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. 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 perpendicularly, 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 Friedreich'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.

ELDON R. SMITH, M.D.
ROBERT N. ANDERSON, M.D.
Dalhousie University,
Halifax, Nova Scotia, Canada

References
3. Smith EB, Heffernan LP, Sangalang VE, Gaum WE, Roy DL: Skeletal muscle involvement in patients with asymmetric...
To the Editor:

England Journal of Medicine article was improperly labeled as demonstrating asymmetric septal hypertrophy. Unfortunately, the legend of figure 4 in The New England Journal of Medicine article was improperly labeled as demonstrating asymmetric septal hypertrophy.2 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.1 Application of our own criteria published elsewhere3 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 near 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 problem 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.1 2

Dr. Henry 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 hypertrophy4 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 hypococontractile 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. Ronald M. Rosser, M.D. Richard L. Popp, M.D. Stanford University Medical Center Stanford, California 94305

References
3. Popp RL, Harrison DC: Cardiac chamber size and volume: Echographic measurement of cardiac chamber dimensions, volume and ventricular function. In Ultrasound in the Diagnosis of Cardiovascular-Pulmonary Disease, edited by Jowett CR. Chicago, Year Book Medical Publishers, 1974

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
Letter: Echocardiographic criteria in IHSS.
E R Smith and R N Anderson

Circulation. 1975;52:352-353
doi: 10.1161/01.CIR.52.2.352
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1975 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/52/2/352.citation

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/