Echocardiographic Diagnosis of Idiopathic Hypertrophic Cardiomyopathy without Outflow Obstruction


SUMMARY

The echocardiographic findings of eight patients with hypertrophic cardiomyopathy without outflow obstruction (HMC) and of 15 normal (Norm) individuals are presented.

The characteristic features in HMC were: (1) interventricular septal width much greater than normal (HMC = 2.5 ± 0.3 cm, Norm = 1.0 ± 0.2 cm, P < 0.005); (2) normal or only slightly increased posterior left ventricular wall thickness; (3) the ratio of interventricular septal to posterior wall thickness ≥2.0; (4) ejection fraction greater than normal (HMC = 0.76 ± 0.08, Norm = 0.68 ± 0.06, P < 0.025); (5) reduced velocity of the early diastolic closing motion of the anterior mitral leaflet (HMC = 60 ± 23 mm/sec, Norm = 124 ± 29 mm/sec, P < 0.005); (6) absence of abnormal systolic movement of the anterior mitral valve, as seen in hypertrophic obstructive cardiomyopathy. The diagnosis of hypertrophic cardiomyopathy can be made with echocardiography, even when outflow tract obstruction of the left ventricle is absent.

Additional Indexing Words:
Interventricular septal hypertrophy
Idiopathic hypertrophic obstructive cardiomyopathy

HYPERTROPHIC cardiomyopathy is difficult to diagnose clinically. Cardiac catheterization, angiocardiology, and coronary arteriography are often necessary for the correct diagnosis of chest pain, dyspnea, an unexplained heart murmur, or an abnormal electrocardiogram with which these patients may present. When obstruction to the left ventricular outflow is present in this condition, systolic approximation of the mitral valve to the interventricular septum can be demonstrated as a characteristic deformity by echocardiography.1-2 However, in nonobstructive hypertrophic cardiomyopathy,3-5 there is no gradient across the left ventricular outflow tract and an abnormal systolic movement of the mitral valve is not seen on echocardiography, even after amyl nitrite or isoproterenol provocation.6 The diagnosis in cases without outflow obstruction is based on angiographic demonstration of left ventricular wall and interventricular septal hypertrophy, diminished end-systolic volume, and papillary muscle hypertrophy.7-8 In some cases the hypertrophy may be restricted to the ventricular septum.8-10 The ventricular septal hypertrophy is difficult to assess with isolated left ventricular angiograms and simultaneous left and right ventriculography has been advocated for this diagnosis.11 These procedures have been advocated for this diagnosis.11

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the obvious drawbacks of potential risk, expense, and patient inconvenience.

Recent advances in ultrasound technics have made it possible to estimate left ventricular volume, left ventricular wall thickness, and width of the interventricular septum noninvasively.12-15

We are reporting the ultrasound findings of eight patients with hypertrophic cardiomyopathy without left ventricular outflow obstruction.

Methods

Eight patients were studied; three were men and five were women. The ages of the patients ranged from 26 to 64 years with a mean age of 44 years. Four patients presented with chest pain as their leading symptom. Five patients had dyspnea on exertion; two were referred because of an abnormal electrocardiogram, suggesting old myocardial infarction in one and left ventricular hypertrophy in the other.

Five patients had cardiac catheterization, angiocardiology, and coronary arteriography. None of these five patients had a gradient between the left ventricle and the brachial artery at rest, and only one had a gradient of 20 mm Hg during isoproterenol infusion. The diagnosis of hypertrophic cardiomyopathy in these patients was based on the angiocardiology features described by Simon7 and Cohen.8 Three patients who did not have cardiac catheterization had clinical evidence suggesting hypertrophic cardiomyopathy.9 10 The clinical features included evidence of left ventricular hypertrophy with normal blood pressure, rapid carotid upstroke, prominent fourth heart sound, and absence of ejection click. A short systolic murmur grade 1-2/6 was also present at the apex or lower left sternal border. Echocardiograms were also taken of 15 healthy volunteers.

Echocardiographic Measurements

Echocardiograms were performed using a commercially available ultrasonoscope* utilizing a 2.25 MHz, 0.5 cm-diameter transducer with a 10 cm focus† and a repetition rate of 1,000 impulses per sec. The calibration dots on the oscilloscope were set so that adjacent dots were separated horizontally by 0.5 sec, and represented 1 cm tissue depth vertically. The technic of recording the echogram was that described by Feigenbaum et al.13 and Popp and Harrison.14 In brief, the left ventricular cavity size was determined by placing the transducer at the fourth or fifth left intercostal space, close to the sternum. The transducer was then rotated slightly superiorly and mediolaterally to localize the characteristic motion of the anterior mitral leaflet, and a record of its motion was made. From this position, the transducer was

Table 1

Echocardiographic Measurements of Eight Patients with Hypertrophic Cardiomyopathy and 15 Normal Subjects

<table>
<thead>
<tr>
<th>Pt</th>
<th>BSA (m²)</th>
<th>Sd (cm)</th>
<th>Ss (cm)</th>
<th>EDV (ml)</th>
<th>ESV (ml)</th>
<th>EF</th>
<th>IVS (cm)</th>
<th>LVPW (cm)</th>
<th>IVS/LVPW</th>
<th>MVDS (mm/sec)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1.58</td>
<td>3.8</td>
<td>2.2</td>
<td>57.5</td>
<td>11.2</td>
<td>0.81</td>
<td>3.0</td>
<td>1.5</td>
<td>2.0</td>
<td>54</td>
</tr>
<tr>
<td>2</td>
<td>1.88</td>
<td>4.0</td>
<td>2.4</td>
<td>67.1</td>
<td>14.4</td>
<td>0.79</td>
<td>2.2</td>
<td>1.1</td>
<td>2.0</td>
<td>70</td>
</tr>
<tr>
<td>3</td>
<td>1.51</td>
<td>4.4</td>
<td>3.0</td>
<td>89.2</td>
<td>28.2</td>
<td>0.68</td>
<td>2.5</td>
<td>1.2</td>
<td>2.2</td>
<td>32</td>
</tr>
<tr>
<td>4</td>
<td>1.65</td>
<td>5.0</td>
<td>3.5</td>
<td>130.9</td>
<td>44.8</td>
<td>0.66</td>
<td>2.5</td>
<td>1.0</td>
<td>2.5</td>
<td>110</td>
</tr>
<tr>
<td>5</td>
<td>1.66</td>
<td>4.5</td>
<td>3.0</td>
<td>95.4</td>
<td>28.2</td>
<td>0.70</td>
<td>2.7</td>
<td>1.2</td>
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<td>2.2</td>
<td>130.9</td>
<td>11.2</td>
<td>0.91</td>
<td>2.0</td>
<td>0.8</td>
<td>2.5</td>
<td>64</td>
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<td>3.0</td>
<td>101.8</td>
<td>28.0</td>
<td>0.72</td>
<td>2.5</td>
<td>1.0</td>
<td>2.5</td>
<td>60</td>
</tr>
<tr>
<td>8</td>
<td>1.79</td>
<td>5.2</td>
<td>3.0</td>
<td>147.2</td>
<td>28.0</td>
<td>0.81</td>
<td>2.6</td>
<td>1.0</td>
<td>2.5</td>
<td>42</td>
</tr>
<tr>
<td>Mean</td>
<td>1.73</td>
<td>—</td>
<td>—</td>
<td>102.5</td>
<td>24.3</td>
<td>0.76</td>
<td>2.5</td>
<td>1.1</td>
<td>2.3</td>
<td>60</td>
</tr>
</tbody>
</table>

≈ SD

| Mean | 1.85 | — | — | 125.4 | 40.1 | 0.68 | 1.0 | 0.9 | 1.17 | 124 |

p = 0.16

| Mean | — | — | — | 29.0 | 14.0 | 0.06 | 0.2 | 0.2 | 0.14 | 29 |

Abbreviations: BSA = body surface area; Sd = end-diastolic short axis; Ss = end-systolic short axis; EDV = end-diastolic volume; ESV = end-systolic volume; EF = ejection fraction; IVS = interventricular septum; LVPW = left ventricular posterior wall; MVDS = mitral valve diastolic slope; SD = standard deviation; NS = not significant.

*Cokine 20, Smith-Kline Instruments.
†Aerotech Laboratories.

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rotated slightly laterally and inferiorly to record the posterior left ventricular wall echo just behind the mitral echo. In order to delineate the ventricular septal echo, the depth compensation was adjusted so that maximum gain was placed close to 0 cm on the oscilloscope. In this way, the entire width of the septum was clearly seen separated from the anterior heart wall. All the patients had echocardiograms taken before and after inhalation of amyl nitrite.

The left ventricular minor diameter was measured echocardiographically between the endocardial echo of the posterior left ventricular wall and the endocardial border of the left septal surface. The minor diameter at end-diastole (Sd) was measured at the time of the R wave of the simultaneously recorded electrocardiogram, and that at end-systole (Ss) was taken at the point where the endocardial surface of the septum and the posterior wall were nearest to each other. The ventricular volumes were estimated by: EDV = 1.047(Sd)^3; ESV = 1.047(Ss)^3, where EDV is end-diastolic volume and ESV is end-systolic volume. The ejection fraction (EF) was calculated as: \( EF = \frac{EDV - ESV}{EDV} \). The estimation of the ventricular volume from the echocardiographic short axis of the left ventricle has been validated recently. The reproducibility of the measurements among observers, and at different times in the same patient, has been documented by Pombo et al. The measurement of posterior left ventricular wall thickness and ventricular septal width was made at end-diastole. The early diastolic slope of the mitral valve was estimated as described by Edler.

**Results**

Table 1 shows echocardiographic features of the patients with nonobstructive hyper-
Hypertrophic cardiomyopathy (HMC) and of 15 normal individuals. In patients with HMC, both end-diastolic and end-systolic volumes were relatively smaller than normal, but the difference was not statistically significant. The ejection fraction was significantly greater in HMC than in normal subjects ($P < 0.025$). The most striking difference was seen in the interventricular septal (IVS) width, which in HMC was two and a half times greater than the normal (with no overlap of values between HMC and normals). There was no significant difference between posterior left ventricular wall thickness in the two groups. The early diastolic slope of the anterior leaflet of the mitral valve in HMC was significantly less than in the normal group ($P < 0.005$); only one patient with HMC had a valve slope in the normal range. An abnormal systolic movement was not seen in any patient, even after amyl nitrite inhalation.

Figure 1 shows an echocardiogram of a normal individual. The IVS (0.8 cm), the short axis at end-diastole (Sd) and at end-systole (Ss), the endocardium and epicardial-pericardial junction of the posterior left ventricular wall, and its thickness (0.8 cm), are well seen.

Figure 2 shows a record of a patient with HMC. Of note are the wide ventricular septum (2.5 cm), the relatively small left ventricular cavity size, and normal posterior left ventricular wall thickness (0.8 cm). Also of interest is the strong echo from the left side of the ventricular septum, suggesting a thickened endocardium.

**Figure 2**

Echocardiogram of a patient with hypertrophic cardiomyopathy without outflow obstruction. Note the wide IV Septum (2.5 cm) and normal posterior left ventricular thickness (0.8 cm). Abbreviations same as in figure 1.
Figure 3 shows the mitral valve motion of a patient with HMC without obstruction. Note the absence of anterior systolic movement of the mitral valve. This echocardiogram was taken after inhalation of amyl nitrite. In contrast to this, figure 4 shows the motion of the mitral valve of a patient with hypertrophic obstructive cardiomyopathy. The systolic anterior movement of the mitral valve toward the ventricular septum is quite well seen.

Discussion

Autopsy studies of patients with hypertrophic cardiomyopathy with or without outflow obstruction have shown two types of myocardial hypertrophy. The most common variety is an asymmetric hypertrophy\textsuperscript{9,19–22} mainly involving the interventricular septum and, to some extent, the free anterolateral wall of the left ventricle. The posterior left ventricular wall may be entirely normal. A much less common variety is that of diffuse hypertrophy\textsuperscript{21–23} involving the left ventricle in more or less symmetric fashion. Our cases appear to be of the asymmetric type, as the posterior left ventricular wall was within normal limits, except in one case. The ventricular septum, however, was considerably thicker than normal.

Menges\textsuperscript{24} in his three autopsy cases of hypertrophic cardiomyopathy reported ventricular septal thicknesses of 2.8, 2.0, and 3.1 cm. In his series of normal cases, the thickness of the ventricular septum was 1.0–1.5 cm (average 1.3 cm). The values for ventricular septal width in the present study agree well

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure3.png}
\caption{The mitral valve of a patient with hypertrophic cardiomyopathy without obstruction, after inhalation of amyl nitrite. Both the anterior (AML) and posterior (PML) valve leaflets are seen. Note the absence of systolic anterior motion of the mitral valve.}
\end{figure}
The anterior mitral leaflet (AML) of a patient with hypertrophic cardiomyopathy with outflow obstruction. Note the prominent systolic anterior motion of the mitral valve (SAM).

with those found by Menges. In the three cases of hypertrophic cardiomyopathy that Menges analyzed, the ratio of thickness of ventricular septum to that of free anterolateral wall ranged between 1.55 and 1.76. In 20 normal hearts, this ratio averaged 0.45 and in 50 hypertrophied hearts (other than hypertrophic cardiomyopathy) it averaged 0.98, exceeding 1.25 in only one instance. Thus, it seems that cardiac hypertrophy associated with hypertension and aortic stenosis is symmetric, affecting uniformly the left ventricular wall and the ventricular septum. However, in hypertrophic cardiomyopathy, the ventricular septum is disproportionately more hypertrophied.

Since the posterior left ventricular wall is least affected by hypertrophy in hypertrophic cardiomyopathy, the ratio of the ventricular septum to the posterior left ventricular wall becomes even more significant. In our series of hypertrophic cardiomyopathy, this ratio was $\leq 2.0$ in every case.

The septal width, as measured by echocardiography, may overestimate the actual width of the septum because the ultrasound beam may not be directed at a right angle to the septal surfaces. However, the technique we employed was highly standardized and reproducible, and the empiric difference between the hypertrophic cardiomyopathy and normal is highly significant ($P < 0.005$).

The echocardiographic ventricular volumes in our normal series and in hypertrophic cardiomyopathy without outflow obstruction are in agreement with the angiocardiographic volumes in normal individuals and in hypertrophic cardiomyopathy with or without outflow obstruction reported by Conn and
associates, particularly when corrected for body surface area.

The rate of early diastolic closing motion of the anterior mitral leaflet was significantly reduced in all but one of the patients with hypertrophic cardiomyopathy. However, this has also been noted in patients with aortic stenosis, and may reflect slow left ventricular filling due to reduced left ventricular distensibility in these patients.1 25

The hallmark of hypertrophic cardiomyopathy is an abnormally thick ventricular septum with a ratio of septum to posterior left ventricular wall thickness $\geq 2.0$. Unlike hypertrophic obstructive cardiomyopathy, there is no abnormal systolic movement of the mitral valve in hypertrophic cardiomyopathy without left ventricular outflow tract obstruction.

Acknowledgment

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References


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