Nonobstructive IHSS

To the Editor:

The editorial by Pravin M. Shah on IHSS (A.K.A. HOCM, MSS, ASH, and Hypertrophic Cardiomyopathy) should serve as a stimulus to stop and reflect on the body of facts, fallacies, and fancies that have characterized descriptions of the disease since its clinical and hemodynamic discovery in the late 1950s.

In his overview of the changing concepts and current state of knowledge of the disease, Dr. Shah passed quickly over the nonobstructive concept of the disease. The justifications to examine alternatives to the obstructive concept of the disease are 1) patients without pressure gradients have severe symptoms (angina, syncope, and breathlessness), and indeed die of the disease more readily than those with gradients; 2) the left ventricle in patients with gradients empties more rapidly and more completely than the normal ventricle; 3) the pattern of early rapid outflow followed by reduced or absent late systolic ejection occurs in the absence of pressure gradients, and therefore cannot be explained by late obstruction. There are two mechanisms by which pressure gradients can be generated in the absence of obstruction: cavity obliteration and catheter entrapment.

Cavity obliteration is a phenomenon in which a ventricle creates a pressure gradient between regions encompassed by walls that are moving rapidly inward (i.e., the apex and body of the ventricle) and relatively noncontractile regions of the left ventricle. The phenomenon occurs in normal ventricles (i.e., dogs infused with isoproterenol, dogs and patients in hemorraghic or septic shock) but occurs more readily in hypertrophied ventricles (coarcted dogs and postoperative aortic stenosis patients). In the normal ventricle, less of the cavity obliterates, and the gradient may exist between apex and body of the ventricle, while ventricular hypertrophy promotes obliteration of virtually all of the ventricle up to the mitral valve, and a gradient exists between inflow and outflow portions of the ventricle.

Catheter entrapment connotes an artificial high pressure created in essentially bloodless recesses of the ventricle, and according to Wigle it can be differentiated from true obstruction by demonstrating that the inflow tract pressure is equal to aortic pressure, that blood cannot be withdrawn from the catheter during systole, and that the high left ventricular pressure often persists after the diastolic notch.

There is also a need to re-examine the specificity of ASH, SAM, and the postectopic beat phenomenon in establishing the diagnosis of this condition. ASH, as defined by the NIH criterion (septum/free wall >1.3) was seen in 14 of 30 patients with malignant hypertension studied in our laboratory. The septal thickness >1.5 cm, and septum/free wall ratio >1.5, used by Abbasi excluded 11 of the 14 "positive" patients. We have failed to find ASH in two thoroughly studied families of patients with established hypertrophic cardiomyopathy which would be statistically unlikely if the disease is always hereditary, as has been stated by the NIH investigators. We have found SAM in patients with left ventricular aneurysm, prolapsed mitral leaflet syndrome, and patients with hypertrophic cardiomyopathy without gradient. The postectopic beat phenomenon occurs in experimental hypertrophic dogs and patients with valvular aortic stenosis. To explain these coincidences as a chance association of "IHSS" and hypertension, "IHSS" and aortic stenosis, etc. is begging the question.

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References


The author replies:

To the Editor:

Drs. Criley, Blaufuss and Abbasi make some very important observations regarding the IHSS (HOCM, MSS) spectrum of disorders. At risk of repetition, it must be emphasized that patients with asymmetric myocardial hypertrophy fall into two broad categories (A) those in whom ventricular outflow (generally the left, occasionally the right) obstruction is present or provocative, and (B) those in whom no obstruction can be stimulated. There is no question that both groups can be equally symptomatic. The "obstructed" group forms nearly 80% of symptomatic