Atroventricular discordance

Sir,
As cross sectional echocardiography visualises dynamic in vivo anatomy with great accuracy, it is no surprise that conclusions reported in the study by Sutherland et al (Br Heart J 1983; 50: 8–20), which concern the identification of ventricular morphology by echocardiographic description of the associated atrioventricular valve and subvalve apparatus, are in broad agreement with those of earlier authors including ourselves.1 Nevertheless, their failure to demonstrate separate levels of connexion of intraventricular valve leaflets into the central fibrous body (the crux) of the heart does not invalidate this criterion for morphological definition but rather reflects the population which they selected. This clearly included patients with inlet ventricular septal defects large enough to disturb the inflow portion of the ventricle in the region of the crux and thus to disturb the area of normal leaflet connexion. Patients in our study, which was not restricted to patients with so-called congenitally corrected transposition as misstated by these authors, were included specifically to look for the markers of ventricular morphology undisturbed by associated lesions such as large ventricular septal defects. As we have stated, criteria other than valve leaflet insertion into the crux are obviously useful if leaflet anatomy is ambiguous.

The authors also use their own findings2 in support of the claim that equiplanar insertion of the leaflets at the level of the crux is a pathognomonic feature of an inlet ventricular septal defect as it is in all atrioventricular defects. In our preliminary report of experience in patients with a single ventricle,3 this finding occurred in all patients with a double inlet connexion to the main chamber. Although this anatomy could be regarded as lying within the spectrum of inflow defects the conditions are clearly morphologically, functionally, and prognostically distinct.

Finally, we believe that the authors should have made it quite clear that the anatomical specimens used to illustrate the stop frame images were not necessarily necropsy specimens from that echocardiographic study, as implied in the title, but were “drawn from the cardiopathologic collection” by methods not stated. Should not their methods include details of how the two were matched? This practice undoubtedly provides convenient illustrations for general lectures, teaching, etc; however, it is, we believe, inappropriate in a journal of this merit.

R A Foale, Jane Somerville, A F Rickards, National Heart Hospital, London W1M 8BA.

References
2 Smallhorn JF, Sutherland GR, Anderson RH, Macartney FJ. Cross-sectional echocardiographic assessment of conditions with atrioventricular valve leaflets attached to the atrial septum at the same level. Br Heart J 1982; 48: 331–41.

This letter was shown to the authors, Dr Sutherland and colleagues, who reply as follows:

Sir,
We thank Drs Foale, Somerville, and Rickards for their letter. We have to admit to some confusion as to the source of their criticisms. They suggest, firstly, that we questioned the value of the reversed offsetting of the atrioventricular valves in the diagnosis of atrioventricular discordance. This is not so. On page 19 (Br Heart Journal 1983; 50: 8–20) we clearly stated “when present it was diagnostic . . . .” Our point was that in the majority of patients with atrioventricular discordance the association of an inlet perimembranous ventricular septal defect results in the valves being inserted at a common level into the central fibrous body. Thus it is essential to turn to other diagnostic criteria. Since the majority of patients with congenitally corrected transposition do have such defects it is specious to argue that this feature disturbs the anatomy. It is precisely this anatomy which is the norm in an unselected population presenting to the paediatric cardiologist. Secondly, they argue that all their patients did not have congenitally corrected
transposition. Yet all the patients that they studied with atrioventricular discordance also had ventriculo-arterial discordance. As far as we are aware (and we are supported by Metcalfe and Somerville) this combination is the one usually referred to as congenitally corrected transposition. It is quite clear that our comment was made specifically in the context of atrioventricular discordance. Thirdly, they accuse us of raising the equiplanar insertion of the atrioventricular valve leaflet to the status of a pathognomonic sign of an inlet ventricular septal defect. We do not know how they reached this conclusion. This statement is not to be found anywhere in our writings. Indeed, in the introduction to the paper they refer to, we stated—alluding to the valve attachments being at the same level—"This finding is of course characteristic of a double inlet atrioventricular connexion, but in this case its importance is trivial compared to the other abnormalities present." There is another equally important reason why we would not make this statement. The offsetting of the atrioventricular valve leaflets and the presence of a muscle bar projecting downwards from the atrioventricular junction to roof a defect clearly differentiates an inlet muscular defect from a perimembranous inlet defect.

We are grateful to take this opportunity to clarify our figure legends in the four instances in which pathological specimens were shown with the corresponding echocardiograms. As these stand in the text they could lead to some confusion. In Fig. 3 and Fig. 4 the pathological specimens were not from the patient whose echocardiogram is shown but were sections from similar patients drawn from the cardiological-pathological collections of the three centres concerned. In Fig. 9 and Fig. 10 the pathological specimens were from the patient whose echocardiogram is shown.

Finally we wish to defend the principle of using corresponding anatomical specimens to illustrate echocardiographic stop frame images. This has now become a widely adopted form of presentation, and such correlative papers have appeared in the majority of leading cardiological journals. Indeed a large number of such papers have undergone review and publication in the British Heart Journal. We feel strongly that such a correlative presentation leads to a greater appreciation of the information which can be derived from echocardiographic images obtained from patients with complex heart disease. We would further suggest that Foale et al are in the minority in voicing criticisms of what is now a widely accepted form of correlative presentation.

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Reference

Myocardial disorganisation in hypertrophic cardiomyopathy

Sir,

"Whatever is only almost true is quite false, and among the most dangerous of errors, because being so near truth it is the most likely to lead astray."

I was reminded of the words of Henry Ward Beecher when reading Dr Maron's editorial in the July issue of this journal (1983;50:1–3). In his rebuttal of our work Dr Maron makes errors not only in misunderstanding what we had to say but also in pointing in the wrong direction when complaining that others have generated a controversy regarding the role of myocardial disorganisation (or disarray). Indeed, Dr Maron does not hesitate to extrapolate from his interpretation of the works of others, including the paper by Becker and Caruso, that those investigators lack any meaningful experience with hypertrophic cardiomyopathy.

Regarding the issue at stake it may be necessary to briefly reiterate the most salient points. Firstly, the rather loose use of terms such as "characteristic," "typical," and "specific," when referring to myocardial disorganisation in cases with hypertrophic cardiomyopathy, has led to the misconception that this particular histological feature is a useful diagnostic