RADIOLOGIC NOTES IN CARDIOLOGY

Angiographic Appearance of Idiopathic Hypertrophic Subaortic Stenosis

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The functional nature of obstruction in idiopathic hypertrophic subaortic stenosis (IHSS) was initially proposed by Brock. This implies that the obstruction is not fixed but develops progressively during ventricular contractions, and that the obstruction can be variable so that the patient may manifest different pressure gradients in the left ventricular outflow tract at different times.

In contrast to fixed obstruction, in which a specific anatomic site can be demonstrated at postmortem, the predominant morphologic change in patients with IHSS is severe asymmetric hypertrophy of the interventricular septum. Characteristic angiocardiographic changes are present in the left ventricle in IHSS, reflecting the altered morphology as well as demonstrating a point of obstruction.

Generally, the most striking abnormality present angiographically is severe distortion of the left ventricle (figs. 1–4). This is seen in diastole in the lateral projection as a prominent bulge into the anterior aspect of the outflow tract, caused by the hypertrophied muscular septum. The normal forward position of the anterior mitral leaflet brings the structure in proximity and occasionally into contact with the hypertrophied septum. In the frontal projection, the left ventricular outflow tract is indented along the left lateral aspect by the hypertrophied septum, as is the inferomedial aspect of the ventricle.

During systole (figs. 5–8), in the lateral projection, one sees further projection of the muscular septum into the anterior portion of the outflow tract, as well as abnormal position of the mitral leaflet. Instead of the usual posterior excursion of the anterior mitral leaflet, the leading edge of the leaflet is held in the outflow tract, creating a curved appearance to the leaflet and a shelflike projection into the outflow tract. Occasionally, the position of the posterior mitral leaflet is seen as well; this structure follows the course taken by the anterior mitral leaflet and is held in the outflow tract. In the frontal projection (figs. 6, 8), a radiolucent band is present, caused by contact of the anterior mitral leaflet with the hypertrophied interventricular septum. The features which characterize this radiolucent band as mitral valve are its V shape, which corresponds to the leading edge of the leaflet, and the location in relation to the aortic valve.

Cineangiographic studies have demonstrated in detail the motion abnormality of the mitral leaflet and hypertrophied septum. The ventricular septum moves inward to encroach on the outflow tract in early systole while the attached portion of the anterior mitral leaflet moves posteriorly, away from the ventricular septum in the direction of a normal mitral valve.
In diastole, in the frontal projection, the asymmetric septal hypertrophy distorts the ventricle by indenting the left lateral aspect of the outflow tract as well as the inferomedial portion of the ventricular cavity (arrows).

In the lateral projection in diastole, the bulge of septal musculature can be seen indenting the anterior portion of the outflow tract (arrow). The normal forward position of the anterior mitral leaflet (arrow) brings it in close proximity to the septal hypertrophy.
Figure 3

Frontal plane in diastole. See description of figure 1.

Figure 4

Lateral plane in diastole. See description of figure 2.
In the lateral projection in systole, the bulge of septal musculature is visible (arrow). The anterior mitral leaflet (pointer) remains with its leading edge in the outflow tract, contributing to the obstruction.

Mitral Regurgitation

Mitral regurgitation occurs frequently in IHSS. The regurgitation, in general, tends to
In the frontal projection in systole, there is a V-shaped radiolucent defect in the outflow tract (arrows) caused by contact of the anterior mitral leaflet with the hypertrophied septum.
**Figure 7**

*Lateral plane in systole. See description of figure 5.*

**Figure 8**

*Frontal plane in systole. See description of figure 6.*
be mild to moderate and occurs in early systole.

The incidence is subject to some dispute. One study has claimed that in 50% of patients in whom pressure gradients were present at rest there was mitral insufficiency. Another study has demonstrated mitral regurgitation in all patients in whom pressure gradients were present at rest. One possible explanation for this discrepancy would be that biplane large-film angiocardiography, which was the method employed by the first-mentioned study, is not as sensitive a technic for appreciation of mitral regurgitation as is cineangiocardiography, the method used by the second group, and that very minimal amounts would not be detectable by large-film angiography.

Right Ventricular Obstruction

Marked hypertrophy of the interventricular septum frequently impinges on the outflow tract of the right ventricle, and pressure gradients ranging from 5 to 15 mm Hg are quite commonly demonstrated in the right ventricular infundibulum in patients with IHSS. Rarely, the disease is more dominant on the right side than the left; this has been generally found in children. Angiographically, the obstruction is seen as a large muscular band separating the inflow from the outflow tract (fig. 9). This resembles angiographically the “anomalous muscle bundle of the right ventricle.”

Differentiation from Nonobstructive Cardiomyopathy

Experimentally and clinically, intraventricu-
lar pressure differences can be recorded in the absence of obstruction when the ventricle empties rapidly and completely and the catheter tip lies within obliterated portions of the left ventricular cavity. This phenomenon is termed "cavity obliteration," and the recording by the catheter of the left ventricular wall tension has led to spurious diagnoses of intraventricular obstruction. Various hemodynamic maneuvers are employed to differentiate patients with cavity obliteration from patients with true IHSS, the most reliable being measurement of left ventricular inflow pressures simultaneously with aortic pressure.

The angiographic appearance of patients with cavity obliteration can be a small but normally shaped ventricle in diastole (fig. 10). In some patients there is some septal hypertrophy with indentation of the outflow tract as well as the inferomedial surface of the ventricle. During systole there is marked emptying of the left ventricle so that all that remains is the inflow portion of the cavity. The apex is completely obliterated, and all that can be seen of this portion is a ribbon of contrast material separating the two papillary muscles. There is no evidence of obstruction in the outflow tract, and the mitral leaflets have a normal appearance in systole.

References

Figure 10
Cineangiocardiographic frames, frontal projection, of a patient with cavity obliteration. In diastole (left), there is slight distortion of the apex of the left ventricle. In systole (right), the apex is completely obliterated (arrow). The outflow tract shows no evidence of the radiolucent line or obstruction.
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Circulation. 1972;46:614-622
doi: 10.1161/01.CIR.46.3.614
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/46/3/614.citation

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