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Commentaries

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Divided left atrium with totally anomalous drainage of normally connected pulmonary veins

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Abstract

In the December 2021 issue of ***Cardiology in the Young***, Hubrechts and colleagues, from Brussels and Leuven in Belgium, describe their experience in which the pulmonary veins were normally connected to the morphologically left atrium. By virtue of the presence of a shelf dividing the morphologically left atrium, however, the venous return was to the morphologically right atrium, with no evidence of formation of the superior interatrial fold, meaning that there was no obstruction of flow into the systemic venous circulation. The question posed by the Belgian authors is whether the shelf dividing the morphologically left atrium is a deviated primary atrial septum, as the arrangement has previously been interpreted. As they discuss, it is currently impossible to arbitrate this conundrum. In our commentary, we discuss the background to the dilemma. We point out that, as yet, it is not possible to code accurately this congenital cardiac malformation within The International Paediatric and Congenital Cardiac Code (IPCCC), nor within the newly produced 11th Revision of the International Classification of Diseases (ICD-11).

Commentary

The article that is the subject of this commentary[1] has undergone a long gestation. It started life as a brief report. Both the authors and ourselves, however, are convinced that it is much improved in its current state as a review. The authors are to be commended for the patience they showed during the process of peer and editorial scrutiny. By the same token, we are indebted to the authors for the insight we have gained during our exchanges as authors, editors, and referees. The end result serves as an example, at least in our eyes, of the advantages that can accrue when peer review works properly. More importantly, the authors have identified a phenotypic entity that, again at least to our eyes, lacks either a current definition or code within the newly produced 11th Revision of the International Classification of Diseases (ICD-11).[2–4] A potential code does exist in the current version of The International Paediatric and Congenital Cardiac Code (IPCCC) [https://ipccc.net/], but that code is not entirely accurate.

This fact is important since, in the concluding sentence of their review, the authors argue that "correct diagnosis is important to facilitate surgical repair and to predict prognosis." This statement is unequivocally true. But how are others to be able to share their own experience and help to predict prognosis, if they are unable accurately to code the phenotypic entity under discussion? This conundrum poses the major question. How are we properly to describe the lesion encountered by the investigators from Brussels and Leuven?[1] The title of their review points to the potential problem. Was there "leftward deviation of the primary septum," or was the problem a "dividing left atrial shelf"? The need for the question comes from the ongoing debate regarding the origin of the partition that produces the problem in the first place. As the authors state, "leftward prolapse of the primary septum is considered as the deviation of the superior margin of the primary atrial septum, dividing the left atrium." But as they also correctly conclude "there is currently no evidence to substantiate that the described partition is initially a deviated primary septum rather than a dividing atrial shelf." There lies the rub. Since we do not know for certain the nature of the shelf, is there any justification for continuing to presume that the lesion was initially the flap valve of the oval foramen, as is frequently the case?[5–8] Is it not better to opt for certainty, and to describe the lesion on the basis of the feature on which all agree? Thus, irrespective of the origin of the partition, it no longer subserves a septal function but instead divides the morphologically left atrium.

This fact is then itself pertinent, since although the entity has frequently been reported in the literature on the basis of deviation of the primary atrial septum, it is also possible to recognise the same entity illustrated as a variant of divided left atrium.[9] Approaching the lesion on the basis of

division of the left atrium, furthermore, focuses attention on the main physiological feature of the lesion. It produces exactly the same effect as do the other variants of divided left atrium.[10] Its surgical repair, however, is subtly different. Not only must the surgeon remove the obstruction to left ventricular inlet, he or she must also septate the atrial chambers such that the pulmonary venous return is able to pass through the mitral valve. In this regard, the Belgian authors are incorrect when they suggest that the entity "may be incorrectly diagnosed as partial or totally anomalous pulmonary venous return." On the contrary, in the absence of any effective atrial septation, and in the presence of a dividing left atrial shelf, the entirety of the pulmonary venous return has no alternative other than to enter the systemic circulation. And the lesion has also been illustrated as a variant of totally anomalous pulmonary venous connection.[11]

The authors are correct, nonetheless, when they emphasise that the lesion is not one of anomalous pulmonary venous connection. The pulmonary veins are unequivocally connected in anatomically normal fashion to the morphologically left atrium. It is because of the effective absence of not only the flap valve of the oval foramen but also the superior interatrial fold, that there is totally anomalous pulmonary venous drainage. Recognition of this fact emphasises the need to distinguish between pulmonary venous connections and pulmonary venous drainage. It is well accepted that the pulmonary veins can drain in totally anomalous fashion whilst being normally connected in the setting of the channel known as the levoatrial cardinal vein. The fact that the vein is neither "levo," "atrial," nor "cardinal" does not detract from its presence in resulting in anomalous drainage of normally connected pulmonary veins. Exactly the same situation is created by the presence of the abnormal left atrial partition that exists when there is absence of the superior interatrial fold, and the flap valve of the oval foramen is either entirely absent, or else is itself malpositioned so as to produce the obstructing shelf.[1]

Hubrechts and her colleagues, furthermore, have correctly summarised the situation when stating that, at present, we do not know whether the dividing shelf is the malpositioned primary atrial septum.[1] We do know, however, that malposition of the primary septum can itself take several guises, as was revealed by the investigation of Cohen and her colleagues.[6] It is insufficient, therefore, simply to describe the phenotypic entity on the basis of malposition of the primary septum, the more so since, as discussed, we cannot be sure that the partition is the primary septum. That is why, in our opinion, the phenotypic entity is best described as division of the left atrium in the setting of totally anomalous pulmonary venous drainage. We should point once more to our use of "drainage," and not "connection," since the pulmonary veins are normally connected. When described in our suggested fashion, the arrangement can be listed as a "daughter" code of divided left atrium, or "cor triatriatum sinister," in the newly produced 11th Revision of the International Classification of Diseases (ICD-11).[2–4] (In the typical heart with divided left atrium, an obliquely orientated fibromuscular partition divides the morphologically left atrium into a compartment connected to the pulmonary veins, and a second component in communication with the atrial appendage and the mitral valvar vestibule. The two components of the divided left atrium are therefore named the pulmonary venous component and the vestibular component.)

Totally or partially anomalous pulmonary venous connection, of course, has long been recognised as one of the associated lesions to be found when the left atrium is divided.[9] But, to the best of our knowledge, totally anomalous pulmonary venous drainage, as described in the current setting, has yet to be considered as part of the spectrum of the divided left atrium. The arrangements, as illustrated in the original classification credited to Lucas, show a very similar lesion. In that drawing, however, the variants described as "Type A2a" or "Type B1" show a communication, either potential or real, between the right atrium and the vestibular component of the divided left atrium. Such a communication is lacking in the example described by Hubrechts and colleagues.[1] When recognised on the basis of totally anomalous pulmonary venous return in the setting of divided left atrium, the lesion finds a suitable resting place. Coding in this fashion also circumvents the controversy with regard to malposition of the primary atrial septum. Only when the lesion can properly be described and coded, are we likely to be able to resolve the ongoing problem regarding its anatomical nature. Logic, therefore, points to recognising the lesion for what it is, namely division of the morphologically left atrium so as to produce totally anomalous drainage of normally connected pulmonary veins.

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Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008.

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