cardiomyopathy may not be “Teare’s disease.” Such distinctions can offer a framework for further needed studies of etiology, prognosis, treatment, and particularly the molecular and genetic bases of these disorders.

References


**KEY WORDS:** echocardiography • idiopathic hypertrophic subaortic stenosis • right ventricular hypertrophy
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