Hypertrophic obstructive cardiomyopathy

Assessment by echocardiographic and Doppler ultrasound techniques

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Hypertrophic obstructive cardiomyopathy is a disease of the myocardium that can be assessed by echocardiographic and transcutaneous Doppler ultrasound techniques. Four patients are presented with various patterns of the disease, and the frequently familial incidence is illustrated. The importance of ultrasonic evidence for asymmetric septal hypertrophy in all stages is emphasized and evidence of reduced septal contractility demonstrated. Abnormalities of mitral valve motion, slow diastolic closure rate and systolic anterior movement of the anterior leaflet, are shown in the obstructive form of the disease. Also partial mid-systolic aortic valve closure and aortic cusp flutter are shown with outflow obstruction. The outflow tract gradient can be calculated from mitral valve to septum systolic distances. Transcutaneous Doppler ultrasound shows a normal aortic velocity pattern in nonobstructive disease while consistent abnormalities are present with severe obstruction. Isoproterenol can be used to alter the normal velocity pattern associated with a minimal resting gradient to an abnormal pattern indicating the development of significant obstruction.

Hypertrophic obstructive cardiomyopathy (HOCM) is a disease characterized by hypertrophy of the interventricular septum with variable left ventricular outflow obstruction (Wigle, Heimbecker, and Gunton, 1962; Cohen et al., 1964; Braunwald et al., 1964). The anatomical features and clinical expression of the disease vary widely from totally asymptomatic septal hypertrophy without left ventricular obstruction to very high outflow gradients and, finally, to poorly compliant, poorly contractile ventricles without outflow obstruction. Symptomatic patients may experience chest pain, arrhythmias, or heart failure, and sudden death has been observed in all varieties. Since HOCM is a relatively common form of cardiomyopathy it is important to identify this disorder in both symptomatic and asymptomatic patients. The recent advent of ultrasound techniques has provided a safe, inexpensive method for diagnosing and assessing HOCM.

There are a number of pathognomonic findings not always evident on angiography that make echocardiography and Doppler velocity techniques valuable adjuncts to catheterization. Four representative cases, from those we have studied, have been chosen to illustrate this.

Methods

Echocardiography was performed using a Unirad Model 100 ultrasonoscope and 2.5 MHz transducer. The echocardiograms were recorded on Polaroid film and on a Honeywell No. 1856 strip chart recorder. Patients were not receiving any medication when studied; they lay in the supine position, with the transducer applied to the fourth left intercostal space.

Transcutaneous directional Doppler velocity recordings were made using the technique described by Light (1969) with a 2.2 MHz beam of ultrasound focused at a depth of 8 cm. The transducer was positioned in the suprasternal notch and aimed approximately towards the left scapula crossing the descending aortic arch until maximal forward flow was seen with the least possible level of background noise. Simultaneous electrocardiogram, phonocardiogram, and carotid pulse tracings were recorded with the velocity trace, using a Litton recorder.

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Case 1

A 10-year-old boy was seen during the assessment of all first degree relatives of another patient with proven hypertrophic cardiomyopathy. This patient was asymptomatic and normally active. Physical examination revealed a healthy looking boy with a normal blood pressure and pulse rate and no abnormal heart sounds or murmurs. The chest x-ray and electrocardiogram were normal.

The echocardiogram showed an interventricular septum 12 mm thick and a posterior wall thickness of 8 mm with a septum to posterior wall ratio of 1.5:1 (Fig. 1). Septal thickness in systole was 28 per cent greater than in diastole (normal 30 per cent: Rosen et al., 1974). The left ventricular cavity was normal with a calculated end-systolic volume of 10 ml, a cardiac index of 2.8 l/min per m², and a mildly increased ejection fraction of 83 per cent. The mitral valve movements were normal, with a diastolic closure rate (DCR) of 83 mm/s and no evidence of systolic reopening. The aortic valve motion was normal. The transcutaneous Doppler aortic velocity tracing was normal with a uniform parabolic contour during systole (Fig. 2).

Since the septum to posterior wall ratio exceeded the upper limit of normal of 1.3:1 (Clark, Henry, and Epstein, 1973), it was concluded that the patient had asymmetric septal hypertrophy, probably an early stage of HOCM.

Case 2

A 15-year-old boy was seen for assessment of electrocardiographic abnormalities. His mother had been shown 2 years previously by cardiac catheterization to have HOCM and her two sons had been subsequently found to have abnormal electrocardiograms.

This patient was asymptomatic. Physical examination showed a muscular healthy looking boy with a pulse rate of 64/min and a blood pressure of 95/65 mmHg (12.6/8.6 kPa). The cardiac apex was normally placed and a soft fourth heart sound was audible but not palpable. A grade 2/4 mid-systolic ejection murmur was audible along the left sternal border and at the apex.

The chest x-ray was normal and the electrocardiogram showed right axis deviation and deep narrow Q waves from V2 to V6.

The cardiac catheter study showed a resting left ventricular pressure of 130/5 mm Hg (17.3/0.7 kPa) and outflow tract pressure of 112/9 mm Hg (14.9/1.2 kPa) with no aortic valve gradient. After 7.5 μg isoprenaline had been injected intravenously, a gradient of 70 mmHg (9.3 kPa) was produced across the left ventricular outflow tract. Narrowing of the outflow tract could be seen on the left ventriculogram.
cent. The mitral diastolic closure rate was reduced at 28 mm/s and systolic reopening of the mitral valve was visible, but the leaflet remained distant from the septum (Fig. 3). The aortic valve movement was normal. The calculated outflow gradient by the method of Henry et al. (1973b) was 12 mmHg (1.6 kPa). The transmural systolic anterior velocity tracing was consistently normal at rest, but after 7.5 μg isoprenaline intravenously a striking delay in the rise to peak velocity appeared indicating early systolic outflow obstruction (Fig. 4).

Similar angiographic and echocardiographic findings were present in this patient's brother.

**Case 3**

A 49-year-old woman was seen for assessment of progressive exertional dyspnoea and a cardiac murmur that had been present for 7 years. The patient was unable to walk on level ground for more than 100 to 200 yards because of dyspnoea, but had not experienced chest pain. She also complained of nocturnal ankle oedema and brief episodes of sudden rapid palpitation.

Physical examination revealed a healthy appearing middle-aged woman, with a pulse rate of 60/min and a blood pressure of 130/70 mmHg (17.3/9.3 kPa). The chest was clear to auscultation and there was no jugular venous distension or ankle oedema. The carotid and brachial pulses showed a brisk rise. The apex beat was in the fifth interspace in the anterior axillary line and was accompanied by a left ventricular heave. A palpable systolic thrill was present over the lower left sternal border. The first and second heart sounds were soft and no third or fourth sounds were heard. A harsh grade 3/4 systolic murmur was present at the apex, lower left sternal border, and at the base, but did not radiate into the carotid arteries. Chest x-ray showed minimal cardiac enlargement and clear lung fields. The echocardiogram showed normal sinus rhythm, with left ventricular hypertrophy but no abnormal Q waves.

On cardiac catheterization an apical left ventricular pressure of 160/0 mmHg (21.3/0 kPa) and outflow tract pressure of 100/20 mmHg (13.3/2.7 kPa) was recorded, resulting in a 60 mmHg (8.0 kPa) systolic gradient within the chamber at rest.

Echocardiography done subsequently showed a thickened interventricular septum at 28 mm and a thickened posterior wall at 17 mm (normal <14 mm) with a septum to posterior wall ratio of 1.6:1. The septum was 4 per cent thicker in systole than in diastole. The left ventricular cavity was small with an estimated end-systolic volume of 8 ml; the cardiac output was 3.1 l/min, and the ejection fraction 89 per cent. The mitral valve diastolic closure rate was reduced at 26 mm/s and systolic opening of the mitral valve was visible, but the leaflet remained distant from the septum (Fig. 3). The aortic valve movement was normal. The calculated outflow gradient by the method of Henry et al. (1973b) was 12 mmHg (1.6 kPa). The transmural systolic anterior velocity tracing was consistently normal at rest, but after 7.5 μg isoprenaline intravenously a striking delay in the rise to peak velocity appeared indicating early systolic outflow obstruction (Fig. 4).

Similar angiographic and echocardiographic findings were present in this patient's brother.
Doppler aortic velocity tracing showed early rapid flow in systole with sudden attenuation at mid-systole and subsequent slower late systolic flow (Fig. 7).

**Case 4**

A 41-year-old woman had presented to her family physician with unexplained congestive heart failure. She was successfully managed with digitalis and diuretics and, when referred for further evaluation, was still experiencing dyspnoea on moderate exertion.

On physical examination there was prominent double apical impulse associated with a loud fourth heart sound. No cardiac murmurs were present and the rhythm was regular. The electrocardiogram showed left ventricular and left atrial hypertrophy and the chest x-ray was unremarkable.

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**FIG. 5** Mitral valve echocardiogram obtained in Case 3 with accompanying line drawing. The thickened septum, reduced diastolic closure rate, and large systolic anterior mitral valve movement is seen. Abbreviations as in Fig. 3.

**FIG. 6** Aortic valve echocardiogram in Case 3. Mid-systolic aortic valve closure and vibration of the valve can be seen.

**FIG. 7** Transcutaneous Doppler aortic velocity trace with electrocardiogram, phonocardiogram, and carotid pulse from Case 3. The velocity trace is bi-phasic, with maximal flow in early systole, sudden decrease, and slower late systolic velocity.

On cardiac catheterization the wedge pressure showed large 'a' waves, up to 25 mmHg (3.3 kPa), and a slow 'y' descent indicating prolonged ventricular filling. The left ventricular pressure was 85/22 mmHg (11.3/2.9 kPa) and aortic pressure 85/50 mmHg (11.3/6.7 kPa). The end-diastolic pressure rose to 32 mmHg (4.3 kPa) after ventriculography and the cardiac index was 1.8 l/min per m². No outflow tract gradient could be provoked by isoprenaline infusion. The left ventricle appeared heavily trabeculated with the elongated shape of HOCM but was poorly contractile.

Echocardiography done 18 months later showed a noncontractile septum 15 mm thick and a poorly contracting posterior wall 7 mm thick giving a septum to posterior wall ratio of 2.3:1 (Fig. 8). There was no perceptible thickening of the septum from diastole to systole. The left ventricle was not grossly dilated, with a diastolic minor axis diameter of 50 mm, but the ejection fraction was reduced to 20 per cent. The right ventricle was dilated. The rhythm was atrial fibrillation and the mitral valve movements were otherwise normal with a diastolic closure rate of 104 mm/s and no evidence of mid-systolic reopening. The aortic valve motion was not recorded and the Doppler aortic velocity tracing was not done.

This patient had an advanced stage of HOCM with a rigid ventricle and no remaining outflow obstruction. She was the mother of Case 2.

**Discussion**

The principal abnormality of the echocardiography in HOCM is a thickened interventricular septum...
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The interventricular septum may be poorly contractile (Rosen et al., 1974) with decreased systolic thickening and excursion. In all four cases this thickening fell below the lower limit of normal (30%) though Case 1, with asymmetric septal hypertrophy alone, was near that limit (Table).

A small ventricle with a raised ejection fraction is characteristic of the obstructive form of HOCM, as seen in Cases 2 and 3, while the loss of obstruction in the latter stages of the disease process may result in a 'normal' sized chamber with a much reduced ejection fraction (Case 4).

Mitral valve movement in the obstructive stage of HOCM is distinctly abnormal and, within limits, characteristic. The diastolic closure rate of the anterior leaflet is usually much reduced while in the 'early' stage, e.g. septal hypertrophy alone as seen in first degree relatives of HOCM patients, the diastolic closure rate is frequently normal. Also in the late stage when the outflow obstruction has disappeared the diastolic closure rate may return to normal. The explanation for these findings probably lies in the dynamics of mitral valve motion. In model studies the diastolic closure rate has been shown to depend on vortex formation in the ventricular outflow tract closing the valve in mid-diastole (Bellhouse, Bellhouse, and Gunning, 1973).

In HOCM the proximity of the greatly hypertrophied septum may result in poor vortex formation in the abnormally shaped small cavity, and thus provide little force to reclose the anterior mitral leaflet in diastole. When the outflow tract size is adequate, as in Cases 1 and 4, the diastolic closure rate is normal.

### TABLE Summary of echocardiographic and Doppler findings

<table>
<thead>
<tr>
<th></th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Septal thickness</td>
<td>12 mm</td>
<td>32 mm</td>
<td>28 mm</td>
<td>16 mm</td>
</tr>
<tr>
<td>(normal 8 to 13 mm)</td>
<td>8 mm</td>
<td>10 mm</td>
<td>17 mm</td>
<td>7 mm</td>
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<tr>
<td>Posterior wall thickness</td>
<td>1.5:1</td>
<td>3.2:1</td>
<td>1.6:1</td>
<td>2.3:1</td>
</tr>
<tr>
<td>(normal &lt; 1.4:1)</td>
<td>28%</td>
<td>5%</td>
<td>4%</td>
<td>0%</td>
</tr>
<tr>
<td>Ratio septum to posterior wall thickness</td>
<td>Normal</td>
<td>Small</td>
<td>Small</td>
<td>Normal</td>
</tr>
<tr>
<td>Systolic septal thickening</td>
<td>2.8 l/min</td>
<td>2.4 l/min</td>
<td>3.1 l/min</td>
<td>1.8 l/min</td>
</tr>
<tr>
<td>(normal &gt; 30%)</td>
<td>per m²</td>
<td>per m²</td>
<td>per m²</td>
<td>per m²</td>
</tr>
<tr>
<td>Ventricular cavity size</td>
<td>83%</td>
<td>87%</td>
<td>89%</td>
<td>20%</td>
</tr>
<tr>
<td>Cardiac output or index</td>
<td>83 mm/s</td>
<td>28 mm/s</td>
<td>26 mm/s</td>
<td>104 mm/s</td>
</tr>
<tr>
<td>Ejection fraction (normal 60 to 75%)</td>
<td>Absent</td>
<td>Present</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Mitral diastolic closure rate (normal &gt; 40 mm/sec)</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Mitral systolic anterior movement</td>
<td>Normal</td>
<td>Abnormal with isoprenaline</td>
<td>Abnormal</td>
<td>—</td>
</tr>
<tr>
<td>Aortic valve motion</td>
<td>Normal</td>
<td>Abnormal</td>
<td>Abnormal</td>
<td>—</td>
</tr>
</tbody>
</table>

(Feigenbaum, 1972). The characteristic feature is a septum to posterior wall ratio greater than 1.3 to 1. This occurs in most symptomatic and asymptomatic patients with HOCM and has been found in a high proportion of first degree relatives of afflicted patients (Clark et al., 1973). With a persistently high outflow gradient the myocardium may become more symmetrically hypertrophied to resemble the hypertrophy of fixed outflow obstruction but the ratio remains high as in Case 3 (Henry, Clark, and Epstein, 1973).

In addition to the abnormal hypertrophy the site from which retrograde aortic flow occurs has been described as follows.

**FIG. 8 Echocardiogram obtained in Case 4 with line drawing. The thickened and poorly contractile septum as well as poorly contractile posterior wall are shown. The mitral diastolic closure rate is normal and no systolic anterior movement can be seen. RV indicates right ventricle. Other abbreviations as in Fig. 3.**
In systole the anterior mitral valve leaflet is normally seen as part of the systolic closure line lying well posterior to the septum throughout, then moving rapidly anterior in diastole. In the non-obstructive form of HOCM no abnormality of the systolic closure line may be noted and the most important finding on the echogram will be the asymmetric septal hypertrophy (Henry et al., 1974) as seen in Cases 1 and 4. In the obstructive form the anterior leaflet moves sharply towards the septum in early systole. Only a small portion of the leaflet is actually involved in this abnormal movement and careful scanning with the transducer is needed to demonstrate it. Provocative manoeuvres such as amyl nitrite inhalation, Valsalva’s manoeuvre, and isoprenaline infusion are valuable in bringing out this leaflet motion in suspected cases where it has not been visualized (King et al., 1973). This movement is distinct from the bowing forward of the mitral valve ring and closure line that is also so often apparent in HOCM (Shah et al., 1971). The second case shows that systolic reopening does not depend upon the presence of an intraventricular gradient and tends to support the theory that altered ventricular geometry rather than a Venturi effect is the mechanism of systolic reopening.

Care must be exercised to avoid confusing the systolic anterior mitral movement of HOCM with the systolic anterior movement recorded in some patients with mitral valve prolapse syndrome. In HOCM the mitral reopening clearly returns to the systolic closure line in late systole (Fig. 5) while in prolapse we have occasionally seen an anterior movement starting in early systole, which does not return to the baseline and is continuous with the diastolic opening of the mitral valve (Fig. 9). We have recorded this echocardiographic pattern in 6 of 43 patients with angiographically proven mitral prolapse. Extensive prolapse with anterior as well as posterior leaflet involvement appeared to correlate with this finding but the exact explanation for the pattern remains unclear. Confusion with the systolic anterior movement of HOCM may result if poor visualization of the late systolic portion of this systolic anterior movement is obtained.

Measurement of the mitral valve – septal distance can be used as a means of calculating left ventricular obstruction (Henry et al., 1973b), whether or not it is the close apposition of these two structures that causes the obstruction. An obstruction index is calculated by dividing the duration of systolic reopening of the average mitral – septal distance. The gradient is then expressed as \((1.8 \times \text{obstruction index}) - 35\), which corresponds well with the catheter findings in the original series, and in our two patients with obstruction.

With significant disease the aortic valve closes in mid-systole, presumably in response to the sudden attenuation of blood flow as obstruction develops, but reopens late in systole. The valve was seen to vibrate throughout systole in the patient with severe obstruction (Fig. 6), reflecting turbulent flow across the valve which may be caused by subvalvular stenosis of any type (Popp et al., 1974). The systolic closure and reopening, however, are specific for the variable obstruction that occurs only in HOCM.

The abnormal pattern of aortic blood velocity seen in HOCM was first described by Hernandez, Greenfield, and McCall (1964) using the intraventricular pressure gradient technique and was characterized by a rapid early systolic velocity followed by an abrupt decrease in flow velocity at mid-systole with a slower late systolic velocity. Blood velocity can be measured by transcutaneous Doppler ultrasound techniques either from the carotid artery (Joyner, Harrison, and Grubner, 1971) or, as we have done, from the descending aortic arch. Case 1 without evidence of obstruction showed a normal velocity trace obtained from the descending aortic arch by transcutaneous Doppler ultrasound (Fig. 2). The velocity pattern was symmetrical throughout systole with a maximum peak velocity occurring at mid-systole. Case 2 with minimal resting obstruction but a 70 mmHg (9.3 kPa) outflow tract gradient after isoprenaline showed a pronounced alteration in the aortic velocity pattern after infusion of that drug (Fig. 4). The development of obstruction in

FIG. 9 Unusual mitral echogram from patient with extensive mitral valve prolapse. A systolic anterior movement is seen that does not return to the systolic closure line, unlike HOCM, but is continuous with anterior leaflet opening. RV indicates right ventricle. Other abbreviations as in Fig. 3.
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References


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FIG. 10 Aortic valve motion from Case 3 transcribed on rectangular co-ordinates with aortic velocity trace and electrocardiogram. The aortic valve movement parallels the aortic blood velocity.

ECG

RIGHT CORONARY CUSP

AORTIC VALVE

NON-CORONARY CUSP

AORTIC VELOCITY

This case produced a biphasic velocity pattern with the peak velocity occurring in late systole. Here the isoprenaline apparently produced the obstruction early in systole such that there was an initial rise in velocity, then a delay or fall, and a final rise to peak velocity in late systole. Case 3, with resting obstruction, showed the aortic velocity pattern predicted by Hernandez et al. (1964), with a sharp early peak followed by a rapid decrease in velocity in mid-systole then a second slower peak extending to the end of systole (Fig. 7). As shown in Fig. 10, the velocity trace corresponded in timing to the movement of the aortic valve. The aortic valve motion recorded on the echocardiogram was redrawn to remove the to-and-fro movement and then replotted against the aortic velocity trace using the electrocardiogram as a timing reference.

These 4 cases represent examples of the spectrum of the HOCM and the usefulness of ultrasound in its evaluation. Characteristic features of the disease are well demonstrated by echocardiography and, in the obstructive forms of the disease, Doppler ultrasound can demonstrate the abnormal aortic velocity profile.

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