Isolated Noncompaction of the Myocardium

To the Editor:

McCrohon and colleagues present a case of isolated ventricular noncompaction (IVNC) diagnosed by magnetic resonance (MR) imaging. They suggest that MR may improve the detection rates of IVNC as, in their case, the diagnosis would have been missed if only echocardiography had been used. Cardiac ultrasound, previously performed in their case, had shown no significant abnormalities.

It is correct that this disease is not widely known, and, as a consequence of ignorance, its diagnosis is frequently missed. To facilitate its diagnosis, we have recently established 4 clear-cut echocardiographic diagnostic criteria, namely: (1) absence of coexisting cardiac abnormalities; (2) a 2-layered structure of the left ventricular wall, with the end systolic ratio of noncompacted to compacted layer; (3) finding this structure predominantly in the apical and mid-ventricular areas; and (4) blood flow directly from the ventricular cavity into the deep intertrabecular recesses as assessed by Doppler echocardiography. This study clearly demonstrated that IVNC can be accurately diagnosed by echocardiography, as we validated our results against pathological-anatomical preparations. Although MR imaging may also be helpful, no diagnostic criteria have yet been proposed. Furthermore, as is the case for echocardiography, MR imaging also necessitates experienced operators.

The MR images of the present case show multiple intertrabecular recesses. We are surprised that a skilled echocardiographer would miss the diagnosis in ultrasound, particularly if he had previous experience of such conditions. Furthermore, the corresponding left ventricular angiogram might have given an additional hint, as this has been demonstrated to be indicative in another previously published report.

We conclude that echocardiography currently remains the gold standard for the diagnosis of IVNC, at least when performed by experienced investigators.

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Response

We accept that the data thus far available on isolated ventricular noncompaction (IVNC) is derived from echo series and we welcome the echo classification. Nevertheless, in this particular case study, the echo (even on subsequent review and further acquisitions) was unable to clearly document the abnormality. As mentioned in the case report, the echo only revealed upper limit of normal wall thickness, and the ventriculogram failed to show any abnormality in the inferior wall, where most of the intertrabecular recesses reside.

The case we presented is not a florid example of IVNC and makes the point that cardiovascular magnetic resonance (CMR) may enhance the detection of more subtle forms of noncompaction that may or may not progress with time. The provision of formal diagnostic criteria using CMR will require further study, along with heightened awareness of the potential of new imaging techniques to define earlier and/or more subtle forms of pathology.

There is little reason to expect that CMR diagnostic criteria will differ significantly from echo criteria, although as this example suggests, CMR techniques may provide the ability to detect a broader spectrum and more subtle variants of noncompaction.

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