Echocardiographic Criteria in IHSS

To the Editor:

In the recent paper by Rossen and colleagues (Circulation 50: 747, 1974), the authors make potentially important observations concerning the echocardiographic features in IHSS. The description of a patient with hemodynamic IHSS and symmetric left ventricular hypertrophy casts doubt on the sensitivity of a previously suggested pathognomonic echocardiographic feature of this disease, that of asymmetric septal hypertrophy or ASH.1 However, whereas the technical quality of the echocardiogram from this patient (fig. 2) is adequate for accurate measurement of the septal thickness, the same is not true for the posterior left ventricular wall. Thus, the most anterior echo of the posterior wall structure has been labeled as endocardium despite the fact that this echo moves almost parallel with that from the pericardium. If this did represent the endocardium, there would be little or no systolic thickening of the posterior wall. A more likely explanation for this echo is failure of the ultrasound beam to strike the posterior wall perpen-

dicularly, resulting in a false impression of excessive wall thickness.1

Perhaps the most disturbing feature of figure 2 becomes apparent when one compares it with figure 4 in a more recent paper by these same authors.2 Even cursory inspection reveals that, although labeled differently, these two illustrations are the same echocardiographic complexes. In the latter illustration, however, a more posterior echo has been labeled as posterior wall endocardium, giving the tracing the characteristics of asymmetric septal hypertrophy. It is difficult to understand why these authors have chosen the same echocardiogram to illustrate symmetric left ventricular hypertrophy in one communication and asymmetric septal hypertrophy in another.

The interpretation of this echocardiogram assumes considerable importance in view of the current intensive efforts to better understand the unusual nature of the IHSS disease spectrum. Our recent finding of electrical, histochemical and histologic abnormalities in voluntary muscles of patients with IHSS3 indicates that the process is not limited to the myocardium. Moreover, the nature of these histologic abnormalities suggests that the changes might be neurogenically induced, an observation supported by the detection of ASH and similar voluntary-muscle abnormalities in patients with Friedrich’s ataxia.4 Both of these findings suggest that the myocardial involvement in this disorder represents only one aspect of a much broader disease spectrum, possibly reflecting genetically determined abnormalities of neuronal structure or function.

Studies designed to define the total spectrum of this disorder would be greatly facilitated by the availability of an easy and sensitive means for the detection of affected patients. Although not yet firmly established, the echocardiographic detection of ASH promises to be such a marker. We felt that the contention of Rossen and co-workers that IHSS may exist without ASH is not substantiated by their illustration — a conclusion supported by the author’s own use of the same echocardiogram in another communication to depict IHSS with ASH.

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LETTERS TO THE EDITOR


The authors reply:
To the Editor:

We appreciate the opportunity to reply to the thoughtful letter by Drs. Smith and Anderson. These gentlemen properly point out some aspects of IHSS. Unfortunately, the legend of figure 4 in The New England Journal of Medicine article was improperly labeled as demonstrating asymmetric septal hypertrophy. Obviously it demonstrates symmetric thickening of the septum and posterior wall in this unusual patient. This patient does demonstrate that symmetric thickening may occur in this disease process and for this reason we included the patient in our article in Circulation. Application of our own criteria published elsewhere made us realize that we had improperly labeled the endocardial surface of the left ventricular posterior wall in this early illustration. Movement of the endocardium and resultant obvious thickening of the posterior wall is at the lower limit of the values found in normal patients in our laboratory. However, the interventricular septum does not thicken during systole. For this reason we used an illustration from the same patient to demonstrate the lack of septal thickening associated with IHSS emphasizing the point of our later article. That is, though equal in thickness, the posterior wall thickened to a normal degree while the septum did not thicken at all. This leads us to believe the septum is involved with the abnormal muscle process seen in IHSS while the posterior wall may be secondarily hypertrophied.

While we apologize to the readers of Circulation and The New England Journal for this confusion, we are sure that this confusion is matched by our embarrassment in overlooking the letter "a" that appears on the legend of figure 4 in the article published in The New England Journal of Medicine. We originally had planned to include a case of asymmetric septal hypertrophy in addition to the case shown and when the illustration from the case with asymmetric septal hypertrophy was deleted, the wrong legend was applied to the figure showing symmetric septal hypertrophy.

For us this situation emphasizes the problems that can occur from an oversight in proofreading a manuscript despite the care with which we believed we were carrying out this process. We sincerely hope that this technical error will not detract significantly from the observations reported in our two papers.

Drs. Smith and his colleagues at the National Heart and Lung Institute have made an extremely valuable contribution to our understanding of hypertrophic cardiomyopathy. The basic principles pointed out by them have been confirmed in our laboratory as well as many other laboratories. However, asymmetric septal hypertrophy alone may occur as a result of right ventricular hypertrophy and symmetric left ventricular hypertrophy may be seen in the spectrum of IHSS. An extremely thick septum which does not contract is characteristic of IHSS and may be a clue to the pathophysiology of outflow tract obstruction in this condition. Our studies show that the septum is a hypocontractile rather than hypercontractile area. While we may be able to predict the degree of outflow tract obstruction by comparison of the echocardiographic "obstruction index" with the gradient measured simultaneously in a patient, the corresponding figures for outflow tract gradient and obstruction index may be slightly different from patient to patient.

We are sure the further careful studies of Dr. Henry and his colleagues as well as those of Dr. Smith and his colleagues will shed more light on the various aspects of IHSS — if all of us can avoid the kind of oversight that has partially obscured the intent of our articles.

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Prognosis in Single Vessel CAD
To the Editor:

Doctor Humphries and his colleagues have to be congratulated for an excellent attempt at defining the natural history of ischemic heart disease patients (Circulation 49: 459, 1974). Most of the data are very clearly presented and discussed. I agree with most of their conclusions, except those in relation to single vessel disease. They leave the reader with the impression that patients with single vessel disease have excellent prognosis, although they are very careful to point out that it applies only to those patients who are...
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