Angiographic Anatomy of the Left Ventricle and Mitral Valve in Idiopathic Hypertrophic Subaortic Stenosis

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SUMMARY
The angiographic features of the left ventricle were examined in patients with idiopathic hypertrophic subaortic stenosis who had clinical and hemodynamic evidence of obstruction. Of 36 combined hemodynamic and angiographic studies considered to be technically satisfactory, 33 showed a characteristic combination of abnormalities. In the frontal projection in systole, a linear radiolucent area extended across the left ventricular outflow tract 2 to 2.5 cm below the aortic annulus, at a level corresponding to the site of intraventricular pressure change. In the left oblique and lateral projections, the mitral leaflets did not swing posteriorly in a normal fashion, but projected into the outflow tract during mid and late systole. The radiolucent line, seen in the frontal views, was considered to represent contact of the leading edge of the leaflet with the hypertrophied muscular interventricular septum. The jet of mitral regurgitation, when present, was seen immediately below the anterior mitral leaflet.

Severe hypertrophy was also seen to involve the inferior portion of the muscular septum, causing displacement of the papillary muscles superiorly and to the left. This maldirection of the papillary muscles was postulated to cause abnormal traction on the chordae tendineae and to prevent normal movement of the mitral leaflets away from the septum during systole. The leaflets, held in the outflow tract, form the posterior component of the obstruction, the anterolateral component of which results from severe, asymmetric septal hypertrophy. It was proposed that this mechanism plays an important part in producing the intraventricular pressure gradient in many patients with idiopathic hypertrophic subaortic stenosis.

Additional Indexing Words:
Idiopathic hypertrophic subaortic stenosis
Angiocardiography of left ventricle
Mitral valve
Aortic stenosis, subvalvular

A NUMBER of descriptions of the pathological anatomy in idiopathic hypertrophic subaortic stenosis (IHSS) are now available.1-6 Although postmortem examinations have almost always demonstrated disproportionate hypertrophy of the interventricular septum, narrowing of the outflow tract sufficient to explain the intraventricular pressure gradient often observed during life in such patients usually has not been found. Indeed, it was this discrepancy between the pathological findings and the hemodynamic picture in IHSS that prompted Brock5 to propose the concept of functional obstruction to left ventricular ejection. The theory of muscular or functional narrowing implies that obstruction is not fixed, but develops progressively during systole, and therefore it might be anticipated that a site of obstruction would be evident only in vivo, when the left ventricle is undergoing active contraction; hence, angiography should provide a definitive means for its demonstration. Although it has been suggested that a site of obstruction ordinarily cannot be defined angiographically in patients with IHSS,7 other studies indicate that obstruction can...
be detected\textsuperscript{8–11} and that the area defined angiographically corresponds to the site at which the intraventricular pressure gradient is recorded at cardiac catheterization.\textsuperscript{11}

The present study was undertaken to assess the frequency with which obstruction is demonstrated by angiocardiology in patients with IHSS who are considered to have obstruction on specific clinical and hemodynamic grounds.\textsuperscript{11–13} The group selected included only patients who underwent simultaneous hemodynamic and angiographic evaluation and comprises 36 of the 135 patients with IHSS who have been studied at this institution. The angiographic anatomy of the left ventricle was analyzed in detail, and these observations were then correlated with the clinical and hemodynamic findings. A portion of this work has been presented previously in preliminary form.\textsuperscript{14}

Methods

Patient Selection

On physical examination, each of the patients had an ejection murmur characteristic of IHSS,\textsuperscript{6, 15} the murmur having its maximum intensity at the cardiac apex or left sternal edge, or both (table 1); in 20 patients the murmur was accompanied by a thrill. An atrial gallop sound at the left ventricular (LV) apex was described in 29 patients, and paradoxical splitting of the second heart sound during inspiration was noted in seven patients. A rapid arterial upstroke with a bifid systolic pulse was observed on the indirect carotid pulse tracing or direct arterial pressure pulse, or both, in all but six patients; of the latter six patients, four had minimal or mild obstruction at rest. The electrocardiograms of 27 patients demonstrated an increased amplitude of S\textsubscript{v} and R\textsubscript{v} consistent with left ventricular hypertrophy (table 1); in four additional patients, S-T segments and T-wave abnormalities suggested LV "strain." Abnormal Q waves consistent with ventricular septal hypertrophy were noted in 10 patients. In one patient (T.O.D.), the electrocardiogram (ECG) was considered to be normal at the time he underwent left ventriculography, although several years later the ECG showed left ventricular hypertrophy.\textsuperscript{6}

Obstruction to left ventricular (LV) outflow at rest, and in many patients during provocative maneuvers, was demonstrated at cardiac catheterization (table 1); intraventricular pressure gradients, ranging from 5 to 135 mm Hg, were found at rest in all patients. In 12 patients the pressure was specifically measured in the LV inflow tract.\textsuperscript{11, 12} The arterial pulse pressure response following a ventricular extrasystole was documented in 33 patients, and was abnormal in 32, the pulse pressure being reduced in the first postextrasystolic beat.\textsuperscript{16}

Angiographic Methods

All patients were studied in the postabsorptive state following premedication with sodium pentobarbital, 100 mg. Catheterization of the LV and selective left ventriculography were accomplished either by the transseptal or retrograde arterial technique. Biplane angiograms, exposed in the frontal and lateral projections, were obtained in 47 patients. Thirty-three of these studies were considered satisfactory for analysis, 14 being excluded because of poor opacification of the left ventricle or failure to obtain exposures during systole. The exposure rates were 12 per second (two patients), 6 per second (27 patients), and 4 per second (four patients). Cineangiograms were performed in addition in biplane-film contrast studies in three of these 33 patients, and these combined studies were considered to represent a single evaluation.

Ten selective LV cineangiographic studies were performed, of which three were considered satisfactory. Seven were excluded because of poor ventricular opacification or because angiograms were obtained only in the right anterior oblique projection (see below). Thus, 36 angiographic studies were obtained in 36 patients (33 biplane and three cineangiographic studies).

Cineangiocardiography in the right anterior oblique projection in our experience generally has not revealed a site of obstruction, and in several patients in whom the obstruction could not be visualized in the right anterior oblique projection, it was clearly seen on films exposed in the left anterior oblique or lateral views. Furthermore, in attempting to demonstrate the site of obstruction, it is evident that biplane roentgenograms must be exposed rapidly enough to obtain a sufficient number of films during systole, and the exposure time should be sufficiently short to prevent blurring due to cardiac motion; exposure rates of 12 per second and exposure times of 12 msec or less have proved helpful in demonstrating the site of obstruction. Finally, the kilovoltage of the anteroposterior exposure must be high enough to provide penetration of the contrast-filled outflow tract, which frequently overlies the spine.
### Table 1

**Hemodynamic and Angiographic Findings in Patients with Idiopathic Hypertrophic Subaortic Stenosis**

<table>
<thead>
<tr>
<th>Pt.</th>
<th>LV pr S/ED (mm Hg)</th>
<th>Resting pr grad LV-BA (mm Hg)</th>
<th>LV-BA pr grad during provocative maneuvers</th>
<th>LVH</th>
<th>Q waves</th>
<th>ST-T</th>
<th>Dig.</th>
<th>MI</th>
<th>Obstr</th>
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<tr>
<td>L.W.</td>
<td>158/19*</td>
<td>5-75*</td>
<td>45</td>
<td>70-110</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
<td>0</td>
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<tr>
<td>E.B.</td>
<td>180/20</td>
<td>85*</td>
<td>30</td>
<td>105 110</td>
<td>+</td>
<td>4</td>
<td>0</td>
<td>+</td>
<td>0</td>
</tr>
<tr>
<td>P.T.</td>
<td>229/27</td>
<td>135</td>
<td>145</td>
<td>120</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
<td>0</td>
</tr>
<tr>
<td>R.P.</td>
<td>132/15</td>
<td>30</td>
<td>35</td>
<td>70</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
<td>0</td>
</tr>
<tr>
<td>G.Y.</td>
<td>145/10</td>
<td>15-35</td>
<td>67</td>
<td>60</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
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</tr>
<tr>
<td>L.S.</td>
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<td>16</td>
<td>25</td>
<td>30</td>
<td>+</td>
<td>2</td>
<td>0</td>
<td>+</td>
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<td>85</td>
<td>50</td>
<td>+</td>
<td>2</td>
<td>0</td>
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<tr>
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<td>0</td>
<td>0</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
<td>0</td>
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<tr>
<td>J.P.</td>
<td>160/20</td>
<td>75*</td>
<td>35</td>
<td>20</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
<td>0</td>
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<td>M.O.</td>
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<td>85</td>
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<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
<td>0</td>
</tr>
<tr>
<td>R.S.</td>
<td>185/16</td>
<td>85</td>
<td>35</td>
<td>30</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
<td>0</td>
</tr>
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<td>T.B.</td>
<td>185/16</td>
<td>60-90</td>
<td>65</td>
<td>0</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
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</tr>
<tr>
<td>T.S.</td>
<td>150/24</td>
<td>20-70</td>
<td>0</td>
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<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
<td>0</td>
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<td>J.T.</td>
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<td>50-100</td>
<td>100</td>
<td>0</td>
<td>+</td>
<td>3</td>
<td>0</td>
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<tr>
<td>W.D.</td>
<td>200/0</td>
<td>70*</td>
<td>0</td>
<td>4</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
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<td>C.C.</td>
<td>210/16</td>
<td>135</td>
<td>45</td>
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<td>4</td>
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<td>+</td>
<td>0</td>
</tr>
<tr>
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<td>105*</td>
<td>105</td>
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<td>4</td>
<td>0</td>
<td>+</td>
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<td>M.H.</td>
<td>196/15</td>
<td>87*</td>
<td>35</td>
<td>0</td>
<td>+</td>
<td>4</td>
<td>0</td>
<td>+</td>
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<tr>
<td>J.L.</td>
<td>154/25</td>
<td>10-30</td>
<td>65</td>
<td>0</td>
<td>+</td>
<td>4</td>
<td>0</td>
<td>+</td>
<td>0</td>
</tr>
<tr>
<td>S.B.</td>
<td>190/14</td>
<td>90*</td>
<td>35</td>
<td>30</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
<td>0</td>
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<tr>
<td>M.W.</td>
<td>148/12</td>
<td>46</td>
<td>35</td>
<td>0</td>
<td>+</td>
<td>4</td>
<td>0</td>
<td>+</td>
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<tr>
<td>R.E.</td>
<td>178/9</td>
<td>73*</td>
<td>80</td>
<td>0</td>
<td>+</td>
<td>4</td>
<td>0</td>
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<td>E.Z.</td>
<td>160/22</td>
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<td>64</td>
<td>0</td>
<td>+</td>
<td>4</td>
<td>0</td>
<td>+</td>
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<tr>
<td>B.W.</td>
<td>128/10</td>
<td>6-10</td>
<td>103</td>
<td>3</td>
<td>+</td>
<td>3</td>
<td>0</td>
<td>+</td>
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**ECG**

- LVH, ST-T: 0 +
- LVH, Q waves, ST-T: + +
- LVH, ST-T: + +
- LVH, ST-T: 0 +
- LVH, Q waves: + +
- LVH, ST-T: 0 +
- LVH, Dig.: + +
- LVH, ST-T: 0 +
- LVH, Q waves, ST-T: + +
- LVH, ST-T: 0 +
- LVH, Dig.: + +
- LVH, ST-T: 0 +
- SR alt. with AF, ST-T: + +
- Q waves, ST-T: + +
- LVH, Q waves: + +
- LVH, Q waves, ST-T: + +
- LVH, Q waves, Dig.: + +
Anatomic Anatomy of the Normal Left Ventricle

The normal outflow tract or vestibule is a tubular structure extending between the aortic valve and the body of the interventricular septum. It is lined by the endocardium and is often clearly defined by the radiopaque material from the left ventricular outflow tract. The anterior leaflet of the mitral valve lies just below the aortic valve and is often clearly defined by the radiopaque material from the left ventricular outflow tract. The posterior leaflet of the mitral valve lies just above the aortic valve and is often clearly defined by the radiopaque material from the left ventricular outflow tract. The anterior leaflet of the mitral valve is visible in the lateral view (fig. IC and D). In the anterior-posterior (AP) projection (fig. IA and B), a cardiac shadow is visible in the diastolic phase, while the posterior leaflet of the mitral valve is not visible. In systole, the mitral valve annulus is clearly visible in the diastolic phase, while the posterior leaflet of the mitral valve is not visible. The mitral valve annulus is visible in the systolic phase, while the anterior leaflet of the mitral valve is not visible.

Abbreviations: Pt. = patient; LV = left ventricular; Pr = pressure; S = systolic; ED = end-diastolic; BA = brachial artery; AO = aorta; Val = Valsalva maneuver; Iso = Isoproterenol; VPC = arterial pulse pressure response following premature contraction; S1 = atrial diastolic gallop; Paradox S1 = paradoxical splitting of the second heart sound with respiration; LVH = LV hypertrophy; ST-T = S-T segment + T wave abnormalities; Dig = digitalis; MI = mitral insufficiency; Obstr = obstruction; * = patients in whom left ventricular inflow tract pressure was recorded; + = abnormal response; 0 = normal response; --- = no observation made.
Figure 1

Left ventricular angiograms exposed in diastole in frontal and lateral projections with explanatory line drawings in patients with valcular aortic stenosis and left ventricular hypertrophy. The lateral and frontal views are from two different patients. In the lateral views (C and D), the anterior mitral leaflet (AML) (small arrow) which forms the posterior boundary of the outflow tract.
atrium. The portion of the mitral valve annulus to which the anterior mitral leaflet attaches is easily located in the AP projection, since it is continuous with the annulus of the aortic valve (fig. 1A and B). The opened mitral leaflets are not visible in diastole in the AP projection.

The features of the relatively normal left ventricular outflow tract during systole in one of these patients with valvular aortic stenosis are shown in figure 2. In the lateral projection (C and D), the interventricular septum bulges somewhat into the LV outflow tract; however, the anterior mitral leaflet swings posteriorly with a consequent widening of the outflow region, and there is a considerable pool of contrast material between the edges of the closed leaflets and the anterior wall of the left ventricle. With the aortic valve ring as a reference point, the small segment of membranous septum (MeS) is identified, the bulge inward immediately below this area representing the junction with the muscular septum (MuS). The muscular septum forms the remainder of the anterior wall of the left ventricle, and the septum extends around the left ventricle to form a portion of the inferior wall as well. In the AP projection in normal systole, the posterior fornix is not clearly visible, and the apposed edges of the mitral leaflets are not seen (fig. 2A and B), since a pool of contrast material lies between the interventricular septum and the closed valve leaflets.

The papillary muscles usually can be identified in systole in both the frontal and lateral projections, as distinct impressions along the superior and the inferior margins of the body of the left ventricle (fig. 2). The anterior papillary muscle (APM) is represented by the superior indentation in both views. The posterior papillary muscle (PPM) lies inferiorly. Normally, in the AP view, the direction of the papillary muscles is relatively parallel to the long axis of the left ventricle, and therefore they are aligned toward the appropriate commissures of the mitral valve (fig. 2), the chordae tendineae from the anterior and posterior papillary muscles attaching to both mitral leaflets adjacent to the anterior and posterior commissures, respectively (see figure 7). Neither the chordae tendineae nor the edges of the mitral leaflets are seen on normal frontal angiocardiograms during systole.

**Angiographic Anatomy of the Left Ventricle in IHSS**

On angiograms exposed in diastole in patients with IHSS in the lateral view, the outflow tract is markedly narrowed by the hypertrophied muscular interventricular septum, and an inverted cone, formed by the hypertrophied septum anteriorly and the open anterior mitral valve leaflet posteriorly, is usually visible (figs. 3F, 4C, and 5B). A large myocardial mass, also representing hypertrophy of the septal musculature, is characteristically seen to bulge into the body of the left ventricle inferiorly (figs. 4C and 5B). In the AP projection in diastole, the hypertrophied septum is visualized superiorly, lateral to the aortic root, as well as inferiorly as it bulges into the left ventricular cavity (fig. 5A).

In films exposed during systole in patients...
Figure 2

Left ventricular angiogram and diagrams in systole in the patient shown in figure 1C and D. Abbreviations are the same. In the lateral view (C and D), the anterior mitral leaflet (arrow) is outlined by a slight degree of contrast material regurgitating through the mitral valve. The anterior bulge opposite to it represents muscular interventricular septum (MuS) and the
with IHSS, in the lateral view the hypertrophied interventricular septum protrudes further into the outflow tract (figs. 3D and E, 4A and B, and 5D). However, the mitral leaflets generally do not swing posteriorly to widen the outflow tract, as in the normal heart. Instead, the leading edges of the leaflets appear to be held in the outflow tract, creating a curved, shelf-like deformity (figs. 3D and E, 4A and B, 5D, and 6B). In the AP projection in systole this leading edge contacts the septal musculature and is visible as an irregular linear radiolucency (figs. 3A and B, 5C, 6A, and 8A). This radiolucent band may be scalloped or V-shaped and has a configuration resembling that of the free edge of the anterior mitral leaflet (fig. 7). Several additional factors lead to this conclusion: (1) on cineangiograms made in the left anterior oblique projection, the anterior commissure can sometimes be appreciated and is continuous with the linear density; (2) mitral regurgitation, when present, occurs immediately beneath the shelf-like projection of the anterior leaflet (fig. 3C and D); (3) rarely, the radiolucent line is "W" rather than "V" shaped (fig. 3A and B), a configuration that is consistent with visualization of both anterior and posterior leaflets, or with the occurrence of the anatomic variant termed "commisural cusps." The obstructing position of the mitral leaflets can be seen in mid as well as late systole, and figure 4A and B shows two consecutive exposures one-twelfth second apart, illustrating this point. Above this zone of obstruction, in the region of the membranous interventricular septum, the outflow tract appears to be of normal caliber.

As in other forms of ventricular hypertrophy, the papillary muscles are usually enlarged in IHSS. In addition, the axis of these structures is often deflected in IHSS by asymmetrically hypertrophied interventricular septum, represented by the inferior extent of the large muscle mass comprising the interventricular septum. Thus, the papillary muscles may not point toward the mitral annulus but instead often are displaced leftward from the central longitudinal axis in the AP projection by as much as 6 cm (average of 3.5 cm) (fig. 8). In the lateral view, this displacement is generally reflected by a superior rotation of the axis of the papillary muscles (figs. 3 and 8).

**Clinical and Hemodynamic-Angiographic Correlations**

The clinical, hemodynamic, and angiographic correlations are summarized in table 1. Asymmetric hypertrophy of the interventricular septum was present in all of the patients. Mitral regurgitation was visualized angiographically in 19 of the 36 studies, or 53%. Obstruction in the left ventricular outflow tract caused by the septal hypertrophy and abnormal systolic position of the mitral leaflets was observed angiographically in 33 of the 36 patients, or 92% (table 1). Mitral regurgitation was present in 17 of the 33 studies (52%) in which obstruction was demonstrated. All patients with obstruction demonstrated angiographically had gradients between the LV and aorta or the LV and peripheral artery at rest (table 1). In 12 of these, the pressure was measured in the LV inflow tract with an end-hole catheter and was elevated. The pressure in the inflow tract was not specifically defined in the remaining patients. The arterial pulse pressure response following a premature contraction was recorded in 30 of these 33 patients. It was abnormal in 29 patients, and in one patient it was normal. In addition, in 28 of the 33 patients (74%) exhibiting angiographic evidence of obstruction, the arterial pressure pulse was bifid.

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membranous septum (MeS) lies above this. The outflow tract is wide in systole despite contraction of the muscular septum, because of the posterior excursion of the anterior mitral leaflet (AML) and posterior mitral leaflet (PML). The papillary muscles can be seen lying parallel to the long axis of the left ventricle. Note the extreme degree of emptying or obliteration of the ventricular apex in this patient with valvular aortic stenosis and LV hypertrophy.
Figure 3

Left ventricular angiogram and diagrams of a patient with IHSS (patient E.B.). In diastole, in the frontal projection (C) there is diminished diastolic volume of the chamber as well as distortion of the left ventricular cavity, especially inferomedially, with elevation of the posterior papillary muscle (lower black arrow). A lateral film exposed simultaneously (F) shows the “diastolic cone” deformity composed of the anterior mitral leaflet (upper arrow) and the hypertrophied muscular septum anteriorly. The black and white arrows (C and F) show the fornix under the posterior mitral leaflet. In systole in the frontal projection (A and B), an irregular radiolucent line is present several centimeters below the aortic ring, which has the configuration of the leading edge of the mitral leaflet (arrow, AML). The hypertrophied papillary muscles (APM and PPM) are visible. Note the moderate residual volume remaining in the LV cavity at the apex. In the lateral projection, during systole (D and E), the abnormal shape of the mitral leaflets can be seen (AML, PML) with the leading edges held forward, in the outflow tract, opposite the hypertrophied muscular interventricular septum. Minimal mitral regurgitation (MR) can be seen between the leaflets.
It is also noteworthy that, of the 33 patients in whom obstruction was demonstrated angiographically, 20 (61%) had thrills accompanying the systolic murmur, 26 (79%) had an atrial gallop sound ($S_4$), 7 (21%) had paradoxical splitting of the second heart sound, and 32 (97%) had ECG abnormalities (table 1).

All three patients (J.G., C.B., and R.F.) in whom no obstruction was identified angiographically exhibited pressure gradients at rest (range, 14 to 45 mm Hg), although in one (C.B.) the gradient was intermittent. In two of the three patients mitral regurgitation was visualized. An abnormal response following a premature contraction was present in all three patients, and the arterial pulse was bimodal in two patients. Although all three had atrial gallop sounds, none had an apical systolic thrill, and none had paradoxical splitting of the second heart sound. In all three patients the ECG was abnormal (table 1).

Discussion

These studies in a selected group of patients with IHSS who had both clinical and hemodynamic evidence of obstruction indicate that angiographic signs of obstruction in the outflow tract are more common than has generally been appreciated. It must be recognized, however, that in some patients considered to have this disease on a familial basis intraventricular pressure gradients cannot be demonstrated, nor do such patients exhibit clinical features suggesting obstruction; moreover, as initially described by Criley and associates,7 other patients exist who may or may not have IHSS, in whom an elevated pressure can be recorded or induced only when the catheter tip is positioned within the ventricular apex.7, 12, 13 Again, however, the latter patients usually do not exhibit the clinical and hemodynamic features typically found in patients with IHSS and obstruction,11–13 nor do they exhibit obstruction angiographically. The present hemodynamic and angiographic analysis has tended to include few patients with these forms of IHSS, since combined hemodynamic and angiographic studies generally were performed.
**Figure 5**

Left ventricular angiograms in a patient (T.B.) with IHISS. During diastole (A and B) there is distortion of the inferior border of the left ventricle medially by hypertrophy of the IV septum. The posterior fornix (arrows), a normal diastolic landmark, is shown as a smooth crescentic outline in the AP films several centimeters below the aortic valve and as the posterior extent of the left ventricle in the lateral films. In systole (C and D), the anterior mitral leaflet (arrows) is restricted from posterior motion and held forward in the lateral projection (D) so that it comes in contact with the septum. A V-shaped irregular line is seen in the frontal plane (C), corresponding to the leading edge of the mitral leaflet. This is quite distinct from the smooth arc of the line of attachment of the posterior mitral leaflet.
Figure 6
Systolic left ventricular angiogram in a patient (M.W.) with IHSS. The irregular line (arrows) is seen, corresponding to the leading edge of the mitral valve in the frontal projection (A), and the abnormal systolic position of the valve in lateral projection (B) is also shown.

Figure 7
Photograph of the normal left ventricle, with the anterior wall removed, in the same position as frontal angiocardiograms. The leading edge of the anterior mitral valve leaflet is essentially V shaped, similar to the appearance of the structure that contacts the septum and forms the posterior component of the obstruction in IHSS.

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only in patients with severe clinical manifestations of this disease, who were considered to have obstruction. Therefore, general conclusions concerning the incidence of these angiographic findings of obstruction within the spectrum of IHSS should be made with caution and it must be recalled that other mechanisms, such as diminished ventricular diastolic compliance and mitral regurgitation, are of variable importance relative to obstruction in patients with this disease. In addition, while the incidence of obstruction was high in patients with specific clinical and hemodynamic features, in three patients (8%) who also exhibited these features, the angiogram failed to reveal a site of obstruction.

The bulge of the interventricular septum into the LV outflow tract in patients with IHSS has previously been well demonstrated.
Figure 8

Left ventricular angiogram and accompanying explanatory drawing in the same patient (M.W.) shown in figure 6. These films were exposed earlier in systole, before complete emptying of the left ventricle, and better illustrate the abnormal axis of the papillary muscles (APM, PPM)

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on angiograms of the left ventricle. As discussed earlier, the posterior wall of the outflow tract is formed almost entirely by the anterior leaflet of the mitral valve and if, as indicated by hemodynamic studies, the obstruction resides in the left ventricular outflow tract, this leaflet must provide one component of the narrowed area. The great mobility of this structure could well explain difficulties previously encountered in identifying an area of obstruction at postmortem examination and on angiography, although several investigators have considered previously that there was a posterior component to the narrowed area. Wigle and his co-workers, in 1962, called attention to an inconstant filling defect posteriorly, which was thought to result from posterior extension of the septal hypertrophy. Nordenström and Ovenfors noted, in two of eight patients with IHSS, that the anterior mitral leaflet appeared to move forward in systole against the anterior portion of the outflow tract, producing a sharp indentation in the contrast medium. Fix and associates also observed a posterior filling defect that was thought to result from restriction of the normal posterior excursion of the anterior mitral leaflet. The latter authors also found this leaflet and its chordae to be abnormally long on postmortem specimens and considered that this added length contributed to the abnormal position in the outflow tract. In previous studies from this laboratory, a linear density in the frontal projection observed on left ventricular angiograms exposed during systole was considered to represent impingement of the anterior mitral valve leaflet against the hypertrophied interventricular septum and to coincide with the site of obstruction. Whether or not these angiographic findings represent the same phenomenon that Björk and associates attributed to abnormal insertion of the anterior mitral leaflet remains to be clarified. Although the anatomy of case 1 described by Björk and associates appears consistent with malinsertion of the mitral leaflet, the angiographic features of case 2 resemble more those described in the present group of patients. Recently, in angiographic studies by Dinsmore and associates, the leading edge of the anterior mitral leaflet was also visualized during systole in patients with IHSS; these authors considered that this phenomenon resulted from abnormal traction by the chordae tendineae and that it was of significance relative to the occurrence of mitral regurgitation.

Many of the pathological features of IHSS can be explained on the basis of malposition of the mitral leaflets. Elongation of the mitral leaflet and the chordae tendineae possibly could be caused by abnormal traction on these structures. In addition, the mitral leaflets are often thickened, and the endocardium is also thickened over the anterior septal bulge. The latter site lies immediately opposite the anterior leaflet and corresponds to the point at which the mitral leaflet would contact the interventricular septum.

A tentative concept of the pathogenesis of the obstruction observed in the patients with IHSS in the present study may be developed as follows. Asymmetric hypertrophy of the interventricular septum, which encircles the anterolateral aspect of the left ventricle, could be the primary alteration. Narrowing of the anterior portion of the outflow tract is caused by direct impingement of this hypertrophied muscle. The inferior portion of the hypertrophied septum projects into the diaphragmatic aspect of the ventricular cavity, resulting in a change in the axis of the papillary muscles. This protrusion of the inferior

in relation to the mitral ring. The large bulk of septal musculature beneath has elevated and deflected the posterior papillary muscle (PPM), with resultant maldirection of the anterior papillary muscle (APM). Traction on the mitral leaflets (AML and PML) may be produced by this mechanism, holding them forward in systole. The anterior mitral leaflet then contacts the hypertrophied septal muscle, resulting in visualization of the leading edge of the mitral leaflet (AML) in the frontal projection (A and B).
portion of the septal mass also causes an angulation of the left ventricular cavity, and little apex-to-base shortening of the left ventricle occurs, as noted by Klein and associates.25 The angulation of the papillary muscles and the lack of apex-base shortening cause abnormal traction on the chordae tendineae and hence on the mitral valve leaflets during systole. Thus, while normally the outflow tract is widest during ventricular systole,26 in IHSS the mitral leaflets do not complete their normal excursion from the outflow tract, and thereby they produce the posterior portion of the obstruction. Both leaflets are pulled forward, since the mode of attachment of the chordae tendineae precludes independent motion of one leaflet (fig. 7); however, since the anterior leaflet is longer and more anterior, its contribution to the obstruction is probably considerably greater than that of the posterior leaflet. Restriction of the normal posterior motion of the mitral leaflets could also prevent coaptation of their edges and thereby often result in mitral regurgitation.

References

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IDIOPATHIC HYPERTROPHIC SUBAORTIC STENOSIS


Angiographic Anatomy of the Left Ventricle and Mitral Valve in Idiopathic Hypertrophic Subaortic Stenosis
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