Echocardiography in the Diagnosis of Idiopathic Hypertrophic Subaortic Stenosis Co-existing With Aortic Valve Disease

By Navin C. Nanda, M.D., Raymond Gramiak, M.D., Pravin M. Shah, M.D., Scott Stewart, M.D., and James A. DeWeese, M.D.

SUMMARY
Echocardiographic studies demonstrated the presence of co-existing idiopathic hypertrophic subaortic stenosis (IHSS) in six patients with aortic valve disease (four calcific aortic stenosis, two pure aortic incompetence). The characteristics of IHSS were a narrow left ventricular outflow tract, a systolic anterior movement of the mitral valve (SAM), and asymmetric ventricular septal hypertrophy. Large SAMs were observed in two patients with pure aortic incompetence and one with aortic stenosis. Relatively small, inconstant, and often incomplete SAMs were noted in the remaining three patients with aortic stenosis. In contrast to isolated IHSS, the small SAMs observed in this group did not become prominent with the Valsalva maneuver or amyl nitrite inhalation. These features may be related to the afterload provided by the fixed, distal stenosis. Echocardiographic evidence of aortic valve disease was present in all patients. Clinically, co-existence of IHSS was not suspected in five patients. Associated IHSS was established using provocative measures during cardiac catheterization in three cases, while in the remainder it was substantiated at surgery. Three of four patients who underwent myotomy/myectomy concomitant with aortic valve replacement survived and postoperative echocardiographic studies revealed complete absence of SAMs in two of them. Echocardiography appears to be useful in the diagnosis of associated IHSS in the presence of aortic valve disease.

Additional Indexing Words:
Ultrasound
Aortic incompetence
Aortic valve stenosis
Myotomy
Myectomy
SAM

The co-existence of idiopathic hypertrophic subaortic stenosis (IHSS) and aortic valve disease represents a diagnostic challenge since the presence of aortic valve disease tends to mask the clinical and hemodynamic features of IHSS, thus making precise diagnosis difficult. This co-existence has been recognized preoperatively during cardiac catheterization, but in some instances has not been diagnosed until surgery or autopsy. The number of reported cases suggests this to be a rare combination, but some investigators have estimated that the frequency of co-existence of these two lesions may be as high as 10% of patients with aortic valve disease. Since the presence of IHSS may require alterations in the clinical therapy or surgical management of the patient, recognition of this complex assumes considerable clinical significance. Echocardiography has developed into a very accurate method for the detection of isolated IHSS and has recently been shown to be of value in the recognition of IHSS co-existing with left heart obstructive lesions in children. The purpose of this report is to describe our echocardiographic experience in detecting IHSS associated with aortic valve disease in adults and to stress the value of echocardiography in the noninvasive diagnosis of this complex condition.

Material and Methods
Six patients with evidence of aortic valve disease form the basis of this report. Patients were equally divided between the sexes. Their ages ranged from 44 to 69 years. In five of six patients the presence of co-existing IHSS was not suspected clinically. Their symptoms, which included angina, syncope on exertion, and manifestations of heart failure, and physical findings were consistent with a diagnosis of aortic valve disease alone. Clinical features suggestive of IHSS and aortic incompetence were present in one patient. A grade 3/6 systolic murmur, audible at the apex and left sternal border, which increased in intensity with the Valsalva maneuver, suggested the presence of IHSS in this patient. Patient clinical data are summarized in Table 1. In all patients, right and left heart cardiac catheterization (retrograde in cases 4 and 5, transseptal in the remainder) as well as left ventricular angiography were
performed. Pressures were obtained in the standard manner.

Echocardiographic studies were carried out in all patients using a commercially available echograph (Picker) and a 2 MHz transducer. Continuous records were made on 35 mm film by means of a Fairchild oscilloscope camera and a dual beam oscilloscope operating as a slave. Mitral valve echograms were obtained by placing the transducer in the 3rd or 4th left intercostal space and directing the beam posteriorly and slightly medially. Aortic root echoes were obtained by medial and cephalic rotation of the transducer from the mitral valve position. In three patients, echoes from the ventricular septum and the posterior free wall of the left ventricle were obtained by a method previously described. Mitral valve echograms were studied for evidence indicative of IHSS. Simple provocative tests using amyl nitrite inhalation and/or the Valsalva maneuver were performed when the SAMs were small and inconstant. The left ventricular outflow tract was measured in mm as the minimum space between the closed position of the mitral valve and the left side of the ventricular septum at the onset of systole (fig. 1). Measurements of the width of the ventricular septum and the posterior free wall of the left ventricle were obtained in three cases. In addition, mitral valve echograms were analyzed for the presence of diastolic flutter indicative of aortic regurgitation. Aortic root echoes were studied for the character of cusp motion pattern and for evidence of valve thickening and/or calcification. Echocardiographic studies were repeated in three patients following surgery.

## Echocardiographic Findings

**Table 2**

<table>
<thead>
<tr>
<th>Case no.</th>
<th>LVO (mm)</th>
<th>SAM</th>
<th>VS (mm)</th>
<th>VS/PW</th>
<th>AV</th>
<th>MVF</th>
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<tbody>
<tr>
<td>1</td>
<td>19</td>
<td>Large</td>
<td>23</td>
<td>—</td>
<td>Calcified</td>
<td>Present</td>
</tr>
<tr>
<td>2</td>
<td>18</td>
<td>Small</td>
<td>26</td>
<td>1.7</td>
<td>Calcified</td>
<td>Absent</td>
</tr>
<tr>
<td>3</td>
<td>16</td>
<td>Small</td>
<td>22</td>
<td>—</td>
<td>Calcified</td>
<td>Absent</td>
</tr>
<tr>
<td>4</td>
<td>15</td>
<td>Small</td>
<td>28</td>
<td>1.6</td>
<td>Calcified</td>
<td>Present</td>
</tr>
<tr>
<td>5</td>
<td>14</td>
<td>Large</td>
<td>28</td>
<td>2.0</td>
<td>Thickened with restricted mobility</td>
<td>Present</td>
</tr>
<tr>
<td>6</td>
<td>14</td>
<td>Large</td>
<td>26</td>
<td>—</td>
<td>Thickened with restricted mobility</td>
<td>Absent</td>
</tr>
</tbody>
</table>

Abbreviations: LVO = left ventricular outflow width; SAM = systolic anterior movement of the mitral valve; VS = width of the ventricular septum; PW = thickness of the left ventricular posterior wall (mm); AV = aortic valve; MVF = diastolic mitral valve flutter.

**Figure 1**

Left ventricular outflow tract measurement technique. The space between the ventricular septum (VS) and the closed position of the mitral valve (MV) forms the left ventricular outflow tract. Its width was measured in mm at the onset of systole (shown by arrows). PHONO = phonocardiogram; ECG = electrocardiogram.
Aortic stenosis and regurgitation co-existing with functional left ventricular outflow obstruction. A prominent systolic anterior movement of the anterior mitral leaflet (arrow) extends upward toward the ventricular septum (left panel). Lower instrument sensitivity (right panel) demonstrates the diastolic flutter of the mitral leaflets characteristic of aortic regurgitation. MV = mitral valve; PHONO = phonocardiogram; ECG = electrocardiogram.

Co-existing aortic valve stenosis and IHSS. The mitral valve study (MV) shows a relatively shallow systolic anterior movement (arrow) of the mitral leaflet (left panel). The aortic valve (AV) is calcified and the systolic motion is markedly restricted (right panel). IVS = ventricular septum; PHONO = phonocardiogram; ECG = electrocardiogram.
stenosis, one aortic stenosis with mild incompetence, the SAMs took an atypical appearance. They were small in amplitude, inconstant, and partial or fragmented in continuity of presentation (fig. 3). These abnormalities were of the type described in a previous study to be suggestive, but not diagnostic, of IHSS. Also, unlike isolated IHSS, they did not become prominent with amyl nitrite inhalation or the Valsalva maneuver. A high degree of suspicion was required to recognize the associated IHSS and the recording of many cardiac cycles with variations in beam direction proved useful in the detection of these atypical SAMs. The ventricular septum was hypertrophied in every patient, measuring from 22–28 mm in thickness (normal thickness varies from 7–11 mm in adults). All patients demonstrated narrow left ventricular outflow tract dimensions (normal width in adults ranges from 20–35 mm). In three cases in which adequate echos were obtained from the posterior left ventricular wall, the ventricular septal-to-posterior free wall thickness ratio was greater than 1:3 (fig. 4). Thus, the existence of associated IHSS in these patients was suggested by a combination of narrow left ventricular outflow tracts and either typical or atypical SAMs. In two of the three patients with atypical SAMs, the diagnosis of associated IHSS was further substantiated by echocardiographic, evidence of asymmetric hypertrophy of the ventricular septum.

Echocardiographic evidence of aortic valve disease was present in all subjects. Aortic root echograms revealed multilayered complexes with markedly restricted or no visible cusp motion indicative of moderate to heavy valve calcification in four patients. Less numerous linear diastolic echoes with minimally restricted leaflet motion, suggestive of valve thickening or relatively less severe calcification, were observed in the remaining cases. Echocardiographic evidence for aortic insufficiency, as judged by the presence of a diastolic flutter on the mitral valve echograms, was observed in three patients. Clinical findings of associated mitral stenosis were supported by echocardiography in one patient. The mitral valve echogram suggested parallel movements of the two leaflets in addition to a slow diastolic slope and the absence of an ‘a’ wave. The small atypical SAM in this patient was considerably fragmented.

At cardiac catheterization, co-existing IHSS was suggested in three patients using standard provocative measures (cases 1, 5, and 6, tables 1 and 3). A significant fall in the pulse pressure of the post-extrasystolic beat was observed in two of them. Separate valvular and subvalvular gradients were not obtained. In the remaining three patients, co-existing IHSS was not demonstrated by cardiac catheterization. No provocative tests were performed. Analysis of the pressure tracings in one of these patients failed to

Table 3

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Hemodynamic Findings</th>
<th>Pressures (mm Hg)</th>
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</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>LV cavity</td>
</tr>
<tr>
<td>1</td>
<td>Control</td>
<td>250/20</td>
</tr>
<tr>
<td></td>
<td>Amyl nitrite inhalation</td>
<td>250/20</td>
</tr>
<tr>
<td></td>
<td>i.v. Angiotensin (1.5 μg/min)</td>
<td>310/25</td>
</tr>
<tr>
<td></td>
<td>Post PVC</td>
<td>360/20</td>
</tr>
<tr>
<td>2</td>
<td></td>
<td>228/13</td>
</tr>
<tr>
<td>3</td>
<td></td>
<td>208/15</td>
</tr>
<tr>
<td>4</td>
<td></td>
<td>231/6–36</td>
</tr>
<tr>
<td>5</td>
<td>Control</td>
<td>160/10</td>
</tr>
<tr>
<td></td>
<td>Post PVC</td>
<td>105/12*</td>
</tr>
<tr>
<td>6</td>
<td>Control</td>
<td>172/8</td>
</tr>
<tr>
<td></td>
<td>i.v. Angiotensin (1.5 μg/min)</td>
<td>152/10</td>
</tr>
</tbody>
</table>

Abbreviations: LV = left ventricle; SA = systemic artery; i.v. = intravenous; PVC = premature ventricular contraction.

*The catheter (retrograde) was suspected to be in the LV outflow area and this may explain the absence of any gradient.

Figure 4

Asymmetric ventricular septal hypertrophy in IHSS. Ventricular septal (VS) thickness to left ventricular posterior wall (PW) width ratio of about 1.7 indicates asymmetric septal hypertrophy.

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demonstrate the characteristic post-extrasystolic response.\textsuperscript{18} Left ventricular angiograms, performed in RAO position, showed small ventricular end-systolic chambers and these were interpreted as consistent with left ventricular hypertrophy secondary to aortic valve disease. Angiographically, the aortic valve was rigid and calcified in four patients (cases 1–4). Four patients showed evidence of aortic regurgitation. It was estimated to be moderate (grade 2/4) in case 1 and mild in cases 4, 5, and 6. No resting valvular gradient in case 5 and complete abolition of the gradient with angiotensin in case 6 indicated absence of fixed valvular stenosis in these patients. The diagnosis of mitral stenosis in case 3 was confirmed at cardiac catheterization and the calculated mitral valve area was 0.68 cm\textsuperscript{2}.

Four patients in the present study underwent surgery (table 1). All showed evidence of subvalvular muscular hypertrophy characteristic of IHSS.\textsuperscript{18} A greatly hypertrophied mass of muscle projected as a thick ridge into the left ventricular outflow tract along its septal side. The aortic valve cusps showed moderate to severe degrees of thickening, fusion, and calcification indicating the presence of significant valvular stenosis in all the operated cases. In three patients myectomy was performed in conjunction with aortic valve replacement. The fourth patient underwent myotomy and mitral commissurotomy in addition to implantation of the aortic prosthesis.

Echocardiographic studies were repeated in three patients postoperatively. In two of them (cases 1 and 2) an abnormality of the systolic segment of the mitral valve could no longer be detected either at rest or following amyl nitrite inhalation. Small SAMs which did not become prominent during provocation with amyl nitrite persisted in one patient (case 4).

**Discussion**

Echocardiography has been successfully used in the diagnosis of isolated IHSS\textsuperscript{7, 8, 16, 19} When consistently observed, a large SAM of the mitral valve, together with a narrow left ventricular outflow tract dimension, is highly specific for this condition. Small inconstant SAMs are suggestive, but not diagnostic, of IHSS. Recently echocardiographic demonstration of asymmetric hypertrophy of the ventricular septum has been shown to be of value in the recognition of this entity.\textsuperscript{10, 20}

The mitral SAM has been proposed to be the basis of outflow obstruction. A Venturi effect, due to the rapid early ejection or anatomic distortion of the hypertrophied papillary muscle, may provide the explanation for this abnormal motion.\textsuperscript{9} A large excursion resulting in apposition of the mitral valve with the ventricular septum in systole is usually associated with significant obstruction under basal conditions. Smaller movements have been observed in IHSS in the absence of a resting gradient. However, they become prominent when obstruction is elicited by appropriate provocative procedures. The behavior of the aortic valve in the presence of associated fixed obstruction at the aortic valve level has not been previously studied. The afterload provided by the aortic stenosis may, to some extent, mask the severity of the co-existing functional obstruction. This may explain the relatively small and somewhat atypical appearance of the mitral SAMs observed in three of four patients with pure or predominant aortic valve stenosis. The presence of a rigid mitral valve in the patient with associated mitral stenosis may be expected to make it incapable of sharp systolic reopening required for outflow obstruction. This might explain the atypical and markedly fragmented character of the SAM observed in this patient. In the presence of aortic stenosis, relatively small mitral SAMs may portend the co-existence of subaortic functional obstruction. Additional evidence to indicate the co-existence of IHSS was provided by the narrow left ventricular outflow tract dimensions observed in all patients in the present study. Narrowing of the outflow space results from encroachment upon this area by the hypertrophied ventricular septum. Patients with aortic stenosis without associated IHSS do not appear to have narrow left ventricular outflow tracts even when the stenosis is severe (table 4). Furthermore, asymmetric ventricular septal hypertrophy, typical of IHSS,\textsuperscript{10} was documented echocardiographically in three patients, two of whom demonstrated atypical SAMs.

Echocardiography is also of value in the assessment of aortic valve disease. Valvular thickening and/or calcification can be reliably evaluated\textsuperscript{14} and the presence of aortic incompetence detected by noting the diastolic flutter of the mitral valve.\textsuperscript{13}

**Table 4**

<table>
<thead>
<tr>
<th>Category</th>
<th>LVO width (mm)</th>
<th>Systolic segment of MV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic stenosis* (pure)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>22 cases</td>
<td>25.4 ± 0.72</td>
<td>21–35 Normal</td>
</tr>
<tr>
<td>Aortic regurgitation (pure)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>13 cases</td>
<td>38.6 ± 2.08</td>
<td>27–52 Normal</td>
</tr>
</tbody>
</table>

Abbreviations: LVO = left ventricular outflow tract; MV = mitral valve; AR = aortic regurgitation; SEM = standard error of the mean.

*The peak systolic gradient was 95 mm or over in 10 patients.
The clinical diagnosis of IHSS co-existing with aortic valve disease presents difficulties as the latter may mask the physical findings of IHSS.1 Awareness of the co-existence of IHSS may be of importance in the medical management of these patients. Routine cardiac catheterization without the use of provocative tests may fail to reveal the existence of associated IHSS.1 Angiograms may not provide diagnostic information unless performed in steep LAO or left lateral position to demonstrate the characteristic severe distortion of the left ventricular cavity together with the anterior position of the mitral valve in systole.21 A small left ventricular end-systolic cavity may sometimes be observed in severe aortic stenosis with concentric left ventricular hypertrophy.21

Removal of fixed obstruction may unmask or aggravate the effects of functional obstruction. This has been incriminated as the cause for the lack of clinical improvement or even deterioration in some patients with aortic stenosis following surgery.1,2 Therefore myotomy or myectomy may be carried out at the time of aortic valve replacement in this group of patients. Knowledge of the associated presence of IHSS may thus be of value to the surgeon.

In the present study, the diagnosis of IHSS was suggested by echocardiography in every patient prior to hemodynamic and/or surgical confirmation. Four of six patients in the present study were identified within the last seven months, during which time 36 patients with aortic valve disease referred to our service were specifically examined by echocardiography for evidence of co-existing IHSS. This high incidence may be due to a high index of suspicion and a diligent search for subtle echocardiographic signs of IHSS.

Echocardiography is a sensitive, noninvasive method of diagnosing IHSS. Our experience suggests that it will prove useful even in the presence of associated aortic valve disease. Following echocardiographic evidence for the co-existence of the two diseases, special attempts to confirm this may be carried out at the time of cardiac catheterization and/or at surgery.

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