Recognition of Left Ventricular Outflow Obstruction

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The purpose of this paper is to summarize the clinical, hemodynamic, and angiographic aspects of valvular, subvalvular, and supravalvular aortic stenosis, with emphasis upon recognition of the more severe grades of the diseases that may require operation.

A basic feature of obstruction to left ventricular outflow is the need for the ventricle to produce high systolic pressure; a systolic pressure gradient across the obstruction is present. The chronic elevation of left ventricular pressure is associated with left ventricular hypertrophy, which may also cause hemodynamic abnormalities. The bedside findings constitute the essential first step in assessing these physiologic abnormalities, to select those patients who will require left heart catheterization and may need surgical relief.

Valvular Aortic Stenosis

The spectrum of valvular stenosis is wide, both in terms of severity and age of the patient. Peak systolic pressure gradients across the aortic valve range from a few mm. Hg to 150 or more (fig. 1). Patients range from the newborn to the elderly. Generally, systolic gradients below 40 mm. Hg result in few or no symptoms, and tolerance for average activity is unimpaired. On the other hand, those with exertional dyspnea or syncope have more severe obstruction, usually with pressure gradients of 70 mm. Hg or more. It is well known that individuals with severe stenosis may suffer sudden unexpected death at any age. Angina pectoris is common but is a relatively inexact guide to the tightness of the obstruction. Some patients with moderate pressure gradients (around 50 mm. Hg) have angina of troublesome frequency whereas occasionally others with more severe stenosis are free from chest pain. In this regard associated coronary artery disease presents a difficult diagnostic problem in some patients with aortic stenosis, particularly men over the age of 35. Surgical correction of valvular stenosis of severe degree is very likely to abolish angina in patients without coronary artery disease, whereas mild or moderately severe aortic stenosis when angina is due to coronary artery disease presents a difficult diagnostic and surgical problem. Coronary arteriography probably will be employed more often in the near future to distinguish such patients. Vectorcardiograms have been of help in excluding anterior wall infarction in patients with left ventricular hypertrophy and may be useful in certain patients who present this problem.

True congestive heart failure in adults occurs in a late stage of the disease and survival often is brief. Recognition of aortic stenosis remains important in this situation, however, because corrective surgery can be performed with salvage of some otherwise hopeless patients (fig. 2d).

Severe stenosis in infancy is announced by heart failure and has a dismal prognosis with a high proportion of deaths in the first few weeks of life. Coarctation of the aorta or patent ductus arteriosus is frequently associated, and pulmonary hypertension results in many. Distinction has been made from aortic atresia and other varieties of the hypoplastic left heart syndrome. Many older children with congenital aortic stenosis are completely asymptomatic until the teens or adulthood, even if the stenosis is relatively severe. Thus,

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except for the infant, heart failure is rare in childhood. Nevertheless, sudden death can occur and recognition of the lesion is of obvious import.

Auscultatory findings are often similar in valvular and discrete subvalvular stenosis and, indeed, all the physical findings can at times be identical. The murmur is of ejection type and diamond shape in phonocardiograms.\(^{11,12}\) It is often best heard in the left third intercostal space or even at the cardiac apex, as well as at the classical aortic area. Radiation of the murmur to the neck is characteristic. When the murmur is prominent at the apex, its timing in relation to the first heart sound is of fundamental importance in distinguishing aortic stenosis (ejection murmur) from mitral insufficiency (pansystolic murmur).\(^{13,14}\) Beyond establishing the presence of valvular or subvalvular stenosis, the murmur has not been of great help to us in further estimating the severity of the obstruction. A systolic ejection click is commonly heard in congenital valvular stenosis before valvular calcification or other degenerative changes occur in the valve, but it is not a dependable means of estimating the transvalvular pressure gradient or consistently excluding the discrete subvalvular form.

Other physical findings indicate the physiologic significance of the stenosis more consistently. A slow rising, small peripheral pulse is a helpful sign, and we find the brachial artery most useful to examine, as have others.\(^{1}\) A systolic thrill is uniformly present with aortic stenosis in children though this finding does not prove the existence of a large transvalvular pressure gradient. A thrill is perhaps found less consistently in older adults but is more often indicative of a large gradient. A lifting left ventricular apex beat is an essential component of the diagnosis of important stenosis. A presystolic gallop is commonly present with severe obstruction to left ventricular outflow and undoubtedly is a reflection of left ventricular hypertrophy. It is felt with the hand as a double apical impulse or heard as a fourth heart sound.\(^{2}\) A single second heart sound throughout the respiratory cycle and paradoxical splitting of the second sound have been consistent indicators of serious stenosis.\(^{15,16}\) A normal second heart sound does not exclude important outflow obstruction, however.

Mild aortic insufficiency is present in some patients with congenital stenosis as well as in many with a presumed rheumatic etiology. Though slight insufficiency is more common in the discrete subvalvular type of obstruction, its presence or absence is not of differential diagnostic help.

Chest radiographs may or may not show
enlargement of over-all cardiac dimensions, but commonly there is rounding of the left ventricular portion of the silhouette (fig. 2). In congenital valvular stenosis, poststenotic aortic dilatation is a useful sign but does not always exclude discrete subvalvular stenosis. Poststenotic dilatation is not of marked degree in the latter case, however. With pro-

Figure 2

A, upper left. Valvular aortic stenosis in a 41-year-old woman with a peak transvalvular gradient of 110 mm. Hg. Slight poststenotic dilatation of the aorta is present. B, upper right. Valvular aortic stenosis in a 59-year-old woman with a peak gradient of 160 mm. Hg. Marked poststenotic dilatation of the ascending aorta is apparent. C, lower left. Valvular aortic stenosis in a 74-year-old man (See fig. 1C). D, lower right. Aortic stenosis with insufficiency in a man with congestive heart failure and a cardiac index of 1.5 L./min./M². Valve replacement resulted in marked improvement.
gression of the disease cardiomegaly develops and can become extreme (Fig. 2). Valvular calcification is a dependable indication of the valvular site of the stenosis and is most easily identified with the use of image amplifier fluoroscopy.

The electrocardiogram is of considerable value in assessing the severity of the disease, though exact correlations are somewhat confused by varying criteria for the diagnosis of left ventricular hypertrophy without marked ST-T abnormalities. The pattern of "left ventricular strain," which presents definite ST-T wave abnormalities as well as high QRS voltage, is consistently associated with important stenosis and necessitates further study of the patient. Abnormalities of voltage alone are less dependable because of the wide range in normal individuals. A disquieting report has described sudden death in a child with severe aortic stenosis and an electrocardiogram that failed to show the pattern of left ventricular strain. It can be concluded that the combination of abnormal QRS voltage and ST-T wave abnormalities is a dependable indication of important stenosis, but a normal electrocardiogram in the young patient does not exclude important obstruction, and will be found in about 25 per cent of such cases. A normal record is uncommon in adults with important stenosis. The vectorcardiogram has been reported to be a more sensitive indicator of left ventricular hypertrophy, and a recent study indicates a consistent relationship of vectorcardiographic findings and left ventricular systolic pressure.

If clinical assessment of the patient suggests that important left ventricular outflow obstruction is present, left heart catheterization is indicated. With this procedure the physiologic magnitude of the stenosis can be accurately determined. In the absence of aortic valve calcification, left ventricular or aortic angiography is also indicated to define the site of the obstruction. If valvular stenosis is suspected, the author believes that the transseptal route is the method of choice for left heart catheterization in adults and larger children. Steady-state measurements of left ventricular and systemic arterial pressures can be obtained as well as a determination of cardiac output by indicator dilution, and the right heart can be entered from the same approach. Left atrial pressure is an important factor in these diseases and is also measured by this method. In addition, transseptal angiography can be performed to demonstrate the site of the stenosis. In the absence of mitral stenosis, no difficulty has been encountered in entering the left ventricle when a curved catheter is used in the transseptal technique. It must be pointed out, however, that percutaneous retrograde arterial catheterization of the left ventricle is preferred by many. Although it requires crossing the valvular stenosis, which can be difficult, it does provide a "pull-out" pressure record that localizes the site of obstruction. It also permits contrast studies of the aortic root which may be very useful for the demonstration of aortic insufficiency or the dome-shaped valve in congenital valvular stenosis. In small children we prefer the retrograde route from a cutdown over a brachial or femoral artery. When the foregoing methods fail to provide a measurement of left ventricular pressure, percutaneous puncture of the left ventricle is employed.

With large pressure gradients across the aortic valve, elevations of left ventricular end-diastolic pressure are found rather often. We are convinced that this finding by itself need not indicate left ventricular failure. Raised left ventricular end-diastolic pressure can be considered a reflection of decreased ventricular distensibility due to left ventricular hypertrophy in many patients. Studies of left ventricular volume or dimensions in valvular aortic stenosis fail to show significant enlargement of chamber volume in many patients, and end-diastolic pressure-volume relationships have not been consistent. Left atrial pressure records in this situation demonstrate large atrial contraction waves. In the absence of mitral valve disease this finding in aortic stenosis is believed due to left ventricular hypertrophy, and consistently is associated with a signifi-

Supravalvular angiography in congenital valvular stenosis will often display ballooning of the valve into the proximal aorta with systole. The smooth contour of such a valve indicates absence of gross degenerative changes in it (fig. 3). The valve which is grossly calcified, whether congenital or rheumatic, has a fixed, rough contour, presenting filling defects in the stream of contrast agent (fig. 4).

In terms of surgical correction, a significant transvalvular gradient has been defined as 50 mm Hg or more in younger patients with congenital stenosis who can have open aortic commissurotomy. The criteria for operation are less well defined for those who can be predicted to need aortic valve replacement. At present, most of our patients with isolated valvular aortic stenosis who require valve replacement, have peak pressure gradients that exceed 60 mm Hg or have calculated effective orifice areas from 0.30 to 0.80 cm². Though operative decisions are greatly influenced by the hemodynamic findings, rigid criteria for operability cannot be outlined on this basis at present.

Discrete Subaortic Stenosis

A band or diaphragm across the outflow tract of the left ventricle can produce obstruction of severity equal to that found in valvular stenosis. These patients may have all of the symptoms that occur in the valvular form, such as exertional dyspnea, angina pectoris, syncope, and easy fatigability. Sudden unexpected death also occurs.

Physical findings may be identical to those
found in the valvular type with regard to the heart murmur and collateral signs associated with left ventricular hypertrophy. An easily heard systolic ejection click strongly suggests valvular stenosis without calcification rather than the subvalvular variety, although it fails completely to separate these two lesions in all patients. The location of the maximum intensity of the murmur has not been of differential diagnostic help in our hands. The murmur of slight aortic insufficiency occurs in both types of stenosis but is more common in the subvalvular variety. Physical findings of mitral valve disease in addition to those of left ventricular outflow obstruction clearly indicate valvular aortic stenosis of rheumatic etiology, and thus rule out subvalvular obstruction.

Chest films in discrete subaortic stenosis less often show poststenotic dilatation of the aorta than in valvular stenosis and even when present, it is not of marked degree. Valvular calcification is absent.

As stated in the previous section, the findings of significant left ventricular outflow obstruction indicate the need for left ventricular catheterization and angiography to measure the pressure gradient across the stenosis, the cardiac output, and to prove the site of the obstruction (fig. 5). In discrete subaortic stenosis the narrowed subvalvular zone of the left ventricle can be identified in angiocardiograms and if the retrograde arterial route is chosen to catheterize the ventricle, a pull-out recording of pressure will further confirm the locus of the obstruction.

**Supravalvular Stenosis**

The syndrome associated with this circular constricted zone just above the aortic valve has been more completely elucidated recently, and a clinical diagnosis now can be made from the physical findings. Most patients with supravalvular stenosis bear a remarkable facial similarity to one another, characterized by broad forehead, pouting lips, heavy cheeks, and pointed chin. Mental deficiency is associated. The patients are physically underdeveloped and dental malformation is present.

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**Figure 5**

Discrete, fixed subaortic stenosis. The obstruction is shown by the indentation below the aortic valve in this left ventricular angiogram.

There is an ejection murmur best heard in the right first or second intercostal space. Neither an aortic ejection click nor a diastolic murmur has been reported in this particular syndrome to our knowledge. The blood pressure may be different in the two arms, with the right higher than the left as a rule; but there is no anatomic obstruction of the subclavian vessels.

A clear understanding of the natural history of this disorder is not yet available because of the limited number of cases described so far. It is significant in this regard that most of the patients reported have been children. Some have had no symptoms while limitation of exercise tolerance and angina pectoris have occurred in others. Sudden death has been reported. This disease may be related to idiopathic hypercalcemia of infancy.

Chest films show a diminution in the size of the aortic knob, even though the aorta distal to the stenosis is often of normal caliber. Poststenotic dilatation of the aorta is not found. Electrocardiographic variations are usually similar to those in valvular and discrete subvalvular stenosis, though right ventricular hypertrophy has been reported.

Left ventricular catheterization will demonstrate a systolic pressure gradient between the ventricle and a systemic artery, and retrograde catheterization of the ventricle will per-
mit pull-out recording of pressure. Aortic systolic pressure in the region just above the valve will equal left ventricular systolic pressure and then systolic pressure will drop as the catheter tip crosses the stenotic zone. Left ventricular angiography demonstrates aortic narrowing in the region just above the valve (fig. 6).

Elevated right ventricular pressure has been found in this disease, due to bilateral peripheral pulmonary arterial stenoses, demonstrated angiographically. This was present in all of 10 patients in one group with supravalvular aortic stenosis and probably will prove to be a consistent part of the syndrome. It is not necessarily an incidental finding, since right ventricular systolic pressure as high as 110 mm Hg was found. Right ventricular hypertrophy in the electrocardiogram has been described, and is likely related to the pulmonary arterial stenoses. It would otherwise present a confusing picture in a patient with signs of left ventricular outflow obstruction, and should call to mind this diagnosis.

Supravalvular stenosis is surgically correctable and the reports of success have been summarized recently.

Other anatomic types of supravalvular stenosis have been reported, particularly involving bands in the vessel and apparently not associated with the facial characteristics described above. Aortic insufficiency has occurred with this type due to associated aortic valve abnormalities. The syndrome described as a circular aortic constriction with characteristic facial features is probably the most common variety of supravalvular obstruction.

**Hypertrophic Subaortic Stenosis**

Interest in this fascinating disorder has become widespread in the short period it has been recognized. At present it appears that severe ventricular hypertrophy of unknown cause results in systolic obstruction to left ventricular outflow, often related particularly to septal enlargement. The right and left ventricular free walls are similarly involved by the process, however, and obstruction to right ventricular outflow is not uncommon in the disorder. Some patients have ventricular hypertrophy without outflow obstruction while in others a pressure gradient across the outflow tract is inconstant. In most cases systolic outflow obstruction is more consistently present and often is of severe degree. It is tempting to postulate that patients with outflow obstruction have progressed further in the evolution of their disease than those with hypertrophy without obstruction, but this is not proved. Familial, congenital, and randomly occurring varieties of the disorder have been described.

The patients may have any of the symptoms of other forms of left ventricular outflow stenosis, and sudden death is often noted in the family histories of patients with the familial form. True congestive heart failure is uncommon.

The physical findings provide clues to the diagnosis. Whereas the thrill and murmur in other forms of left ventricular outflow obstruction are most prominent in the upper sternal region with characteristic transmission to the

**Figure 6**

Supravalvular aortic stenosis. A narrow constriction is evident just above the aortic valve, following left ventricular injection of contrast agent.
Two chest radiographs in two patients with idiopathic hypertrophic subaortic stenosis. Poststenotic dilatation of the aorta is not present.

Chest radiographs in two patients with idiopathic hypertrophic subaortic stenosis. Poststenotic dilatation of the aorta is not present.

the ventricle is not impeded by the muscular stenosis. A pressure gradient between the main body of the left ventricle and the outflow tract is demonstrable, and often there is a notch on the upstroke of the ventricular pressure records at the level of systemic arterial systolic pressure (fig. 8). Ventricular end-diastolic pressure is often high, indicating the effect of the marked hypertrophy upon the diastolic compliance of the ventricle.

Left atrial pressure can be recorded by transseptal puncture or an estimate obtained from pulmonary artery wedge pressure. Tall left atrial “a” waves are characteristic. The systemic arterial pressure pulse has a unique appearance with an abrupt sharp percussion wave followed by a lower tidal wave before the incisura (fig. 8). The systemic arterial pressure wave after a premature contraction is often smaller than in the preceding normal beats.58 Right heart catheterization rather consistently shows a similar, but usually much smaller, pressure gradient across the outflow tract of the right ventricle. In acute studies, ouabain, isoproterenol, and nitroglycerin have increased the outflow tract pressure gradient and injections of methoxamine and norepinephrine have abolished the gradient.59-62 It has been postulated that changes of ventricular volume, systemic vascular impedance, or systemic di-

neck, this is not the case in hypertrophic stenosis. Here the murmur is often mistaken for pure mitral insufficiency or a ventricular septal defect because of its maximal location along the lower left sternal border or at the apex and its pansystolic characteristics. A thrill may be associated. Mitral insufficiency often is present to a variable degree, because of mitral valve distortion by the hypertrophied septum. A strong left ventricular apex beat and palpable atrial contraction are consistent findings. A slow rising peripheral pulse is not characteristic, and in fact the systemic arterial pulse is quite abrupt.57 Ejection velocity by the hypertrophied ventricle is quite rapid until the outflow tract closes after the start of ejection.

Chest radiographs may show enlargement of the cardiac silhouette, which is truly remarkable in view of the decreased volume of the left ventricular cavity (fig. 7). The electrocardiogram displays unequivocal left ventricular hypertrophy.

There are remarkable hemodynamic findings at cardiac catheterization, related in part to the fact that the stenosis is not constant throughout the cardiac cycle, and also because it can vary from day to day. In our laboratory, retrograde arterial catheterization of the left ventricle is chosen for study of patients in whom this diagnosis is suspected. Entry into

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astolic pressure may be important operative variables, with increases in any of these factors tending to decrease the outflow tract gradient. The hypertrophied, poorly distensible left ventricle presents a high resistance to diastolic filling; this factor raises left atrial pressure, setting the stage for dyspnea. In addition, left ventricular end-diastolic volume is low, resulting in a low stroke volume and cardiac output.

Left ventricular angiography produces a distinct picture of the abnormality. The zone 2 or 3 cm. below the normal aortic valve is open in diastole and then narrows with systole to a conical stenosis (figs. 9 and 10). Mitral insufficiency may be demonstrated.

Surgery for this disorder remains in an investigative stage, but reports of success have appeared. Medical treatment is not very effective. In view of the undesirable physiologic effects of nitroglycerin in this disorder, it should be used with caution. Digitalis similarly may produce adverse effects, since it en-
hances the stenosis. In regard to digitalis, a difficult problem is posed by the patient with hypertrophic subaortic stenosis who also has atrial fibrillation with a rapid ventricular rate. In this situation digitalis may be required, despite other theoretical reasons to withhold it.

Summary
The purpose of this report is to summarize the syndromes of obstruction to left ventricular outflow. Emphasis is placed upon the correlations between clinical, hemodynamic, and angiographic features that permit an accurate diagnosis. An exact definition of the problem in each patient has become of great importance because of rapid advances in cardiac surgery, which now offers specific treatment for each of the syndromes described.

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