Cardiol Young 2002; 12: 421–423

ISSN 1047-9511

From the Editor-in-Chief

Together with this issue of the journal, we publish an important supplement sponsored by the Association for European Paediatric Cardiology. It contains the first updating of the European Paediatric Cardiac Code, published initially in the supplement which accompanied our last issue of 1999.[1] Since this initial publication, the code has been used increasingly within Europe, and its use is gradually spreading world-wide. The increasing experience has brought to life some deficiencies in the initial listings, and these, along with other suggestions, have been discussed by the Coding Committee of the Association and, where considered appropriate, incorporated into the new version. Perhaps more importantly, the short list of the Code has now been cross-mapped to the codification that was produced almost simultaneously by the Society of Thoracic Surgeons, working with the European Association of Cardiothoracic Surgery.[2] It was unfortunate, in many ways, that these two systems were produced separately and almost simultaneously, the more so since the European Association of Cardiothoracic Surgery also collaborated directly with the Association for European Paediatric Cardiology in the production of the European Paediatric Code. Despite obvious differences, the two systems have much more in common. It was agreed at the meeting held at the World Congress in Toronto, and summarised in these pages,[3] that the first step in unifying them would be to cross-map their short lists. This has now been achieved. This step, in itself, represents a major step in the move towards, eventually, producing a code and nomenclature which will achieve universal approval. As with the initial publication of the code,[1] our supplement also contains explanatory articles reporting the activities of the Coding Committee,[4] the changes made to the Long List,[5] and a brief explanation of the steps involved in cross-mapping.[6] The explanations relating to the Long List are published only in the Supplement. We have deemed the report from the Coding Committee sufficiently importance to be reproduced also within the body of the Journal itself. This article is then accompanied within the Journal by a report describing the background and progress made by the International Nomenclature Committee. This article is authored by the Executive group of the international committee, this group itself having coordinated the majority of the cross-mapping of the lists.[7]

The time and energy needed to cross-map these lists should not be underestimated. We all owe a huge debt, therefore, to Rodney Franklin, Marie Béland, Christo Tchernenkov, and Jeff Jacobs. They have worked over and above the call of duty to link together the two systems. In their article,[7] they give an account of the steps involved in moving towards the current state of rapprochement between the approaches of the groups responsible for producing, on the one hand, the European code and, on the other hand, the surgical system. Attempts to achieve a universally acceptable nomenclature and system of coding for congenital cardiac malformations are far from new. Lodewyk Van Mierop, in the late 1970s, expended much energy in seeking to unify the approaches extant at that time, and came within a hair's breadth of success.[8] We have continued to advance since that time, and the cross-mapping of the two short lists now represents solid and demonstrable progress. It is also encouraging to know that the team involved with cross-mapping is becoming much more representative of paediatric cardiology throughout the World. The structure of the committee responsible for the developments is also becoming much more transparent and democratic, as described in the article by Béland and her colleagues.[7] We should thank all those who attended the recent meeting in Montreal for giving up their valuable time to move the process forward. We are also indebted to our Canadian colleagues, who attracted a significant amount of sponsorship to support the meeting. Although significant progress has been made, there is a huge amount still to be done if we are to meet the goal of unifying the long lists by the time of the next World Congress, scheduled to be held in Buenos Aires in 2005. As viewed from my chair, time slips by with ever increasing speed.

It is no secret that I have great personal interest in the production of the ideal system for describing the various congenital malformations of the heart, as does my good friend and colleague, Richard Van Praagh (Fig. 1). Béland and her associates comment, in their article,[7] on our own contributions to the summit held in Toronto. It has been the cogent criticisms of Van Praagh and his co-workers which have led to the refinements of our own approach to nomenclature, in particular their emphasis of the principle of analysis which they dubbed the "Morphological Method".[9] At an early stage of our investigations, I was greatly

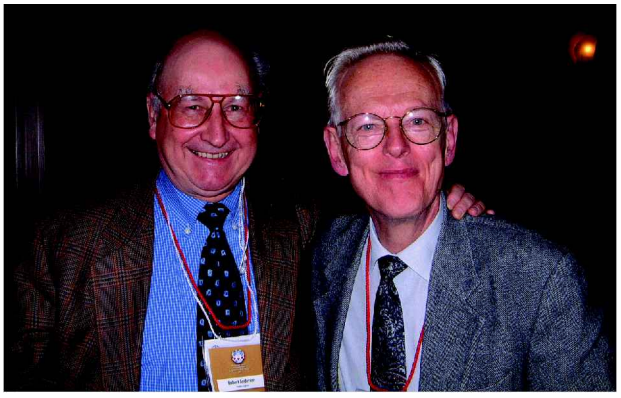

Figure 1.
Richard Van Praagh photographed with the Editor-in-Chief at the Toronto summit, May, 2001.

impressed by a comment made by the English philosopher, A.J. Ayer. Writing in the preface of his book "Language, Truth, and Logic",[10] Ayer commented how his critics had done much to fine-tune his concepts, and to improve his writings, even though they disagreed with many of his arguments. So it has proved with the criticisms of our own early works by Van Praagh and his colleagues. For myself, I hope that such constructive criticism will continue to flourish as we move towards the construction of the International "super-tree".[7] In my opinion, if we are to succeed, we need to minimise semantic arguments, and concentrate on areas of true scientific disagreement. Such areas unequivocally continue to exist, and need to be identified if we are to reach consensus. The key will be to identify the phenotypes of contentious lesions, and then agree how best to describe them. For example, disagreements continue with regard to the ventricular septal defect of "atrioventricular canal type". So as to debate this issue in constructive fashion, we need to establish the battleground. I have no problems in this respect. In our opinion,[11] the phenotype of the "atrioventricular canal malformation" is a common atrioventricular junction, but with the junction shared more or less equally between the ventricles (Fig. 2). On this basis, neither straddling tricuspid valve (Fig. 3), nor the so-called "isolated cleft of the mitral valve" (Fig. 4), can be considered malformations of this type, since they possess separate right and left atrioventricular junctions. I am sure that those who hold contrary views would produce different opinions. Only when the alternative definition of the phenotype has been established, however, can we determine whether differences of opinion reflect the underlying anatomy, or simply the words used to describe the morphologic arrangements. Many areas will need to be considered in this fashion, but it is my belief that

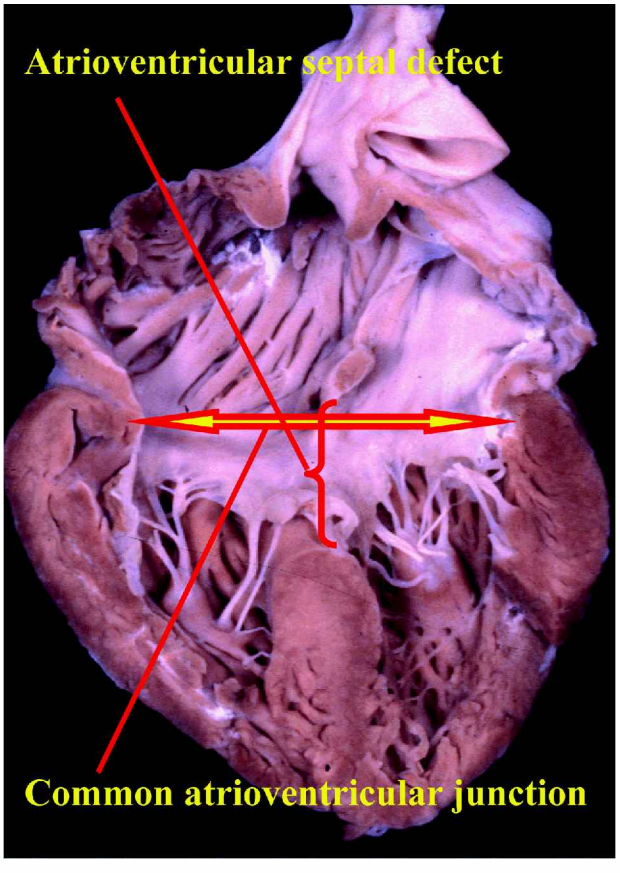

Figure 2.
As shown in this illustration, the essential feature of the heart which we prefer to describe as an "atrioventricular septal defect", but which others call an "atrioventricular canal malformation", is the presence of a common atrioventricular junction.

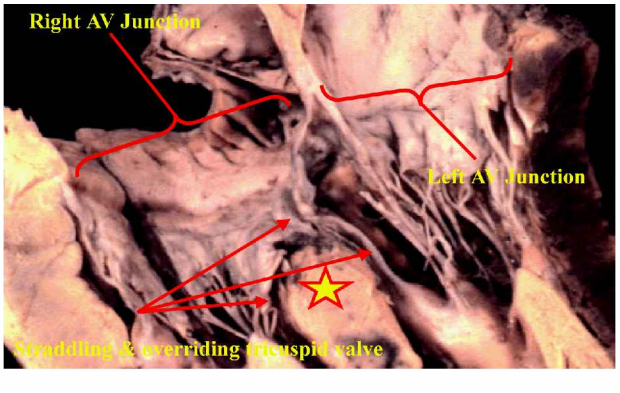

Figure 3.
This heart has overriding of the crest of the muscular ventricular septum (☆) by the right atrioventricular (AV) junction, along with straddling of the tension apparatus of the tricuspid valve. Some describe the interventricular communication seen in this setting as a ventricular septal defect of "atrioventricular canal type". As can be seen, however, the heart itself is phenotypically different from the entity shown in Figure 2 because of the presence of separate right and left atrioventricular junctions.

https://doi.org/10.1017/S1047951102000719 Published online by Cambridge University Press

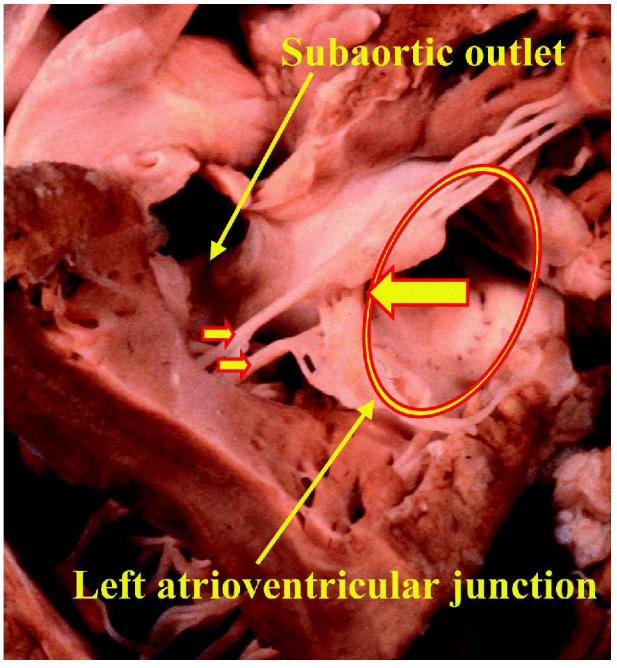

Figure 4.
In this heart, there is a cleft in the aortic (anterior) leaflet of the mitral valve (large arrow). The edges of the cleft are tethered to the septal crest by tendinous cords (small arrows) which cross the subaortic outflow tract. Some argue that this entity is also an "atrioventricular canal malformation". As can again be seen, however, it differs phenotypically from the heart shown in Figure 2 because it possesses a discrete left atrioventricular junction.

agreement on the phenotypic patterns will be the key to success. As always, the columns of our journal, and our website (www.greenwich-medical.co.uk), are open to all who may wish to contribute to the debate.

Robert H. Anderson
Editor-in-Chief

References

1. Franklin RCG, Anderson RH, Daniëls O, Elliott M, Gewillig MHML, Ghisla R, Krogman ON, Ulmer HE, Stocker FP. The European Paediatric Code. Cardiol Young 2000; 10 (Suppl 1): 1–146.
2. Mavroudis C, Jacobs JP, eds. Congenital Heart Surgery Nomenclature and Database Project. Ann Thorac Surg 2002; 69 (Suppl): S1–S372.
3. Anderson RH. From the Editor-in-chief. Cardiol Young 2001; 11: 367–368.
4. Franklin RCG, Anderson RH, Daniëls O, et al. The European Paediatric Cardiac Code – the first revision. Report of the Coding Committee of the Association for European Paediatric Cardiology. Cardiol Young 2002; 12: 1–8.
5. Franklin RCG. The European Paediatric Cardiac Code Long List: structure and function – the first revision. Cardiol Young 2002; 12: 9–16.
6. Franklin RCG, Jacobs JP, Tchervenkov CI, Beland MJ. Bidirectional crossmap of the Short Lists of the European Paediatric Cardiac Code and the International Congenital Heart Surgery Nomenclature and Database Project. Cardiol Young 2002; 12 (Suppl 2): 17–21.
7. Béland MJ, Jacobs JP, Tchervenkov CI, Franklin RCG. First report from the Executive of the International Working Group for mapping of nomenclatures for paediatric and congenital cardiac disease. Cardiol Young 2002; 12: 425–430.
8. Van Mierop, LHS: diagnostic code for congenital heart disease. Pediatr Cardiol 1984; 5: 331–362.
9. Van Praagh R, David I, Wright GB, et al. Large RV plus small LV is not single RV. Circulation 1980; 61: 1057–1058.
10. Ayer AJ. Language, Truth and Logic. London: Gollancz, 1936.
11. Anderson RH, Ho SY, Falcao S, Daliento L, Rigby ML. The diagnostic features of atrioventricular septal defect with common atrioventricular junction. Cardiol Young 1998; 8: 33–49.

https://doi.org/10.1017/S1047951102000719 Published online by Cambridge University Press