A Hemodynamic Technic for the Detection of Hypertrophic Subaortic Stenosis

By Edwin C. Brockenbrough, M.D., Eugene Braunwald, M.D., and Andrew G. Morrow, M.D.

It is now well appreciated that following a successful valvulotomy for valvular pulmonic stenosis, muscular hypertrophy in the right ventricular outflow tract may persist and be responsible for a significant resistance to right ventricular ejection.1-3 Similar muscular obstruction to the left ventricular outflow has also been noted in patients following surgical relief of discrete valvular or subvalvular stenosis. Obstruction to left ventricular ejection resulting from contraction of hypertrophied muscle in the left ventricular outflow tract, without antecedent aortic valvular or subvalvular stenosis, has recently been recognized as a distinct disease entity.4-10 Although a variety of names have been attached to this malformation, "idiopathic hypertrophic subaortic stenosis" has appeared to us to be most appropriate and the clinical, hemodynamic, and angiographic features in 14 patients studied at the National Heart Institute have been detailed elsewhere.10

In contrast to the usual forms of obstruction to left ventricular ejection in which the orifice size is fixed, in hypertrophic subaortic stenosis the obstruction in the outflow tract relaxes during diastole and narrows during ventricular systole.10 This obstruction to left ventricular outflow results from myocardial contraction and it might be anticipated that alterations in the force of contraction could modify the effective orifice size of the left ventricular outflow tract. Accordingly, it was reasoned that the more forceful ventricular contraction which accompanies the compensatory beat following a premature contraction might narrow the orifice, reduce the stroke volume and, therefore, the systemic arterial pulse pressure. In contrast, in patients with the discrete forms of obstruction to left ventricular outflow, the increased force of contraction following the compensatory pause after a premature beat is accompanied by an increase in the systemic pulse pressure. This report consists of an analysis of left ventricular pressure pulses, which was undertaken in order to throw further light on the mechanism of the obstruction to left ventricular ejection in patients with hypertrophic subaortic stenosis, as well as to search for a simple method that might be useful in identifying patients with this lesion.

Material and Methods

The catheterization data from 87 patients with aortic stenosis, studied during the last 18 months, were reviewed. Eleven patients had idiopathic hypertrophic subaortic stenosis and one patient secondary hypertrophic subaortic stenosis, these diagnoses having been established by selective left ventricular angiocardiography or at operation.10 Forty-five patients had acquired aortic valvular stenosis and 30 patients had congenital aortic stenosis of the valvular, discrete subvalvular, or supravalvular type. The diagnosis was proved at operation in all 75 patients in the latter two groups.

Left ventricular pressure measurements were obtained by the transseptal,11 transbronchial,12 and anterior percutaneous13 technics or by direct needle puncture at operation. Simultaneous left ventricular and arterial pressure tracings were analyzed and values for the peak left ventricular systolic pressure, the systemic arterial pulse pressure, and the duration of diastole were determined. The last was considered to be the time interval between the dicrotic notch and the upstroke of the subsequent beat. The average values obtained from three beats that did not follow a premature contraction were compared with the average values from three beats, each of which was the first normally conducted beat to follow the compensatory pause after a premature contraction. In two patients with idiopathic hypertrophic subaortic stenosis only a single premature ventricular contraction was available for analysis.

From the Clinic of Surgery, National Heart Institute, Bethesda, Maryland.
Simultaneous left ventricular (LV) and brachial artery (BA) pressure tracings, in a patient with acquired valvular aortic stenosis (A) and a patient with congenital, discrete subvalvular stenosis (B). An increase in the arterial pulse pressure accompanies the rise in peak left ventricular systolic pressure following the premature contractions.

Results

In all patients, the first normally conducted beat following the compensatory pause after a premature contraction was characterized by an elevation of the left ventricular systolic pressure. This increased left ventricular pressure was accompanied by a widened arterial pulse pressure in the patients with valvular and discrete subvalvular stenosis (fig. 1A, 1B). The opposite occurred, however, in the patients with idiopathic hypertrophic subaortic stenosis; the pulse pressure narrowed, in spite of a significant increase in left ventricular systolic pressure (fig. 2A, 2B). This paradoxical response of the arterial pulse pressure was characteristic of all but one of the patients with hypertrophic subaortic stenosis; in this patient only one premature ventricular contraction was recorded and the pulse pressure of the post extrasystolic beat was identical to the pulse pressure of the control beats. Figure 3 illustrates the relationship between the left ventricular systolic pressure.
and the arterial pulse pressures in normal beats as well as in those that followed premature ventricular contractions in 15 patients with the discrete forms of aortic stenosis. The results in these patients are representative of those obtained from the entire group. In a similar manner the relationship between the duration of diastole and the pulse pressure in these patients is shown in figure 4. In each instance the arterial pulse pressure varied directly with the left ventricular systolic pressure and with the duration of diastole.

Figure 5 illustrates the relationship between the left ventricular systolic pressure and the arterial pulse pressure in eight patients with hypertrophic subaortic stenosis. The relationship between the duration of diastole and the pulse pressure in these patients is shown in figure 6. The arterial pulse pressure varied inversely with the left ventricular systolic pressure and with the duration of diastole in any given patient. In the other three patients with hypertrophic subaortic stenosis the results were identical to those plotted in figures 5 and 6.

Discussion

Certain clinical and hemodynamic features have been useful in suggesting the diagnosis of hypertrophic subaortic stenosis. In contrast to the findings in patients with valvular or discrete subvalvular aortic stenosis, the murmur in these patients is most prominent along the left sternal border or at the apex. Either no thrill, or only a faint one, is present over the base of the heart and along the carotid vessels, and on roentgenographic examination there is no post-stenotic dilatation of the aorta. The arterial pulse is brisk to palpation and recordings of the arterial pressure pulses generally show a rapid upstroke in early systole, followed by a sudden decline in midsystole. While these signs may be helpful in suggesting the diagnosis, they are not specific, and the definitive preoperative diagnosis is based on the localization of the obstruction to the outflow tract of the left ventricle and the demonstration that the outflow tract narrows only during ventricular systole. In general, retrograde catheterization of the left ventricle from the aorta and selective left ventricular angiocardiography have therefore been necessary. In view of the appreciable
risk, discomfort, and expense associated with these procedures, the need for a simple, reliable diagnostic test is clear.

The longer filling period following a premature ventricular contraction results in a greater left ventricular end-diastolic volume and, in accordance with the Frank-Starling mechanism, a greater force of ventricular contraction. The difference in the arterial pressure pulse produced by this increased contractile force has permitted separation of patients with hypertrophic subaortic stenosis from those with fixed obstruction to left ventricular ejection. In patients with valvular or discrete subvalvular stenosis, the stroke volume, and therefore the arterial pulse pressure, increases along with the force of left ventricular contraction. In the absence of aortic stenosis or in the presence of only moderate obstruction, changes in the arterial systolic and pulse pressures generally reflect similar changes in left ventricular systolic pressure. As observed by Breall and Shaffer, however, in patients with severe aortic stenosis the peak arterial pressure remains relatively constant in spite of variations in left ventricular pressure. Even in such patients a longer diastolic filling period is associated with a lower diastolic pressure and a wider pulse pressure in the subsequent beat. In contrast, in patients with hypertrophic subaortic stenosis, the longer filling period following a premature ventricular contraction is followed by a smaller pulse pressure. This paradoxical decrease in pulse pressure is probably an expression of a reduction in orifice size. Since in these patients the obstruction is produced by contraction of hypertrophied muscle, it would not be surprising if the severity of the obstruction were a function of the strength of contraction. Thus, in addition to the angiocardiographic evidence for striking variation in the diameter of the outflow tract during

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**Figure 5**
The same relationship as that depicted in figure 3, in eight patients with idiopathic hypertrophic subaortic stenosis.

**Figure 6**
The same relationship as that depicted in figure 4, in eight patients with idiopathic hypertrophic subaortic stenosis.

**Figure 7**
Comparison of preoperative and postoperative pulse pressure analyses in a patient with hypertrophic subaortic stenosis secondary to a discrete subvalvular obstruction.
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various phases of the cardiac cycle, these pulse pressure changes suggest that beat-to-beat variations in the size of the outflow tract also occur.

One patient with secondary muscular hypertrophy of the left ventricular outflow tract was studied. This 16-year-old boy had a systolic pressure gradient of 120 mm. Hg across an area of discrete subvalvular stenosis. Three weeks after an operation in which a thin subaortic membrane was completely resected, a gradient of 70 mm. Hg persisted. This residual gradient had completely disappeared when he was again catheterized 18 months later, confirming the impression that it had been caused by secondary muscular hypertrophy. Analysis of his pressure pulses demonstrated that preoperatively his responses were typical of those seen in patients with “fixed” stenosis, while immediately after operation the paradoxical response characteristic of hypertrophic subaortic stenosis was present (fig. 7). It seems likely that before operation in this patient the stenotic subvalvular membrane prevented hemodynamic expression of the obstruction produced by the muscular hypertrophy. After operation, however, the persistence of muscular hypertrophy in the outflow tract resulted in dynamics identical to those observed in patients with idiopathic hypertrophic subaortic stenosis.

It is an interesting paradox that the Frank-Starling mechanism, which usually serves to increase the stroke volume when there is greater diastolic filling, appears to increase the stenosis and actually diminishes the stroke volume in patients with hypertrophic subaortic stenosis. It is possible that sustaining this stronger force of contraction for more than a few beats may be deleterious and that inotropic drugs should be used with caution in these patients.

The ultimate clinical usefulness of the hemodynamic sign described for differentiating systolic from fixed aortic stenosis will depend upon its specificity, but also, before it can be applied, pressures must be recorded after several extrasystolic beats. These were recorded in all but one patient with hypertrophic subaortic stenosis in whom simultaneous left ventricular and arterial pressures were available for analysis. If a deliberate attempt were made to record pressures during the occurrence of extrasystoles, their frequency in the course of left heart catheterization would permit the hemodynamic analysis described above to be made in almost every patient undergoing this procedure.

The observation that the pulse pressure varies inversely not only with the peak left ventricular systolic pressure but also with the duration of the preceding diastole makes it possible to distinguish hypertrophic subaortic stenosis from the discrete forms of obstruction to left ventricular outflow when only an arterial pressure pulse is available. Such recordings may be obtained during right heart catheterization, when premature ventricular contractions are frequently induced. In the presence of spontaneously occurring premature contractions, it would be possible to distinguish the two forms of obstruction by inspection of direct or indirect arterial pressure tracings obtained without simultaneous intracardiac pressures and perhaps by careful palpation of the peripheral pulse.

Summary

In the presence of valvular aortic stenosis or of discrete subvalvular stenosis, the narrowed orifice is constant and its size is not altered by changes in the force of ventricular contraction. In hypertrophic subaortic stenosis the orifice narrows during systolic contraction of the hypertrophied muscle in the left ventricular outflow tract and the orifice size therefore is a function of the force of left ventricular contraction. In 75 patients, proved to have the discrete, “fixed” type of aortic or subaortic stenosis the beats following the compensatory pause after a premature contraction were always characterized by higher left ventricular systolic pressures and larger systemic arterial pulse pressures than the normal beats, i.e., arterial pulse pressure varied directly with left ventricular systolic pressure and with the duration of diastole. In 12 patients proved to have hypertrophic subaortic stenosis, the beats following premature
contractions always exhibited lower arterial pulse pressures than did the normal beats, i.e., arterial pulse pressure varied inversely with left ventricular systolic pressure and with the duration of diastole. The distinction between these two types of obstruction to left ventricular outflow may be established with confidence by this analysis of the effects of premature ventricular contractions on either the simultaneously recorded left ventricular and arterial pressure pulses, or on the arterial pressure pulses alone. The recognition of hypertrophic subaortic stenosis is essential in formulating a rational plan of treatment for all patients with obstruction to left ventricular outflow, and the hemodynamic technic described affords a simple but reliable diagnostic approach.

Addendum
Since figures 5 and 6 were prepared for publication we have studied three additional patients with idiopathic hypertrophic subaortic stenosis. In each instance the arterial pulse pressure varied inversely with left ventricular systolic pressure and with the duration of diastole.

References
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