Echocardiographic Criteria in the Diagnosis of Idiopathic Hypertrophic Subaortic Stenosis

By Ronald M. Rossen, M.D., Daniel J. Goodman, M.D., Robert E. Ingham, M.D., and Richard L. Popp, M.D.

SUMMARY

Echocardiography has proven to be a useful technique in the diagnosis and assessment of therapy in idiopathic hypertrophic subaortic stenosis (IHSS). Asymmetric septal hypertrophy has been described as the pathognomonic anatomic marker of the disease. A characteristic systolic anterior motion of the anterior mitral valve leaflet has been detected in the presence of hemodynamically significant subaortic left ventricular outflow obstruction. An echocardiographic quantification of the outflow gradient (the obstruction index) has been derived previously.

Four patients were studied by ultrasound at the time of cardiac catheterization. All four demonstrated systolic anterior motion of the anterior mitral leaflet in the absence of a resting gradient. In three of the four, the calculated obstruction index predicted hemodynamically significant resting gradients. All four patients were shown to have labile gradients with provocative maneuvers. A fifth patient with abnormal systolic anterior motion demonstrated a close correlation between the obstruction index and resting gradient; however, symmetric hypertrophy of the septum and left ventricular posterior wall was detected by ultrasound. Therefore, the abnormal mitral valve pattern may be seen in the absence of a resting gradient and symmetric left ventricular hypertrophy may exist in the presence of IHSS.

Additional Indexing Words:
Echocardiography
Obstruction index
Asymmetric septal hypertrophy
Systolic anterior motion

IDIOPATHIC HYPERTROPHIC SUBAORTIC STENOSIS (IHSS) has been recognized as a distinct disease entity within the last fifteen years.\textsuperscript{1}–\textsuperscript{3} The clinical, hemodynamic, pathological and therapeutic aspects of the subvalvular obstruction have evoked considerable interest and discussion.\textsuperscript{4}–\textsuperscript{6} Recently, echocardiography has been proposed as a sensitive and specific noninvasive method for the detection and therapeutic assessment of IHSS.\textsuperscript{7}–\textsuperscript{13} A characteristic systolic anterior motion of the anterior mitral valve leaflet has been described in the presence of hemodynamically significant subaortic obstruction.\textsuperscript{14}–\textsuperscript{16} Asymmetric septal hypertrophy has been suggested as the pathognomonic anatomic abnormality and the subvalvular gradient has been quantified by ultrasound with the derivation of an obstruction index.\textsuperscript{18}–\textsuperscript{20}

In the course of a prospective hemodynamic and echographic study, four patients with asymmetric septal hypertrophy had echograms suggestive of significant gradients at a time when simultaneously determined left ventricular outflow tract pressure gradients were zero. An additional patient with systolic anterior motion of the mitral valve and a resting gradient was noted to have symmetric (concentric) hypertrophy of the left ventricle.

It is the purpose of this paper to emphasize three points: (1) The characteristic systolic anterior motion of the mitral valve may be present without a gradient. (2) The obstruction index and calculated gradient may not correlate with the actual gradient as determined by catheterization. (3) Concentric symmetric ventricular hypertrophy may be part of the IHSS spectrum.

Methods

Five patients, four males and one female, aged 14, 16, 18, 55, and 65, underwent echocardiographic examination during the course of cardiac catheterization. The diagnosis was established in each case by transseptal catheterization of the left ventricle and simultaneous recording of the central aortic pressure, with the standard provocative maneuvers and left ventricular angiography.\textsuperscript{5}–\textsuperscript{14} Simultaneous M-mode sector scan echograms and left ventricular and central aortic pressures were recorded at rest. Pressures were measured with either the Micron MP-15 or Statham P23Db transducers.

All cardiac echograms were performed with a Smith Kline Ekoline 20 instrument using a 2.25 MHz transducer of 0.50 inch diameter with a repetition rate of 1000 pulses/sec. The echograms were recorded either by a Polaroid photograph...
from the oscilloscope or by means of a Honeywell 1856 strip
chart recorder. The technique of M-mode sector scan ul-
trasound examination has been previously described.

The patients were studied in the supine position with the
transducer in the fourth or fifth intercostal space along the
left sternal border. The transducer was directed so that both
the anterior and posterior leaflets of the mitral valve were
recorded. Care was taken to locate the free edge of the
mitral leaflets, observing the full M-mode sector scan. The
ultrasonoscope was adjusted to record all the echoes from
the interventricular septum and left ventricular posterior
wall. The interventricular septum and left ventricular
posterior wall dimensions were measured below the level of
the edge of the mitral leaflets and a ratio was calculated. The
initial diastolic anterior mitral valve slope was determined
for each patient. An obstruction index was calculated for
representative beats from the echogram by dividing the
duration of left ventricular outflow narrowing by the mean
distance between the anterior mitral leaflet and septum during
systole as described by Henry et al. The predicted gradient
was calculated from the regression equation derived by
Henry et al.: Predicted gradient = (1.8 × obstruction index) − 35. Peak subvalvular pressure
gradients were determined from the left ventricular and
central aortic pressure tracings for the beats simultaneously
recorded on the echogram.

Results

Hemodynamic Data

The hemodynamic data obtained at cardiac
catheterization are summarized in table 1. The mean
resting cardiac index was 2.9 L/min/m². Four patients
had no left ventricular-aortic pressure gradient at rest.
However, with appropriate provocative maneuvers,
significant gradients ranging from 10 to 180 mm Hg
were produced in each patient. The most profound
gradients followed the administration of isopro-
terenol.

Echocardiographic Data

A summary of the echographic data is presented in
table 1. The mean end-diastolic septal thickness was
24.6 mm with a range of 20–28 mm, and the mean left
ventricular posterior wall thickness was 14.0 mm with
a range of 10–20 mm. The septum-left ventricular
posterior wall ratio averaged 1.9 in all patients, and
2.1 exclusive of the patient with symmetric hyper-
trophy. The initial diastolic slope of the mitral valve
ranged from 35 to 66 mm/sec with a mean of 43.4
mm/sec. These values are reduced from the normal
value in this laboratory of > 70 mm/sec.

In the four patients without resting subaortic
pressure gradients, the calculated obstruction index of
the simultaneously recorded beats ranged from 20 to
68. Predicted gradients using the regression equation
ranged from 2 to 87 mm Hg. Due to technical factors,
simultaneous echocardiograms and pressures could
not be obtained in all patients during all provocation
maneuvers. In the one patient with a resting subaortic

<table>
<thead>
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<th>Patient</th>
<th>Age</th>
<th>Ultrasound</th>
<th>Peak gradient (mm Hg)</th>
<th>Atrial</th>
<th>Septal ( \text{diam} )</th>
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</tbody>
</table>

Abbreviations: CL = cardiac index at rest. Peak gradient = peak left ventricular-aortic systolic pressure gradient. MV = mitral valve.

Predicted resting gradient derived from regression equation.

*Actual peak-systolic gradient at rest for beats during the echographic recording.

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gradient, there was close agreement between the predicted and directly measured values.

Figure 1 illustrates a representative echocardiogram with simultaneously recorded left ventricular and aortic pressures from patient S.L. Asymmetric septal hypertrophy and a reduced initial diastolic mitral valve slope are demonstrated. In addition, the anterior mitral leaflet moves anteriorly in systole, toward the septum, giving a calculated obstruction index of 40, at a time when there is no gradient recorded.

Figure 2 is the echogram from patient A.A. with a resting subvalvular gradient. The anterior mitral leaflet makes an early, abrupt motion toward the septum and makes contact with it in mid-systole, giving an obstruction index of 40. There is marked hypertrophy of both the septum and left ventricular posterior wall with a ratio of 1.0.

Discussion

The firm clinical diagnosis of IHSS often has proven difficult, requiring cardiac catheterization with provocative maneuvers to reveal the subvalvular obstruction.2, 6, 7, 22 Echocardiography has been shown to be a highly specific and sensitive noninvasive method for the diagnosis of the disorder.14-19 Moreyra first commented upon the use of ultrasound in IHSS, noting contact between the septum and anterior mitral leaflet in diastole.23 This has subsequently proven to be a non-specific finding. A characteristic systolic anterior motion of the mitral valve during systole was observed by Shah and associates and later confirmed by other groups, although this pattern may be absent in patients with latent and labile gradients.14-17 It has been suggested that the abnormal motion is the result of the hypertrophied septum producing geometric distortion of the papillary muscles and is an important factor contributing to the left ventricular outflow tract obstruction.15 Based upon the assumption that contact between the anterior mitral leaflet and hypertrophied septum produces the pressure gradient, and utilizing simultaneous echograms and catheterization data, an obstruction index and regression equation have been derived, allowing noninvasive estimation of the gradient.18 If such an index were reliable, it could provide a convenient objective method for the assessment of therapy.

During simultaneous echographic and hemodynamic evaluation, four patients were noted to have abnormal systolic anterior motion of the mitral valve at a time when no left ventricular outflow tract gradient was recorded. Our group and others have stated that the mitral valve motion abnormalities are present only when a gradient exists.14, 15 The results in our present series of patients seem to contradict our previous statements.15 At a time when no gradient could be measured, systolic anterior motion of the anterior mitral leaflet was clearly recorded. Henry has likewise found patients in whom the leaflet abnormality was present in the absence of a pressure gradient.19 We have also observed disappearance of the systolic mitral valve movement with propranolol therapy in some patients, although this has not been a common finding.17, 18 Successful ventriculomyectomy also reportedly produces disappearance of the mitral motion disorder.17

In a prospective attempt to evaluate the obstruction index, we have applied the regression equation of Henry to our patients. In only two of our five patients did the predicted gradient approximate the directly measured value. The three other patients had predicted gradients ranging from 22 to 87 mm Hg when no gradient could be recorded. The obstruction index and regression data may have to be derived from each patient to allow for quantitative serial study. This is not surprising, since the three-dimensional shape of the orifice formed by the anterior mitral leaflet and septum may not be the same in each patient. Obviously an echographic representation of one portion of a cross-sectional level
of the outflow tract may be misleading in some patients.

Asymmetric septal hypertrophy has been offered as the pathognomonic anatomic abnormality in IHSS, irrespective of the presence of a gradient or the mitral valve motion abnormality. A 16-year-old girl with a family history of the disease demonstrated the characteristic mitral valve motion at a time when a resting gradient was recorded. The interesting feature of her echogram was the presence of symmetric, concentric hypertrophy of the left ventricle with a septal-posterior wall ratio of 1.0. Both the interventricular septum and left ventricular posterior wall measured 20 mm in thickness. This patient is an exception to the reported findings of a septal-free wall ratio exceeding 1.3 in 15 patients with catheterization-documented IHSS. This patient has undergone catheterization on three occasions during her life, and no other type of outflow obstruction or hypertension has been found. It is interesting to postulate that asymmetric septal hypertrophy which may have been present from birth produced subvalvular obstruction, and under this stress, hypertrophy of the free wall subsequently developed.

It would be expected that echographic findings would reflect this anatomic spectrum. Asymmetric septal hypertrophy has been recognized by echocardiography with a striking prevalence in family members of those with IHSS. Appreciation of systolic anterior motion of the mitral valve has helped indicate the site of obstruction, although we have demonstrated that this motion may be present without a gradient. It is probable that the duration, degree, and area of contact between the septum and anterior mitral leaflet is related to the production of the gradient. The pattern of symmetric concentric hypertrophy in the presence of subaortic obstruction was seen in one of our patients. An echographic pattern of concentric hypertrophy, but without the abnormal mitral valve motion, has been seen in patients with the clinical and hemodynamic picture of pure hypertrophic nonobstructive cardiomyopathy in our laboratory. It is possible that such cases represent a later stage in the course of IHSS. When symmetric hypertrophy develops, subaortic obstruction may be reduced, leaving a picture of nonobstructive cardiomyopathy. Exploration of this sequence will require demonstrating progression from a typical echographic picture of asymmetric septal hypertrophy without obstruction, to asymmetric septal hyper-

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**Figure 2**

Echogram of patient A.A. who had a resting gradient and symmetric left ventricular hypertrophy. Note the marked septal and left ventricular posterior wall thickening. EKG = electrocardiogram; AW = right ventricular anterior wall; IVS = interventricular septum; PW = left ventricular posterior wall; MV = mitral valve anterior leaflet.
trophic with obstruction, and subsequently to a hypertrophic nonobstructive pattern. Echocardiography appears to be a good method for such long-term evaluation.

References
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