How best to explain unexpected arrangements of the atrioventricular valves?

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In the two articles published in the current issue of ‘Annals’, the authors describe the fascinating and unusual arrangements of the atrioventricular valves. In the first case, Sapre and colleagues\(^1\) identify the presence of dual orifices in a straddling tricuspid valve, with one of the orifices committed to the morphological left ventricle. Although they do not comment on this particular feature, as we will see, the arrangement produces an apparent paradox of a heart with three atrioventricular valves. In the second case, Shetkar and Kothari describe a patient having a heart with an absent right atrioventricular connection, but where the left atrium was connected to both ventricles through seemingly separate atrioventricular valves; the valves then superficially resemble the tricuspid and mitral valves.\(^2\) Here we have the paradox of the heart seemingly possessing two atrioventricular valves, yet in the setting of the absence of one atrioventricular connection. Shetkar and colleagues then discuss the possibility of how the arrangement at the atrioventricular junctions, which they describe in terms of double outlet from the left atrium, would have produced the appearance of three atrioventricular valves had the right atrioventricular junction been patent. In their discussion, they also highlight how these rare lesions, and others like them, can produce significant difficulties in arbitrating the most appropriate manner in which to describe the findings.

To my eyes, the key to providing the optimal description is emphasized by the first group of authors.\(^1\) They have recognised that the dual orifices in their patient are within the tricuspid valve, but that the valve is supported by an overriding atrioventricular junction. Thus, even though one of the orifices within the tricuspid valve is connected to the morphologically left ventricle, because of the normally connected mitral valve, the heart may be considered to have three atrioventricular valves. The heart itself, nonetheless, still has only two atrioventricular junctions. I suggest that it is the attention to the morphology of the atrioventricular junction or junctions, rather than the number of valvar orifices, which clarifies the seemingly paradoxical situations.

If we analyse the atrioventricular junctional morphology, we find that the normal heart possesses two atrioventricular junctions, with each junction committed exclusively to its own ventricle. In the setting of a deficient atrioventricular septation, the two atrioventricular junctions become part of a common structure, which is usually guarded by a common atrioventricular valve. My own preference is to describe this situation in terms of atrioventricular septal defects in the setting of common atrioventricular junctions.\(^3\) The arrangement is equally well described in terms of an atrioventricular canal.\(^4\) It is less satisfactory, however, than to argue that the left side of the atrioventricular valve guarding the canal is no more than a “cleft mitral valve”.\(^4\) The valve is certainly made from the same building blocks as the normal mitral valve. The normal mitral valve, nonetheless, is described as such because of its resemblance to the Episcopal mitre. This fact is pertinent to the subsequent discussions. In the normal heart, the mitral valve has an aortic leaflet that guards one-third of the circumference of the left atrioventricular junction, and a mural leaflet that guards two-thirds of the left junction. Such a valve can have a cleft in the aortic leaflet, but the cleft then points into the subaortic outflow tract.\(^5\) In the setting of a common atrioventricular junction, the left half of the common valve is guarded by three leaflets, two of which bridge the ventricular septum. The third leaflet, the mural leaflet, guards less than one-third of the left junction. The left valve seen in the setting of the atrioventricular canal defect, or the atrioventricular septal defect with common atrioventricular junction as I prefer to call it, bears no resemblance to the normal mitral valve.\(^3,5\) During surgical repair of such lesions, it is necessary to close the zone of apposition between the left ventricular components of the bridging leaflets so as to create a new left valve, which is also competent. Such a surgical maneuver, nonetheless, does not recreate a mitral valve, since the mural leaflet of the surgically created left valve guards only one-third of the newly formed left atrioventricular junction.

As I have already intimated, all of the above discussion is pertinent to the hearts described in the two cases.

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reported in the current issue. Confusion can be avoided if the valves are described on the basis of the junctions from which they are hinged, rather than on their perceived individual morphology. As is described by Sapre and colleagues,[1] their patient had duplication of the tricuspid valve. One of the valvar orifices was draining to the left rather than the right ventricle, but the tongue of leaflet tissue producing the dual valvar orifices was bound down to the crest of the malaligned ventricular septum. Because of this, there was no potential for shunting at the ventricular level. Instead, the incompetence of the valvar orifice permitted shunting from the left ventricle to the right atrium. The authors treated their patient in the Operating Room simply by closing the valvar orifice that connected the right atrium with the left ventricle. They did not comment on the fact that their patient could have been considered to have three atrioventricular valves. Had they drawn a simplistic cartoon in the fashion of Figure 2b, as prepared by Shetkar and Kothari,[2] however, then the potential presence of three atrioventricular valves would have become apparent. Indeed, as pointed out by Shetkar and Kothari,[2] a patient with dual orifices in a straddling and overriding left atrioventricular valve has already been described. Because of the presence of a left-handed ventricular topology in that case, the arrangement produced the apparently paradoxical situation of double inlet left ventricle co-existing with discordant atrioventricular connections.[6]

In my opinion, it is straddling and overriding of a left atrioventricular valve which has dual orifices in the setting of tricuspid atresia that provides the most logical explanation for the arrangement encountered by the second set of authors.[2] In their patient, however, the overriding leaflets of the left valve were not bound down exclusively to the crest of the malaligned ventricular septum. There was, therefore, a co-existing ventricular septal defect. The authors describe the defect as being of the inlet variety. In that the overriding left atrioventricular valve provided an inlet to the morphologically right ventricle, this is a justified description. It should be remembered, however, that the patient also had absence of the right atrioventricular connection. It is this feature that produces tricuspid atresia. Taken together, the absence of one atrioventricular connection, combined with overriding of the other atrioventricular junction, produces the uniatral, but biventricular, connection.[7] Most usually, overriding of one atrioventricular junction is found when the other atrioventricular junction is both present and patent. All patients with overriding atrioventricular junctions, nonetheless, have double outlet from the atrium supporting the junction. Such double outlet can also be found in patients with common atrioventricular junction when there is gross malalignment of the primary atrial septum. This is seen most frequently with leftward deviation of the primary septum, and then the only exit from the left atrium is through the ostium primum defect.[8] When the double outlet left atrium is seen with common atrioventricular junction and rightward deviation of the primary atrial septum, the lesion is comparable to the patient described in the current issue of the journal, but the atrial shunt would be through a primum defect rather than through the oval fossa.[9]

There are then fundamental differences in the orientation of the ventricular septum in hearts with straddling and overriding of one of two atrioventricular junctions.[10] This is key to the comments of the authors with regard to the location of the ventricular septal defect found in their patient.[2] When it is the mitral valve that is straddling and overriding the ventricular septum, then the valve usually straddles through a ventricular septal defect opening to the outlet of the morphologically right ventricle. In this setting, there is no obvious malalignment between the atrial and ventricular septal structures. Indeed, the septums are well aligned at the cardiac crux. Malalignment between the atrial and ventricular septal structures at the crux of the heart is the pathognomonic feature of straddling and overriding of the tricuspid valve, as would have been the situation in the case described by Sapre and colleagues.[1] These differences in atrioventricular septal malalignment provide the key to understanding the disposition of the atrioventricular conduction axis.[10] The axis will be anticipated to arise from the regular atrioventricular node when there is alignment at the crux, but from an anomalous posteroinferior node when there is septal malalignment at the crux. Shetkar and Kothari, however, describe their patient as having an inlet ventricular septal defect. Since I would argue that the patient has overriding of the mitral valve, I would have anticipated the presence of an outlet septal defect. The explanation for this discrepancy is that the overriding of the solitary left atrioventricular junction provides an inlet to the morphologically right ventricle, even though there is coexisting tricuspid atresia. And, although there is then malalignment of the ventricular and atrial septal structures so as to permit the overriding, the two septums will remain in alignment at the crux. I would predict, therefore, that the atrioventricular conduction axis in their patient arose from a regularly positioned atrioventricular node.

All of this discussion brings into focus the advisability of describing the valves residing within the right and left ventricles as the tricuspid and mitral valves, respectively. When there are biventricular atrioventricular connections, and each atrium is connected to its own ventricle, the tricuspid valve certainly does reside within the morphologically right ventricle, and the mitral valve in the morphologically left ventricle. The valves then have distinctive features, with the tricuspid
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valve being septophilic, and the mitral valve being septophobic. Such normal valves, however, also have distinctive arrangements of their papillary muscles. When the arrangement is complicated by the finding of dual valvar orifices in hearts with uniatrial but biventricular connections, then the components of the solitary valve shared between the ventricles are still capable of being perceived as having septophilic and septophobic characteristics, as is shown by the case described in the current issue. Therefore, because of the overriding nature of the valve, nonetheless, the valve itself also has features resembling a common valve, as is again suggested by the authors. The anatomy of the papillary muscles supporting the components of the overriding valve, however, is most unlikely to be comparable to that found in the presence of normal tricuspid and mitral valves, or to those that support a common valve. In the circumstances of the second case, therefore, I would describe the valves within the two ventricles as representing the right and left ventricular components of an overriding left atrioventricular valve, rather than as tricuspid and mitral valves. I would also prefer to describe the valve itself as being a left valve, since it is hinged from the left atrioventricular junction. In that it coexists with tricuspid atresia, it could justifiably be considered to be a mitral valve in its own right. As we have seen in the other case described in the current issue of the journal, the valve with dual orifices was itself well described as being tricuspid, even though one of its orifices was committed to the morphologically left ventricle. It is appreciation of the overriding nature of the atrioventricular junction supporting the valves in question that clarifies the situation. Differentiation of the nature of the atrioventricular junctions, therefore, permits valves to be described in meaningful fashion, even when it seems there might be three such entities within a given heart.

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