Cardiol Young 2005; 15 (Suppl. 3): 7–16
© Cambridge University Press
ISSN 1047-9511

The functionally univentricular circulation: anatomic substrates as related to function

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Keywords: Single ventricle; double inlet ventricle; atrioventricular valvar atresia; hypoplastic left heart syndrome; hypoplastic right heart

PATIENTS WITH A FUNCTIONALLY UNIVENTRICULAR circulation represent a challenge for both surgeons and cardiologists.[1–4] In this introductory paper, we aim to describe the variability in cardiac phenotype amongst these patients, and to review how function might relate to anatomy.

Anatomic substrates

Candidates for construction of the Fontan circulation, the endpoint nowadays for most patients possessing functionally univentricular arrangements, represent a heterogeneous group of anomalies that have in common the feature that only one of the chambers within the ventricular mass is capable of supporting independently either the pulmonary or systemic circulation. Although they all have, therefore, a functionally univentricular circulation, it is incorrect to describe the majority of these patients as having univentricular hearts, since almost all will usually possess two ventricles within their ventricular mass, one large and the other small.[5,6]

The true univentricular heart

The exceptions to the rule of most patients possessing two ventricles, with one being small, are exceedingly rare. Occasionally, nonetheless, there may be no anatomic evidence of a second chamber. The heart can then be described as possessing a solitary ventricle, and hence as being truly univentricular.[7] In this situation (Fig. 1), the ventricular mass is typically coarsely trabeculated throughout its apical components, the trabeculations being even coarser than those seen in the normal morphologically right ventricle. The developmental basis for this anomaly is, as yet, not at all understood, albeit that it is reasonable to propose that it represents failure of ballooning of separate apical components for the morphologically right and left ventricles as proposed by Christoffels et al.[8] Anatomic studies of the conduction system in such hearts show a bizarre arrangement, confirming the possibility for such markedly abnormal morphogenesis. Usually there are dual atrioventricular nodes, situated inferiorly and laterally within the atrioventricular

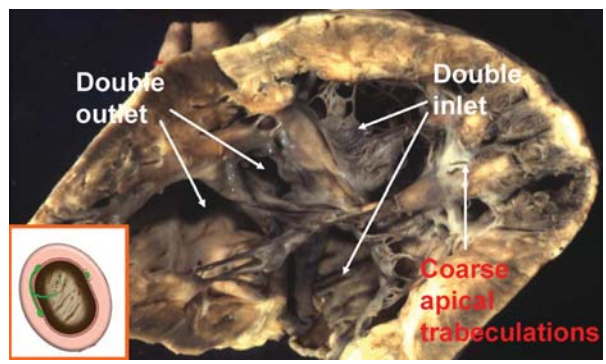

Figure 1.
An example of the very rare situation where there is only a solitary chamber within the ventricular mass. The ventricular morphology is best described as solitary and indeterminate and the ventricular trabeculations in this situation are extremely coarse. In this heart there is also double inlet and double outlet from the solitary ventricle. The inset shows the bizarre arrangement of the conduction system in a similar heart sectioned histologically. There is a sling of conduction axis connecting dual atrioventricular nodes, and giving rise to a solitary strand which activates the ventricular mass. This pattern is strong evidence that the ventricle is neither morphologically right nor left, but solitary and indeterminate.

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https://doi.org/10.1017/S1047951105001563 Published online by Cambridge University Press

junction, which are connected by an inferior sling of conduction tissue, together with a branching strand reminiscent of a right bundle branch[9] (Fig. 1 inset).

Hearts with one big and one small ventricle

For most patients with a functionally univentricular circulation, there will be two ventricles present, one large and one small. Why then, does the presence of a second ventricle matter, if the circulation is functionally univentricular? As we will see, identification of two ventricles is the first step in determining the type of atrioventricular connections, the nature of which can have a profound effect on ventricular function. By definition, identification of both ventricles will also determine the location of the ventricular septum, as well as the relative position of the chambers. Both these features impact on the location of the ventricular conduction tissues, an important variant for the surgeon seeking to palliate in optimal fashion this group of patients.[10]

How is it best to make sense of this very heterogeneous group of anomalies? The key to unravelling these seemingly complex anatomic arrangements is by using the process of sequential segmental analysis.[11–13] Using this system, the heart is broken down into three components, or segments, namely the atriums, ventricles and great arteries. Having determined the morphology of these parts, the next step is to analyse the manner in which they are connected together at the atrioventricular and ventriculo-arterial junctions. Central to the process is the use of "markers" for the chambers in each component, such that a morphologically right ventricle, for instance, can be identified as such regardless of its relative position with respect to the remainder of the cardiac components. When determining the "marker", we use the most constant "building block" for the chambers or arterial trunks seen within any of the three segments when looking across an entire range of cardiac malformations. The notion for using such key components to identify the segments came from the so-called "Morphologic Method", as set out initially by Van Praagh et al.[14]

Overall segmental arrangements

The process of sequential segmental analysis provides the means for grouping all patients with a

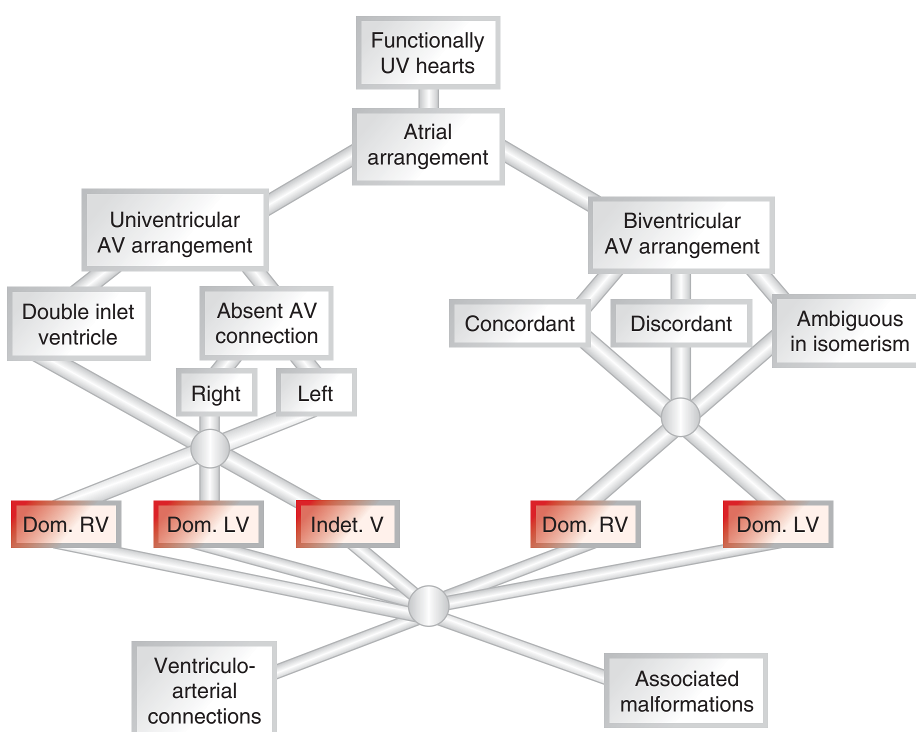

Figure 2.
Schematic representation to show the possible segmental combinations that can result in a functionally univentricular heart. Any of the four types of atrial arrangement can be present. There are then two major divisions, those with univentricular atrioventricular connections, double inlet, or absent connection, or else those with biventricular connections which can be concordant, discordant or ambiguous when the atrial appendages are isomeric. Any of these types of atrioventricular connection can potentially co-exist with either a dominant left, right or indeterminate ventricle. Likewise any form of ventriculo-arterial connection is possible and associated malformations need also to be documented separately.

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functionally univentricular circulation into two broad categories, those with biventricular and those with univentricular atrioventricular connections (Fig. 2). Either of these types of connection can occur with any of the four forms of atrial arrangement, and in the presence of a variety of ventriculo-arterial connections. The latter must also be determined for complete analysis, as must the presence of important associated lesions, such as a ventricular septal defect, or sub-pulmonary or sub-aortic stenosis (Fig. 2). The morphology of the main pumping chamber, which as already indicated will usually be of either morphologically right or left types, is then a crucial part in this analysis. It is now customary to divide the ventricles into three component parts, namely the inlet, apical trabecular component and outlet (Figs 3 and 4).[11–13] It is the nature of the apical trabeculations that determines ventricular morphology, and also permits the recognition of the overall topologic arrangement of the ventricular mass. The apical trabeculations are coarse within the apex of the morphologically right ventricle (Fig. 3), but much finer and mesh-like within the apical portion of the morphologically left ventricle (Fig. 4). The topologic arrangement of the normal right ventricle is right-handed, since it is only the palmar surface of the right hand that will fit on

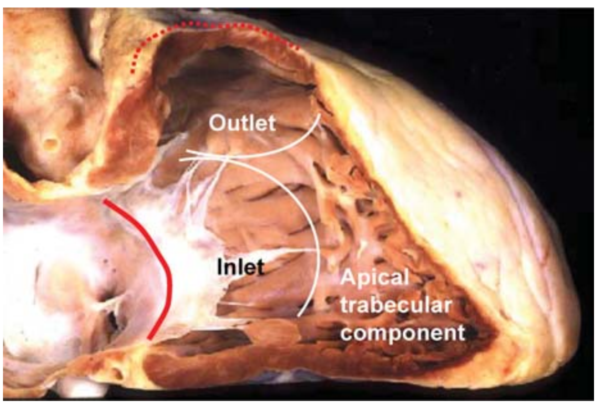

Figure 3.
Analysis of the ventricular mass in functionally univentricular hearts is based on the principle that the ventricles in the normal heart extend from the atrioventricular junction (solid red line) to the ventriculo-arterial junction (dotted red line). The muscular mass thus delineated, shown here for the morphologically right ventricle, can be analysed in three parts, inlet, apical trabecular and outlet components.

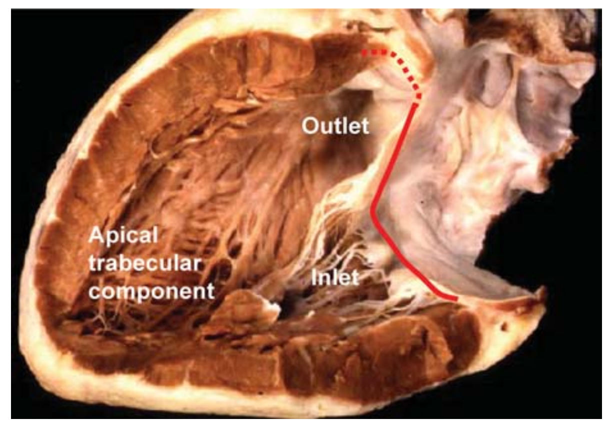

Figure 4.
As for the morphologically right ventricle, shown in Figure 3, the normal morphologically left ventricle extends from the atrioventricular junction (solid red line) to the ventriculo-arterial junction (dotted red line). In the morphologically left ventricle, these junctions are in fibrous continuity along the inner curvature. The normal left ventricle, nonetheless, can also be analysed in three parts, inlet, apical trabecular and outlet components.

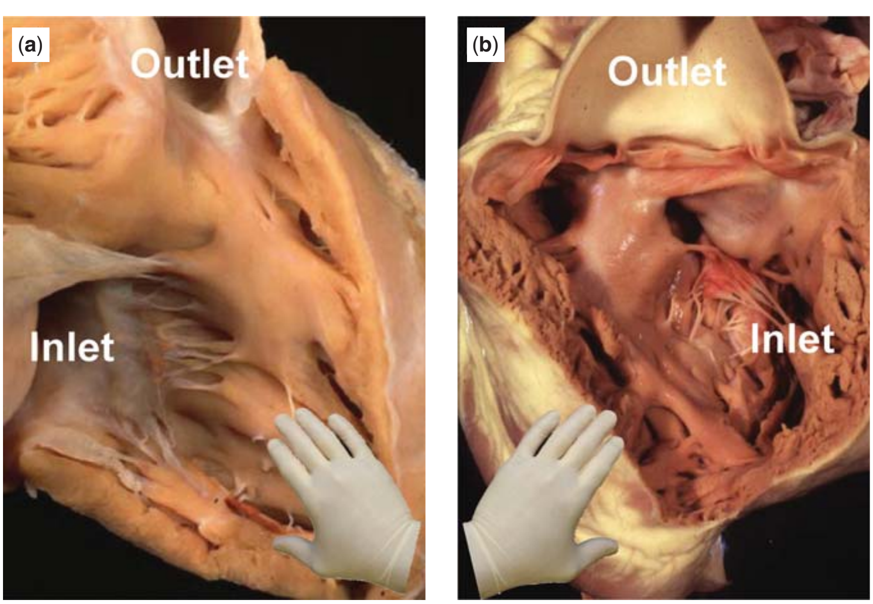

Figure 5.
*These specimens show the principle underscoring ventricular topology. Whichever hand fits within the morphologically right ventricle such that the palmar surface is towards the septum, thumb in the inlet and fingers in the outlet, defines the topology of the ventricles. The morphologically right ventricle seen in (**b**), from a heart with congenitally corrected transposition, is then effectively the mirror image of the normal right ventricle shown in (**a**).*

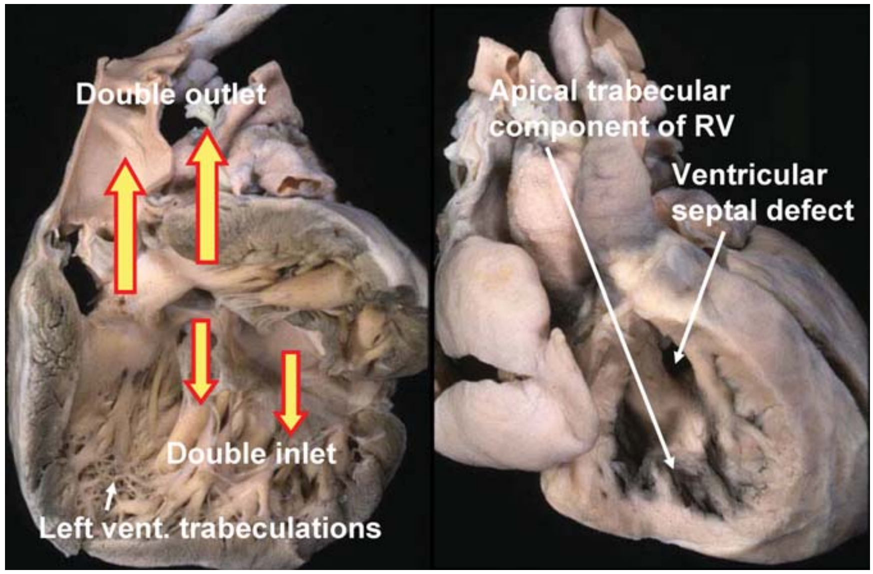

Figure 6.
This specimen shows the essence of the tripartite approach to analysis of the ventricles in functionally univentricular hearts. The left-hand panel shows that there is double inlet and double outlet to a morphologically left ventricle (arrows). As a consequence, the rudimentary right ventricle, shown in the right-hand panel, contains neither inlet nor outlet component. It is still recognisable as a morphologically right ventricle, nonetheless, on the basis of the coarse trabeculations within the remaining apical component.

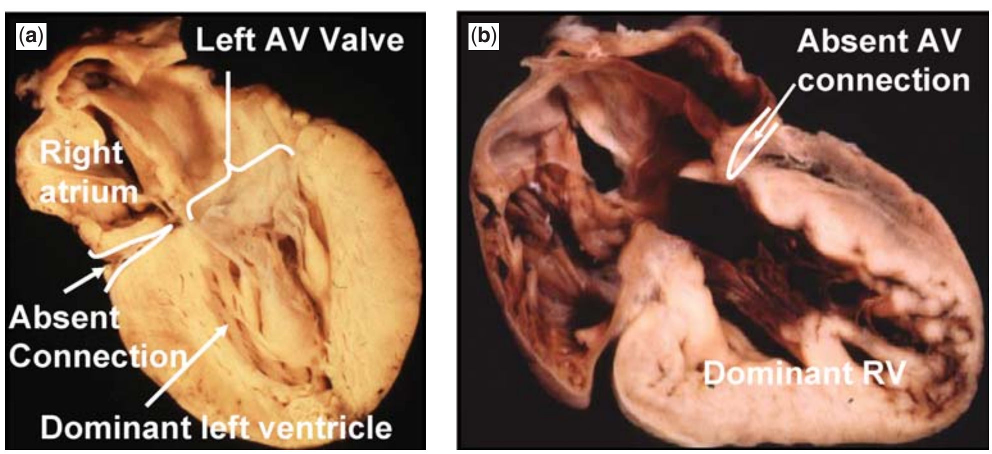

Figure 7.
*Examples of absent right (**a**) and absent left (**b**) atrioventricular connection as seen in classical tricuspid (**a**) or mitral atresia (**b**). The remaining atrioventricular connection is concordant, such that the dominant ventricle is finely trabeculated and of left morphology in (**a**) and coarsely trabeculated and of right morphology in (**b**).*

the septal surface of the right ventricle with the thumb in the inlet and fingers in the outlet. The topology of the left ventricle is left-handed in the normal heart.[15] These arrangements are mirror-imaged when there is left-handed topology, as seen, for example, in the commonest variant of congenitally corrected transposition (Fig. 5).

In congenitally malformed hearts, such as the majority of patients who are candidates for a Fontan circulation, the apical trabecular patterns are retained within both the dominant and small ventricles, even when the small ventricle lacks both its inlet and outlet components (Fig. 6). Thus, following the principle of the "Morphologic Method", it is this apical trabecular component that becomes the morphologic "marker" used for identification of the ventricles, be they complete or incomplete, and be they large or small.[14] In the patient with a functionally univentricular circulation, it is possible, at least for the morphologist, to compare the apical trabeculations within the two ventricles, and hence to determine the morphology of the dominant pumping chamber. In

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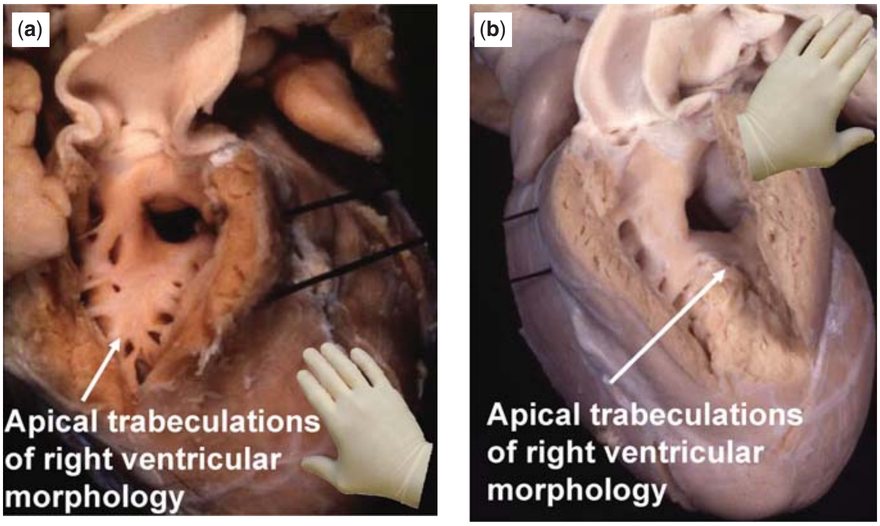

Figure 8.
*The same principle can be applied to ventricles from patients with functionally univentricular circulations. In the absence of a patent inlet, the thumb is placed towards the potential site of the inlet. These two hearts both have double inlet left ventricle with discordant ventriculo-arterial connections. In (**a**), the rudimentary right ventricle is located on the right shoulder of the dominant morphologically left ventricle, and the inference is made that there is right-handed ventricular topology. Panel (**b**) shows the situation with presumed left-hand ventricular topology.*

the setting of classical tricuspid atresia, then, the apical trabecular pattern of the dominant ventricle will usually be of fine, or left, type (Fig. 7a). For classical mitral atresia, it will be coarse, and of right type (Fig. 7b).

As mentioned above, the topology of the ventricles can also vary, and can be the mirror image of normal, depending on whether it is the right or left hand that can be placed within the morphologically right ventricle with the thumb within the inlet, or potential side of the inlet, and the fingers towards the outlet (Figs 5 and 8).

The univentricular atrioventricular connection

In the setting of those patients with univentricular atrioventricular connection, or connections, there are a very limited number of ways in which the atrial and ventricular segments can join together at the atrioventricular junction. There can either be complete absence of the right or left-sided atrioventricular junction, or else a double inlet connection. In either setting, the atrioventricular junction or junctions can then lead into either a dominant right, a dominant left, or a solitary and indeterminate ventricle (Fig. 2). Most frequently, of course, in the setting of an absent atrioventricular connection, the remaining atrioventricular connection will be concordant. This gives the substrate for so-called classical forms of tricuspid and mitral atresia (Fig. 7). In the setting of double inlet, both atrioventricular valves will usually lead into a morphologically left ventricle, and much more rarely to morphologically right, or indeterminate ventricles. Note should be taken, nonetheless, of the fact that more complex connections can co-exist at the level of the atrioventricular junctions (Fig. 9). So as to recognise these rarer variants, and to differentiate them from the more common forms, it is crucial, once again, to follow the principles of sequential segmental analysis.[16]

In clinical practice, the relative position of the ventricles provides the most valuable clue to their morphology. There may rarely be exceptions to the rule, but almost always, a small and incomplete morphologically right ventricle will sit on the shoulder of a dominant morphologically left ventricle. Conversely, a small and incomplete morphologically left ventricle will sit beneath a dominant right ventricle, being carried on its diaphragmatic surface.[17] In cross-section, this means that the atrioventricular valve, or valves, will usually be posterior to the ventricular septum in the setting of a dominant morphologically left ventricle, as is seen in classical tricuspid atresia or double inlet left ventricle (Fig. 10). They will then usually be anterior in the setting of a dominant right ventricle, as seen in classical mitral atresia or double inlet right ventricle (Fig. 11). The exceptions to these arrangements are those rare instances of so-called criss-cross hearts, in which the spatial position of the ventricles and their morphology are mismatched.[18,19]

Biventricular atrioventricular connections

On occasion, hearts with biventricular atrioventricular connections can be functionally univentricular. Although the morphology of hearts within this group is even more heterogeneous than those with a univentricular atrioventricular connection (Table 1),

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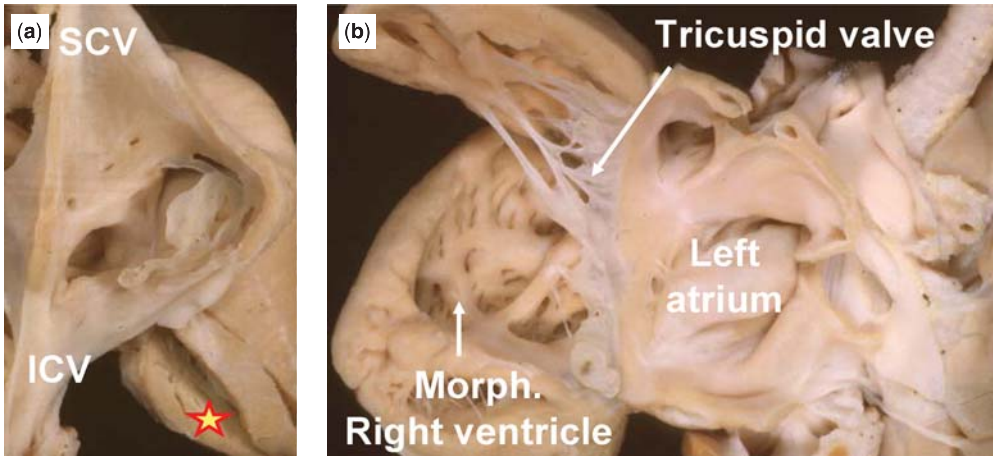

Figure 9.
*An example of a more complex form of atrioventricular valvar atresia. Initially, it appears as though this heart has classical tricuspid atresia, since the floor of the right atrium is completely muscular and there is complete absence of the right atrioventricular connection (panel (**a**)). The valve-like tissue seen in panel (**a**) is a prominent and fenestrated Eustachian valve. Panel (**b**), however, shows that the left atrium leads not into a dominant morphologically left ventricle, as might be expected, but into a coarsely trabeculated morphologically right ventricle, via a morphologically tricuspid valve. There was a slit-like incomplete left ventricle found in the right-sided diaphragmatic margin of the ventricular mass (star in panel (**a**)), pointing to the presence of left-hand ventricular topology. Had the right-sided atrioventricular connection been formed, it would have been guarded by a mitral valve. Careful sequential segmental analysis and description is required in cases such as these. SCV: superior caval vein; ICV: inferior caval vein.*

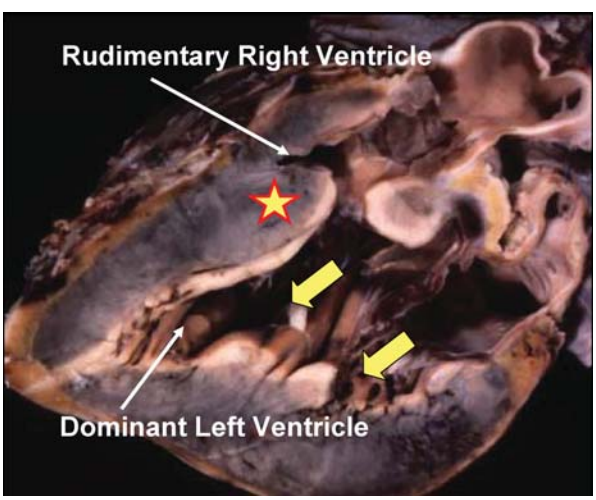

Figure 10.
This long axis section of a heart with double inlet left ventricle shows the relative position of the rudimentary right ventricle and ventricular septum with respect to the inlets. The incomplete right ventricle is situated superior to the dominant left ventricle. The two inlet valves (yellow arrows) leading the dominant left ventricle are posterior-inferior to the ventricular septum (star).

analysis should again start using the sequential segmental approach. Then, once the atrial arrangement has been established, it should always be possible to work out, with appropriate knowledge of ventricular morphology, whether the atrioventricular connections are concordant, discordant, or biventricular in

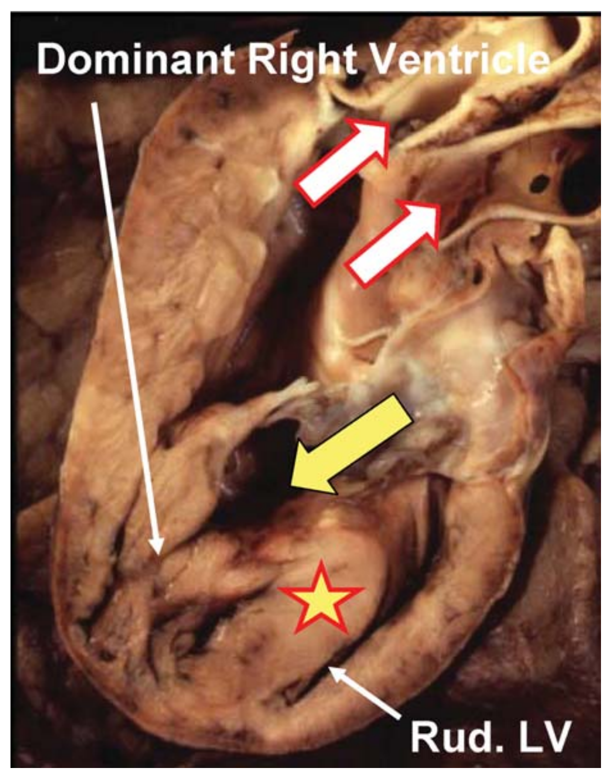

Figure 11.
A similar long axis section of a heart with double inlet right ventricle shows the opposite relationship to that shown in Figure 10. In this instance, the incomplete ventricle, which is of morphologically left type, is located postero-inferiorly. The common atrioventricular valve (yellow arrow) is supero-anterior to the plane of the ventricular septum (star).

Table 1. Candidates for a Fontan circulation amongst patients with biventricular atrioventricular connections.

Straddling atrioventricular valve
Imperforate atrioventricular valve
Hypoplastic left heart syndrome
Hypoplastic right heart (pulmonary atresia with intact ventricular septum)
Unbalanced atrioventricular septal defect
Ventricular imbalance associated with coarctation
Idiopathic right ventricular hypoplasia
Double outlet right ventricle with non-committed interventricular defect
Huge ventricular septal defect

the setting of isomerism of the atrial appendages. Equally important will be establishing the type of ventriculo-arterial connection, and any associated malformations (Fig. 2).

Within this group are to be found hearts with biventricular atrioventricular connections that are unseptatable, such as those with straddling atrioventricular valves, or a huge ventricular septal defect, or those with double outlet and a non-committed interventricular communication, or those with imperforate atrioventricular valves. There are also those with biventricular atrioventricular connections and obstruction to either outlet in the setting of an intact ventricular septum, this combination producing the so-called hypoplastic left or right heart syndromes. This final group of hearts is the one in which the ventricles contain all their component parts, but in which one or other ventricle is of insufficient size to support separately the entirety of the systemic or pulmonary circulations. The inclusion criterion for this category is particularly hard to define in anatomic terms, and can be equally hard on clinical grounds, particularly if diagnosis is made prenatally.[20,21] Within this group we would include, amongst those with unequivocal hypoplastic right and left heart syndromes, those with unbalanced atrioventricular septal defects and marked ventricular disproportion, usually in the setting of left ventricular hypoplasia associated with coarctation of the aorta, but sometimes with marked hypoplasia of the right side of the heart.

The location of the atrioventricular conduction system

As already indicated, perhaps the most important corollary of knowing the morphology of the ventricles in the functionally univentricular heart, in the setting of either univentricular or biventricular atrioventricular connections, is being able to identify the location of the atrioventricular conduction system. We have already made reference to the unusual course of the ventricular conduction tissues in the rare

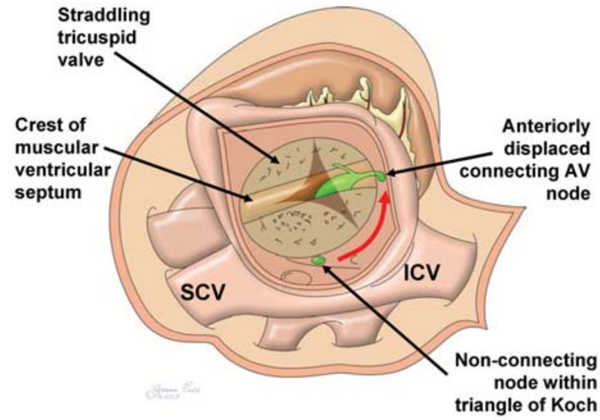

Figure 12.
A surgeon's view of a straddling and overriding tricuspid valve, as seen through the opened right atrium. In this setting, the connecting atrioventricular node is located at the point where the ventricular septum meets the atrioventricular junction, and is thus effectively displaced anti-clockwise around the right atrioventricular junction to varying degrees (red arrow).

situation of a solitary and indeterminate ventricle (Fig. 1). In other functionally univentricular hearts, the location of the atrioventricular node, and the course of the ventricular conduction tissues, will depend both on the morphology and topology of the ventricular mass, and also on the degree of alignment between the inferior portion of the ventricular septum and the atrial septum.

In hearts with a dominant morphologically right ventricle, and normal, or right-hand ventricular topology, there is usually good alignment between these two structures, resulting in a regularly positioned atrioventricular node within the morphologically right atrium, and a normal course for the ventricular conduction system. Thus, patients with classical mitral atresia, double inlet right ventricle, or straddling and overriding mitral valve, for example, most of whom will have right-hand ventricular topology and a left-sided hypoplastic left ventricle, the conduction tissues will be normally disposed. If the ventricular mass is the mirror image of normal in this setting, and there is left-handed ventricular topology, usually associated with a right-sided morphologically left ventricle, then there are typically dual atrioventricular nodes, which form a so-called sling of conduction tissue across the crest of the hypoplastic ventricular septum. The bundle branches will then be the mirror image of normal.[22,23]

In patients with classical tricuspid atresia and a dominant left ventricle, the ventricular septum retains some alignment with the atrial septum because the entire right atrioventricular connection is absent. The atrioventricular node is then found at the site of the "dimple" in the right atrium. In contrast, in other

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patients with univentricular connection to a dominant left ventricle, the atrioventricular node cannot connect with the bundle branches in its regular position. In most patients with double inlet left ventricle, or in those with straddling and overriding tricuspid valve, for instance, there is malalignment between the inferior portion of the ventricular septum and the atrial septum. In this situation, the atrioventricular node is found at the point where the ventricular septum meets the atrioventricular junction.[24,25] In patients with a straddling tricuspid valve, the atrioventricular node "moves" anteriorly around the right atrioventricular junction with greater degrees of malalignment of the ventricular septum (Fig. 12). The situation seen in double inlet left ventricle is then the extreme end of this spectrum, and the atrioventricular node is then positioned antero-superiorly within the atrioventricular junction at the mouth of the right atrial appendage[26] (Fig. 13). The morphology of the bundle branches will then follow the topology of the ventricular mass, being the mirror image of normal in the setting of left-hand ventricular topology.

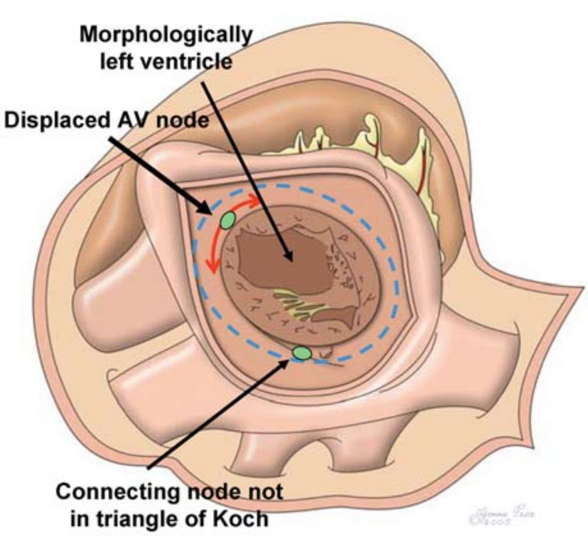

Figure 13.
The situation, again as seen by the surgeon through a right atriotomy, in double inlet left ventricle. In this entity, there is marked malalignment between the ventricular and atrial septums. The atrioventricular node is now to be found within the region of the atrioventricular junction, below the tip of the right atrial appendage. If a patch is placed to exclude the right atrioventricular valve from the Fontan circulation, it is usually inserted above the level of the atrioventricular junction so as to avoid the node irrespective of its location.

Morphology related to function

In terms of function, the ability of morphologically right ventricle to function as systemic ventricle is uncertain in the long term, but undoubtedly adaptation starts prior to birth. Take, for example, the morphologically right ventricle in hypoplastic left heart syndrome. Figure 14 shows the mean z-scores for the dimensions of the right heart across a variety of lesions afflicting the chambers of the left heart at a median gestation of 21.4 weeks.[27] In all groups, changes in the conformation of the morphologically

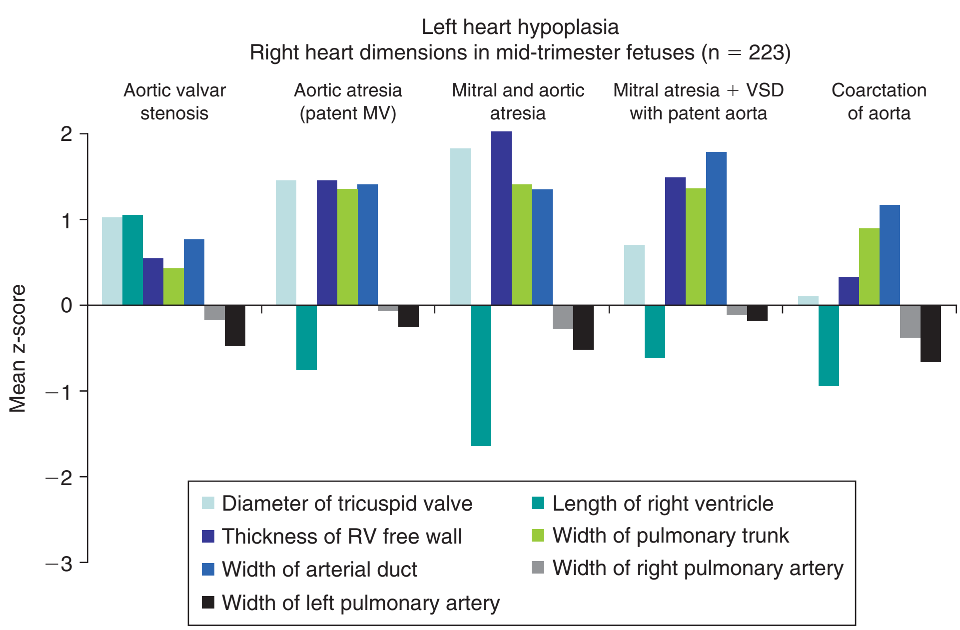

Figure 14.
This graph illustrates the change in conformation of the components of the right heart in fetuses with 5 different forms of left-sided hypoplasia. The median gestation is 22.3 weeks, and the bars show the mean z-score for the dimensions of the right heart thereby indicating the degree of deviation from the mean. The right ventricle is wider, shorter and thicker walled than normal even at a mean gestation of 22.6 weeks. The degree of change correlates with the severity of left heart disease.

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right ventricle are evident, with the chamber becoming wider, shorter, and thicker-walled concomitant with diversion of flow to the right heart. The changes are more marked for the more severe forms of hypoplasia of the left heart. There is also other evidence from postnatal specimens of an alteration in the trabecular patterning of the morphologically right ventricle, and alterations in morphology of the tricuspid valve, suggesting that the morphologically right ventricle is far from "normal" in this situation, beginning its adaptation to the functionally univentricular circulation early in development.[28,29]

What is clear is that the function of the dominant ventricle is dependant not only on its morphology but also on the type of atrioventricular connection that is present. We know, for instance, that in the normal heart the myofibres are oriented in three broad layers, although crossover from one layer to another is also crucial.[30,31] Superficial fibres are oriented obliquely, the middle fibres circumferentially, and the deep layer runs longitudinally (Fig. 15). These orientations have been shown to differ in some forms of functionally univentricular heart. So, in tricuspid atresia, and in pulmonary atresia with intact ventricular septum, for instance, the superficial fibres are more longitudinal.[32,33] Although one might think that the same rule would apply to all hearts with a dominant left ventricular chamber the superficial fibres are circumferentially orientated in patients with double inlet left ventricle.[34] The developmental reasons for this difference are again unclear, but determining these, and the time of onset, may well shed light on the reason why ventricular function can be so different between patients with tricuspid atresia versus those with double inlet left ventricle. Similarly, the cause for increase in the fibrous matrix that has been demonstrated in patients with tricuspid atresia is largely unknown, but given that it is present in all groups postnatally, most likely it has its genesis prior to birth.[35]

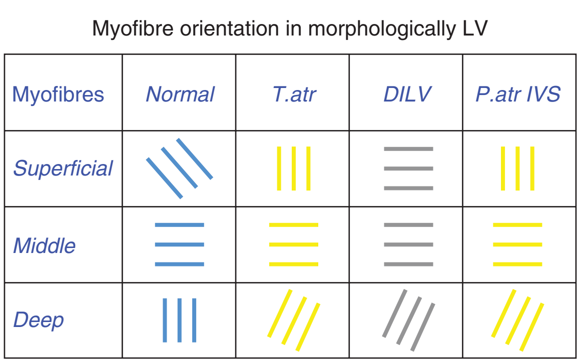

Figure 15.
This schematic representation summarises the major orientation of the myofibres in the superficial, middle and deep layers of the dominant left ventricle in normal hearts, and in hearts with tricuspid atresia (T.atr), double inlet left ventricle (DILV) and pulmonary atresia with intact ventricular septum (P.atr IVS). Although all have a dominant morphologically left ventricle, the orientation of superficial fibres differs between the groups. This may well play a role in observed differences in function.

Conclusions

Most patients with the functionally univentricular circulation will have two ventricles, albeit that one will be small and incapable of supporting either the systemic or pulmonary circulation. In order to make sense of the manifold substrates within this group, it is crucial to begin analysis in sequential segmental fashion, dividing the ventricle into 3 components, and making use of the apical trabecular pattern in order to determine ventricular morphology. Function will depend not only on the morphology of the dominant ventricle, but also on the type of atrioventricular connection. There is increasing evidence that the structure of the dominant chamber is not as in the normal heart, and that changes in conformation and fibrous matrix are likely to begin prior to birth.

Acknowledgement

The research on which this review is based was supported by grants from the British Heart Foundation together with the Joseph Levy Foundation.

Research at the Institute of Child Health and Great Ormond Street Hospital for Children NHS Trust benefits from R&D funding received from the NHS Executive.

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