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Editorial Comment

How do we determine atrial arrangement?

Robert H. Anderson

Cardiac Unit, Institute of Child Health, London, UK

In this issue of the Journal, Machado-Atias and his colleagues present a detailed and careful analysis of the arrangement of the heart and thoraco-abdominal organs in a large series of over one thousand autopsied patients.[1] Their findings bear directly upon the controversy in which I was recently involved at the 3rd World Congress of Pediatric Cardiology and Cardiac Surgery. The controversy sessions took place on the last day of the Symposium. My debate, carried out with Dr Steve Sanders from Duke University, was concerned with the proposition "There is no such thing as Isomerism in the Heart". It attracted a relatively limited audience. This was a pity, since the scientific arguments presented from both sides, as viewed from my obviously biased stance, were formidable. Many of the points we discussed are highlighted in the excellent results and discussion presented here by the team from Caracas.

As has been argued strongly in recent years, it is my own belief that the evidence supporting the existence of cardiac isomerism is overwhelming.[2–4] Within the human heart, however, specifically when it is congenitally malformed, my diagnosis of the presence of isomerism is based exclusively on the arrangement of the atrial appendages. This point needs to be born in mind by those who read the extensive study of the Venezuelan team.[1] Machado-Atias and his colleagues make many points with which I agree most strongly. They make one suggestion which is particularly important.

This bears on one of the criticisms made by Steve Sanders of my own approach to isomerism. In his presentation in Toronto, Steve pointed out that the term "isomerism" was basically derived from the physical sciences. He argued that chemical isomers were exact mirror-images of each other. In the field of biological science, it is very unusual to find exact mirror-imagery. Even in the situation of the limbs, which are essentially mirror-imaged relative to each other, features such as hypertrophy or atrophy of one limb can obscure the basic situation. Considering the heart and thoraco-abdominal organs with this in mind, specifically in the situation of visceral heterotaxy, Machado-Atias and colleagues suggest that the more appropriate term would be "isomorphism". This is the noun they use throughout their description, and all the logical arguments support its use. Whether this means that it will meet with widespread approbation, and enter the terminological lexicon, will only be established on the basis of future usage.

Whilst supporting Machado-Atias and his colleagues in the use of isomorphism, however, there is one area in which we continue to disagree. The team from Caracas have shown great patience with me during the process of review, but we have still to reach final agreement on this one outstanding point. Machado-Atias and his colleagues argue that, in three of their cases with heterotaxy, there is discordance between arrangement of the atriums and the atrial appendages. I find this an impossible statement since, for me, the diagnosis of atrial arrangement is based entirely upon the recognition of the morphology of the appendage. This does not depend upon the shape of the appendage, nor on the presence or absence of the terminal crest, nor on their size. All of these features can be distorted by extraneous agencies. Our definition of the anatomic nature of the appendages is based exclusively on the extent of the pectinate muscles lining the wall of each appendage relative to the atrioventricular junction. In the normal heart, these pectinate muscles in the morphologically right appendage extend all round the vestibule of the tricuspid valve and reach to the crux of the heart. The morphologically left appendage has its pectinate muscles confined within

Correspondence to: Robert H Anderson, Cardiac Unit, Institute of Child Health University College, London, UK. Tel: +44 20 7905 2295; Fax: +44 20 7905 2324; E-mail: r.anderson@ich.ucl.ac.uk

its tubular lumen, with the smooth vestibule of the atrioventricular junction being confluent with the venous component. In a detailed analysis of over 200 hearts with visceral heterotaxy, we showed that these features permitted all atrial appendages to be placed into groups of right and left isomorphism. Thus, in our extensive series, no hearts showed discordance between the arrangement of the appendage and the rest of the atrial chamber simply because we diagnosed the arrangement exclusively according to the morphology of the appendages.[2] In the discussion which took place in Toronto, it became clear that Dr Sanders was not using the same criterions to define isomerism. In his opinion, atrial "situs" was better defined on the basis of other features, such as venoatrial connections. This approach, however, breaks the fundamental rule established by Van Praagh and his colleagues,[5] working from the original studies of Lev,[6] and called the "morphological method". This principle states that one variable feature should not be used to define another feature which is itself variable. Paraphrased, it means that the most constant component of any structure should be used for the purposes of definition. Within the atrial chambers, the appendages are far more constant than the other features, such as venoatrial connections, septal structure, or vestibular morphology. Thus, it is the appendages, and specifically their morphology as judged according to the extent of the pectinate muscles, which should be used to define atrial arrangement.

Because of this reliance on the morphological method, I still have problems with this one aspect of the manuscript published by Machado-Atias and his colleagues. The problem is encapsulated in the heart shown in their Figure 4 (p. 547). As can be seen, the lungs and bronchial tree from this patient are unequivocally right isomorphic. Within the common atrium, however, the group from Caracas suggest that the left-sided appendage is of left morphology. It certainly has a narrow neck, and is unequivocally smaller than its right-sided partner. But examination of Figure 4E shows equally clearly that the pectinate muscles extend to the crux on both sides. To my eyes, this heart shows isomorphism of the right atrial appendages, and thus there is no discordancy between arrangement of the atriums and their appendages. It is my belief that analysis in this fashion will remove the other discordances which are suggested to exist between the atrial chambers and their appendages.

Taken overall, nonetheless, the study by Machado-Atias and his colleagues[1] is almost entirely in keeping with my own approach to sequential segmental analysis. As they rightly point out "[t]he association of cardiovascular malformations; be it right or left isomorphism, cannot be used as an absolute reference for classification of 'situs' ". They conclude that "[t]he precise determination of cardiac arrangement should be based only on the atriums, and in particular on the internal morphology of their appendages". I would rephrase this sentence to end "specifically on the internal morphology of their appendages". In all other respects, we are in total agreement.

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