On the Question of Obstruction in Idiopathic Hypertrophic Subaortic Stenosis

ONLY 10 YEARS have elapsed since the concept of functional obstruction to left ventricular ejection was introduced to clinical medicine (1), and in the interim, the features of obstructive cardiomyopathy, or idiopathic hypertrophic subaortic stenosis, have become generally recognized. It seems certain that the increasingly frequent recognition of this disease has resulted largely from wider application of left heart catheterization techniques; nevertheless, while the diagnosis initially was based on hemodynamic features peculiar to this disease, it has become possible with increasing experience to recognize and assess the severity of this lesion on clinical grounds alone (2).

Work originating in a number of laboratories, recently reviewed in detail (2), has generated the hypothesis that the unique clinical and hemodynamic features of idiopathic hypertrophic subaortic stenosis, such as the sharp upstroke of the arterial pulse, the ejection quality of the systolic murmur, and the variability of the pressure differential across the left ventricular outflow tract, can be explained by invoking two basic premises: first, that systolic obstruction develops progressively in the outflow tract during the course of each cardiac contraction and, secondly, that the muscular nature of the outflow tract allows variations in the degree of obstruction during alterations in such factors as the distending pressure in the outflow tract and the inotropic state of the myocardium.

Renewed interest in the mechanism responsible for the intraventricular pressure difference in patients with idiopathic hypertrophic subaortic stenosis has been stimulated recently by Criley, Lewis, White, and Ross (3) who have supported an alternative explanation. They proposed that the peculiar hemodynamic features of idiopathic hypertrophic subaortic stenosis are not due to an obstructive phenomenon but rather to early and complete emptying of the left ventricle, the elevation of the intraventricular pressure occurring only within essentially empty portions of the left ventricular cavity; the rationale behind operations designed to relieve obstruction in idiopathic hypertrophic subaortic stenosis was therefore questioned by these investigators. This hypothesis had its origins in several observations: extensive data showing that pressure differences within the left ventricle can be induced in normal animals by interventions that empty the ventricle and increase its contractile force (for example (4, 5)); observations in patients with idiopathic hypertrophic subaortic stenosis which indicated that approximately 75% of the stroke volume can be ejected during the first half of ventricular systole (6, 7); and difficulty in defining angiographically a site of obstruction in patients with idiopathic hypertrophic subaortic stenosis (3). The possibility that cavity obliteration might be responsible for the elevated intraventricular pressure therefore offered an attractive alternative to obstruction, and a lively interchange of ideas and observations ensued.

The reasons why, in our opinion, true obstruction does occur in patients with
idiopathic hypertrophic subaortic stenosis are presented at length elsewhere (8). They consist of, in brief: the fact that a pressure gradient is recorded when the catheter tip is not positioned in obliterated portions of the ventricle but lies free just anterior to the mitral valve, in an area where elevated intraventricular pressures cannot be induced in normal animals (9); observations, using an electromagnetic flowmeter at the time of operation, which indicate that only 30% of the stroke volume is ejected before the pressure gradient develops (7); numerous descriptions at postmortem examination and at operation of focal muscular hypertrophy in the left ventricular outflow tract; the dramatic effects on the pressure gradient of surgical excision of muscle in this area; and the finding that angiographic films exposed during systole in many patients with idiopathic hypertrophic subaortic stenosis clearly reveal an area of obstruction that appears to result from apposition of the leading edges of the mitral valve leaflets against the bulging interventricular septum (8).

It has become increasingly clear that the hemodynamic consequences of idiopathic hypertrophic subaortic stenosis are complex. Thus, the intraventricular pressure gradient may vary at different cardiac catheterization studies, as well as during the course of a single study. In addition, the limitation of ventricular filling imposed by reduced diastolic compliance, as well as the effects of mitral regurgitation, are of variable importance. Further, this hemodynamic spectrum is clouded by patients who may have ventricular hypertrophy but who do not have idiopathic hypertrophic subaortic stenosis in whom it is sometimes possible to record an elevated left ventricular pressure when the catheter lies in the apex or an intratrabecular recess. In such patients, the mechanism for the increased left ventricular pressure may well be similar to that responsible for the elevated pressure during pharmacologic maneuvers in normal animals; under these circumstances, the other hemodynamic features characteristic of idiopathic hypertrophic subaortic stenosis, such as the elevated ventricular pressure in the nonobliterated area just anterior to the mitral valve and the decrease in pulse pressure after a premature contraction, are not observed (8, 9). This hemodynamic problem, and the protean manifestations of idiopathic hypertrophic subaortic stenosis per se, need not obscure the central issue, however. The question remains of whether true obstruction to left ventricular ejection exists in patients with idiopathic hypertrophic subaortic stenosis; this question obviously carries important therapeutic implications. In our opinion, the current lines of evidence strongly favor a primary role for true obstruction. As new facets of idiopathic hypertrophic subaortic stenosis are discovered and as surgical or pharmacologic approaches to therapy are developed, it may be anticipated that the questions currently surrounding this fascinating disease will have promoted more complete understanding of its unusual pathophysiology.

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REFERENCES
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The Task of the Clinician

The doctor-patient relationship is central to all treatment procedures, and the clinician, through training and experience, has learned to use it for the benefit of his patient. The mentally sick, the invalid, and the dying patient benefit as much from the human relationship as from any drugs, surgery, or other physical procedures that the physician may apply. The extraordinary combination of scientific knowledge and social skill which goes into the making of a good physician can best be summarized by an enunciation of the goals of clinical medicine, which are:

To adapt scientific information to the treatment of patients and to the prevention of disease; to control the physical environment to make this a healthier world in which to live; and to cope with the social environment through legislative, legal, and educational means.

Within this framework, the tasks of the individual clinician then are: To personally deal with patients and their particular circumstances, diseases, and problems; and to contribute to the psychological well-being of people by enabling them to grow, to develop their potentialities, and to maintain a healthy attitude towards life.—Jurgen Ruesch: Declining clinical tradition. *JAMA* 182: 110, 1962.