Cardiology in the Young

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Do we need to change names to better understand hearts with deficient ventricular septation?

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Editorial

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In this edition of Cardiology in the Young, Anderson et al. try to rejuvenate the long-standing discussion about the nature of the "ventricular septal defect" in hearts with double-outlet right ventricle, by introducing the new concepts of aorto-right ventricular and aorto-left ventricular communications.[1] The authors define the aorto-right ventricular communication as the area closed by the surgeon, not only in double-outlet right ventricle with subaortic "ventricular septal defect" but also in tetralogy of Fallot, outlet malalignment defects, and perimembranous central defects. They distinguish it from the aorto-left ventricular communication that represents the exit of the left ventricle in hearts with double-outlet right ventricle. According to the authors, using these two concepts would avoid the confusion about the signification of the term "ventricular septal defect."

The authors state that the concepts advocated are based on the developmental process of normal ventricular septation, the steps that have been very nicely demonstrated in one of their previous articles.[2] In this article, they show how the primary interventricular communication is remodelled into a secondary, then a tertiary interventricular communication, which is ultimately closed by the membranous part of the ventricular septum, derived from the tubercles of the atrioventricular cushions.[2]

The authors subsequently establish a parallel between the "perimembranous defects" encountered in three situations: hearts with concordant ventriculo-arterial connections, hearts with a malaligned outlet septum, and hearts with double-outlet right ventricle with subaortic ventricular septal defect. The rationale is that in all these defects, the area that will be closed by the surgeon has "comparable anatomical borders," namely the fibrous continuity between the tricuspid and mitral valve at the infero-posterior rim of the defect, and the "muscular outlet septum" as the lateral or superior border of the defect.

This new approach leads to some degree of confusion due to several of the inconsistencies generated by the amalgam proposed by the authors. Particularly perplexing is the extension by the authors of the concept of aorto-right ventricular communication to hearts with perimembranous central defects. This confusion is increased by the fact that the authors rename such defects, a rather strange way, "perimembranous defects in hearts with concordant ventriculo-arterial connections" or, even more surprisingly, "real perimembranous defects."

Indeed, the use of the term "perimembranous" for naming three different types of defects is particularly disconcerting and contradicts one of the authors' early premises. The authors rightly assess in their introduction that "the use of the same term to account for different entities potentially is the road to disaster," referring to the use of the term "ventricular septal defect" for naming the channels that permit potential shunting between the ventricles. This concern can be readily applied to the authors' use of the term "perimembranous." It is this sort of confusion that the International Society for Nomenclature of Paediatric and Congenital Heart Disease has tried to avoid with the publication of the IPCCC ICD11 Nomenclature,[3] which includes the classification of ventricular septal defects published by Lopez et al., on behalf of the International Society for Nomenclature of Paediatric and Congenital Heart Disease, in 2018 (4). This classification encompasses four categories of ventricular septal defects, according to their location on the right ventricular septal surface (trabecular muscular, inlet, outlet, perimembranous central), but also taking into account the nature of the inferior rim of the defect and the structure of the outlet septum (as modifiers), especially for the outlet defects.[4] Although this classification was not initially designed to be embryology-based, these four types of defects correspond to the evolution of the interventricular communication at the successive stages of cardiac development.[2] The classification generated by the International Society for Nomenclature of Paediatric and Congenital Heart Disease in fact provides a clear description of the defects that the authors are attempting to re-categorize in their most recent publication.

Using the same term "perimembranous" to name three anatomically different entities, or phenotypes, in a malformed heart, may therefore appear as a step backwards relative to the IPCCC ICD11 Nomenclature and to the goal pursued by the International Society for Nomenclature of Paediatric and Congenital Heart Disease. The authors claim that

"perimembranous defects in hearts with concordant ventriculo-arterial connections," which they call "real perimembranous defects" and correspond to the perimembranous central defects in IPCCC ICD11 Nomenclature, are comparable, being an "aorto-right ventricular communication," to the outlet perimembranous defects with anterior malalignment of the outlet septum, as found either in isolation or in hearts with tetralogy of Fallot. However, when one refers to embryology, the latter corresponds to the persistence of the secondary interventricular foramen, while the former corresponds to the persistence of the tertiary interventricular foramen.[2] Therefore, the term "aorto-right ventricular communication" is difficult to apply to perimembranous central defects, in which the aorta is entirely above the left ventricle. In all other defects that are part of the authors' arguments, (outlet malalignment defects, tetralogy of Fallot, and double-outlet right ventricle), the aorta overrides the crest of the muscular septum. From the anatomic point of view, there are fundamental differences in geographic location and position of the aorta in these defects that are entirely explained by cardiac development.[2]

An ambiguous consequence of the authors' new proposed classification of replacing the term "perimembranous central defects" with "perimembranous defects in hearts with concordant ventriculo-arterial connections" implies that ventriculo-arterial connections are not concordant in outlet malalignment defects. This is obviously not the case, except for the outlet subaortic defects in the setting of double-outlet right ventricular type of ventriculo-arterial connections.

Another flaw in this article is that the structure defined by the authors as the muscular outlet septum seems to vary according to the type of defect. Still, by reference to normal cardiac development, the outlet septum is formed by the fusion of the proximal part of the outflow tract cushions and thus separates the subarterial outflow tracts, and the aortic and pulmonary valve when these valves are patent. It is the fusion of the outlet cushions with the crest of the ventricular septum, between the two limbs of the septomarginal trabeculation, that closes the secondary interventricular communication. The final step of ventricular septation, after completion of wedging of the aortic valve between the atrioventricular valves, is the closure of the tertiary interventricular communication by the membranous septum. The authors affirm that there is no outlet septum in the normal heart. This is certainly the case since most or all of it disappears during aortic wedging, but then, how could the superior margin of a "real perimembranous defect" be formed by the muscular outlet septum, which according to Figure 7 of the article, would then be part of the posterior limb of the septomarginal trabeculation? This is no more than an embryological, and therefore anatomic, impossibility.

Finally, the ambiguities in the definition of tetralogy of Fallot regarding the degree of aortic override, coupled with the fact that the interventricular communication is always localised between the two limbs of the septal band in isolated outlet defect with anteriorly malaligned outlet septum, in tetralogy of Fallot, and in hearts with double-outlet right ventricle with subaortic, subpulmonary or doubly committed and juxta-arterial defects,[5] confirm that the use of eponyms should be discouraged. Instead, an accurate description of all parts of the cardiac phenotype, in other terms of the building blocks of the heart and their arrangement, should be favoured.

Harmonisation of the terms used to describe the various phenotypes of congenital cardiac diseases has been recently achieved.[3] The aim of the International Society for Nomenclature of Paediatric and Congenital Heart Disease and the IPCCC ICD11 Nomenclature is that all people involved in care of those patients, wherever in the world they live, may be able to speak the same language, in order to facilitate the communication between them, and ultimately to better manage the patients. This of course does not mean that the nomenclature cannot evolve, nor that the complexity of CHDs must be minimised. However, this complexity does not need to be increased. Changing the nomenclature recently established, after years of discussions by a panel of worldwide experts, should require a thorough and thoughtful process of collegial debate, based on scientific morphologic studies.

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