Cardiol Young 2004; 14 (Suppl. 1): 3–12
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ISSN 1047-9511

Anatomical Considerations

Morphology of the functionally univentricular heart

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OF ALL THE COMPLEX MALFORMATIONS THAT affect the heart, those that have produced the greatest difficulty, over the years, in terms of logical description and classification are the ones with the circulations supported by a functionally univentricular mass. The problems in description stem from the fact that, although the ventricular mass functions as a singular entity, the majority of lesions falling within this category, from the stance of morphology, possess two ventricular chambers. For many years, and indeed, even now in many centres, the hearts are described as being "univentricular", or alternatively as "single ventricles". There is still no consensus as to which particular lesions should be included within this "univentricular" category. The paradigm of the group is usually taken as double inlet left ventricle.[1] In this lesion, the dominant left ventricle is accompanied by a rudimentary second chamber, albeit that arguments continue as to whether the second chamber is an incomplete right ventricle,[2] or simply an infundibulum.[3] Further arguments raged as to whether tricuspid atresia should be included within the univentricular category.[4,5] The debate is rationalised when we recognise that all the potentially univentricular hearts are treated nowadays by constructing the Fontan circulation,[6] or one of its variants,[7] as indeed are some unequivocally biventricular hearts.[8] Clarification and simplification of the anatomical arrangement in this functionally univentricular category, as we will show in this review, is provided by rigorous application of the crucial philosophic principle of analysis called the "Morphological Method",[9] coupled with separate analysis of the ventricular mass and the atrioventricular junctions.[10,11]

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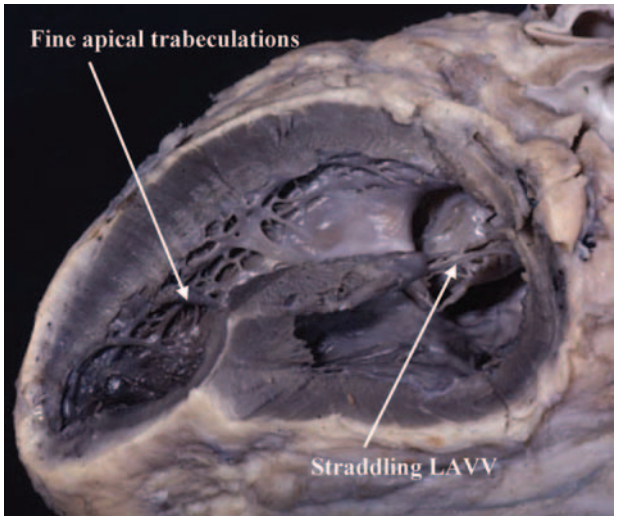


Figure 1.
The postero-inferior chamber found in hearts with double inlet right ventricle, as shown in this example, can be recognised as a rudimentary and incomplete left ventricle because of the fine criss-crossing nature of the apical trabeculations. Note the straddling tension apparatus of the left atrioventricular valve (LAVV).

The morphological method

This important principle was thrust into prominence when a group of European authors, including one of us, suggested that double inlet right ventricle was a "univentricular heart" of right ventricular type.[12] Van Praagh and his colleagues[9] highlighted the obvious deficiency in this analysis, since the majority of the hearts said to be univentricular possessed a second chamber of unequivocally left morphology (Fig. 1). So as to justify the use of "univentricular" to describe this anatomically biventricular arrangement, the European group had arbitrarily chosen to deny ventricular status to the incomplete and hypoplastic left ventricle, justifying this because it lacked its inlet component. In their letter, Van Praagh et al.[9] pointed

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out that this meant that one variable feature, namely the nature of a ventricular chamber, was defined on the basis of another feature that was itself variable, specifically the presence or absence of an inlet component and atrioventricular valve in the small chamber. They argued that this deficiency in logical analysis could be avoided if structures within the heart were defined on the basis of their intrinsic morphology, and not according to other features that were themselves variable.

Another problem, however, had emerged in the European system of analysis. Thus, if a hypoplastic ventricle was connected to less than half the circumference of an atrioventricular valve, it was deemed to be a non-ventricle. The same chamber, when connected to just over half the circumference of the overriding atrioventricular valve, suddenly became a ventricle. This problem was highlighted by Brandt and his colleagues.[13] We immediately recognised the logic and force of their argument, that more so since it emphasised the fact that the anatomic feature that was univentricular in the hearts described was not the ventricular mass, but rather the arrangement of the atrioventricular junctions.[10,11] As we will show, combination of the morphological method with junctional analysis has now served to clarify and simplify anatomical description of these functionally univentricular hearts.

How do we distinguish ventricular morphology?

Another problem underscoring much of the controversy concerning "univentricular hearts" was the concept of describing normal ventricles in terms of "sinus" and a "conus", a convention still employed by some recognised authorities.[3] Examination of normal ventricles, however, reveals no anatomic landmarks that justify this analysis in bipartite fashion (Figs 2 and 3). It is a simple matter, nonetheless, to recognise boundaries permitting the normal chambers to be described in terms of three functional parts. The ventricles are the pumps to the systemic and pulmonary circulations. Efficient mechanical pumps possess inlet and outlet valves, together with a driving piston. So it is with the ventricles. Extending from the atrioventricular to the ventriculo-arterial junctions, each ventricle has an inlet component, surrounding and supporting the atrioventricular valvar complex, an outlet component supporting the leaflets of the arterial valves, and an apical trabecular component (Figs 2 and 3). When all components are present, then each has distinctive features. Thus, the morphologically tricuspid valve, found only in the right ventricle, is attached by its tension apparatus to the septum. The mitral valve, in contrast, lacks septal attachments. In most instances,

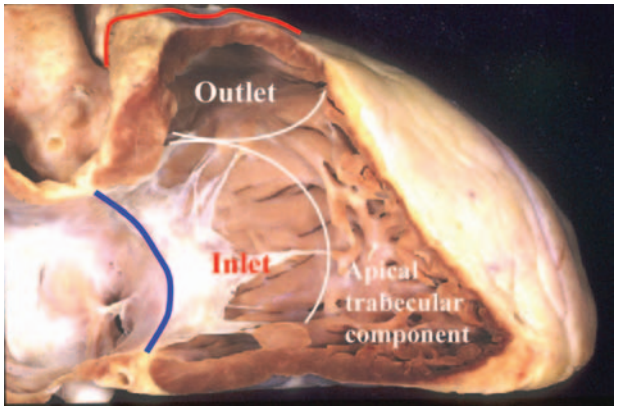


Figure 2.
The right ventricle is shown as revealed by removing its parietal wall. When considered as extending from the atrioventricular (blue line) to the ventriculo-arterial junction (red line), then the ventricle is readily analysed in terms of inlet, apical trabecular, and outlet components.

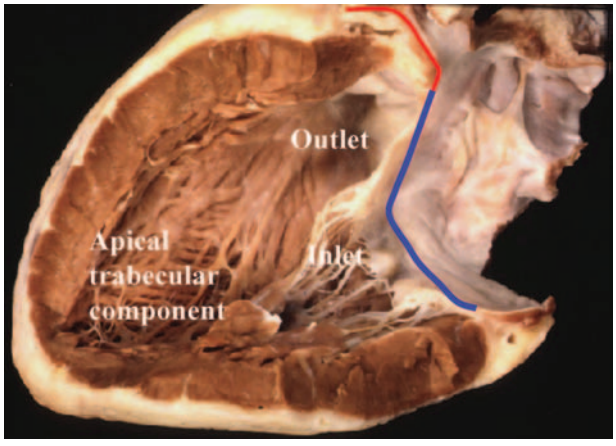


Figure 3.
The left ventricle is shown from the same heart from which the right ventricle was shown in Figure 2. Again, when judged as extending from atrioventricular (blue) to ventriculo-arterial (red) junctions, the ventricle has inlet, apical trabecular, and outlet components.

the arterial valve supported by the morphologically left ventricle has fibrous continuity between its leaflets and those of the mitral valve, whereas the arterial valve arising from the morphologically right ventricle is almost always supported by a complete muscular infundibulum. Not all ventricles, however, possess inlet and outlet components. Because the apical trabecular component is the most constant of the ventricular parts, then using the concept of the morphological method,[9] it is this feature that is best used to distinguish between morphologically right and left ventricles. The apical component of the morphologically right ventricle has coarse apical trabeculations (Fig. 2), while the apical part of the morphologically left ventricle has much finer criss-crossing apical trabeculations

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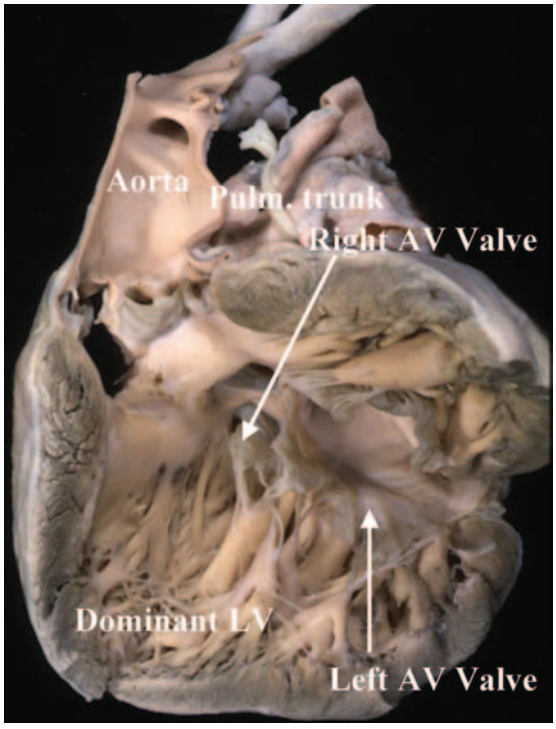


Figure 4.
This heart has double inlet to, and double outlet from, a dominant left ventricle.

(Fig. 3). These features also serve to distinguish the ventricles even when each apical component exists in isolation. Thus, in hearts having double inlet to, and double outlet from, the morphologically left ventricle (Fig. 4), the incomplete morphologically right ventricle, even though no more than a pouch, can still be recognised on the basis of its coarse apical trabeculations (Fig. 5). Similarly, in presence of double inlet to, and double outlet from, the morphologically right ventricle (Fig. 6), the fine apical trabeculations serve to distinguish the remnant of the morphologically left ventricle (Fig. 1).

This tripartite analysis of the ventricular mass permits all chambers to be described as ventricles, removing the illogical need to disqualify certain chambers from ventricular status simply because they do not possess an inlet component.[13] Complete ventricles possess all three components, albeit that the chambers are not necessarily of sufficient size to support the systemic and pulmonary circulations in biventricular fashion. As we will see, some functionally univentricular hearts can possess two complete ventricles when one of the ventricles is hypoplastic. Incomplete ventricles, lacking one of their components, can be sufficiently large on occasion to support one or other of the circulations. Thus, the left ventricle in hearts with double outlet right ventricle is an incomplete chamber, lacking its outlet component. Such left ventricles, nonetheless, are fully capable of supporting the

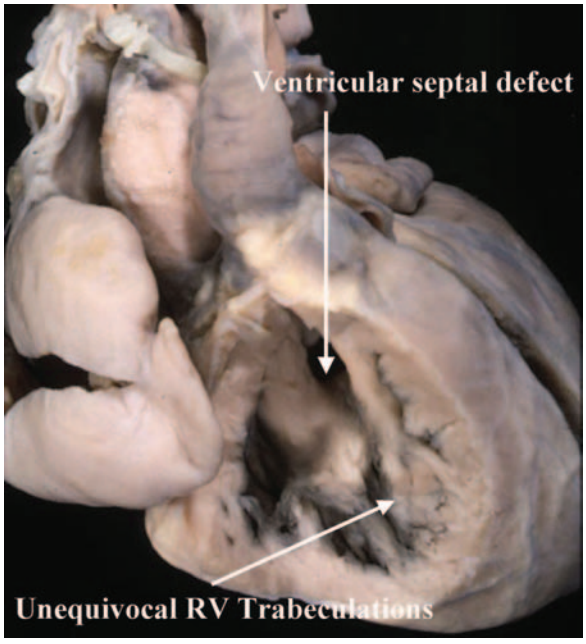


Figure 5.
This photograph shows the second ventricular chamber from the heart shown in Figure 4. Based on the morphology of the apical trabeculations, the pouch is unequivocally an incomplete right ventricle.

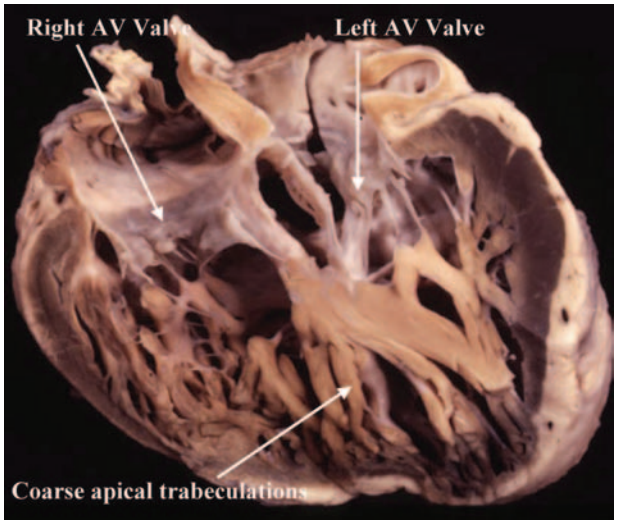


Figure 6.
This "four-chamber" section is through the dominant ventricle of the heart shown also in Figure 1. As it can be seen, there is double inlet to a dominant right ventricle.

systemic circulation when surgical repair has restored to them their outlet component. Incomplete left ventricles lacking an inlet component, in contrast, are unable to support one or other of the circulations. Nowadays, it is also unusual for surgeons to attempt to use an incomplete right ventricle to support one of the circulations, but this has been shown to be feasible in the past. The incomplete right ventricle, for example, was incorporated into the pulmonary circulation

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when using the atrioventricular variant of the Fontan procedure,[14] while incomplete right ventricles were also used to support the systemic circulation subsequent to ventricular septation for double inlet left ventricle.[15] In almost all instances in current practise, nonetheless, diagnosis of an incomplete right ventricle is likely to be an indication for a functionally univentricular repair.

Can there be an anatomically univentricular heart?

The majority of patients submitted for functionally univentricular surgical repair will not possess anatomically univenticular hearts. Instead, they will have unbalanced ventricular masses, with either a dominant left ventricle and a hypoplastic right ventricle, or a dominant right ventricle with a hypoplastic left ventricle. Only in very rare circumstances it is possible to find truly anatomically univentricular hearts. When such a truly univentricular heart is suspected, it is important first to exclude the possibility that one of the ventricles is so hypoplastic as to be virtually unrecognisable. In the clinical situation, it may be very difficult to distinguish this situation from the true anatomic univentricular arrangement. It is much easier for the morphologist to make this distinction, since the location of the incomplete and grossly hypoplastic ventricle is indicated by the delimiting coronary arteries. By searching for the delimiting coronary arteries, which mark the site of the hypoplastic apical ventricular septum, it is possible to identify incomplete right ventricles, always positioned anterosuperiorly within the ventricular mass, or incomplete left ventricles, always found postero-inferiorly relative to the dominant right ventricle (Fig. 7).

Prior to diagnosing the presence of an anatomically univentricular heart, it is also necessary to exclude the possibility that there is a huge ventricular septal defect, with virtual absence of the apical ventricular septum. In this situation, however, despite the virtual absence of the septum, it is still possible to distinguish fine apical trabeculations on one side of the ventricular cavity, and coarse trabeculations on the other side. Having excluded these possibilities of gross hypoplasia of one or other ventricle, or a huge ventricular septal defect, it is then possible to identify anatomically univentricular hearts (Fig. 8). These malformations typically have double inlet to, and double outlet from, a solitary ventricle of indeterminate apical morphology. The apical trabeculations in such ventricles are uniformly coarse, being even coarser than those found in the morphologically right ventricle. The only septal structure present in such hearts is the muscular outlet septum, interposed between the subarterial outlets. Such solitary ventricles of

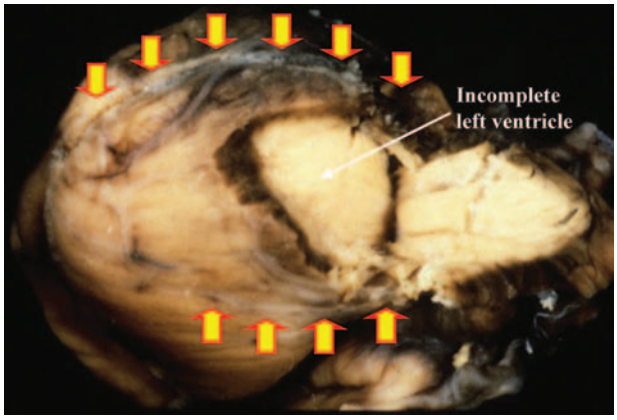


Figure 7.
In this specimen with hypoplasia of the left heart, dissection in the postero-inferior aspect of the ventricular mass between the delimiting coronary arteries (arrows) reveals the location of the incomplete left ventricle.

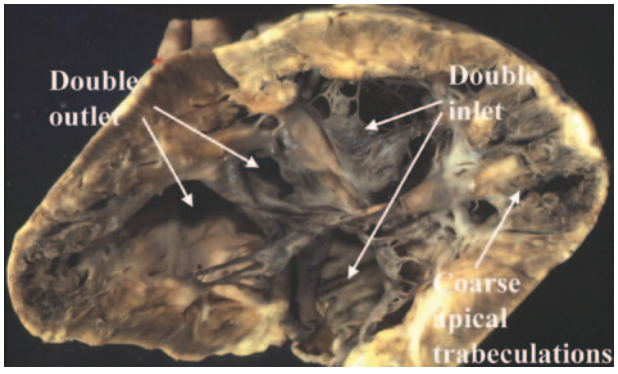


Figure 8.
In this specimen, opened in clam-like fashion, there is double inlet to, and double outlet from, a solitary ventricle. The apical trabeculations are much coarser than those of the right ventricle (Compare with Fig. 6). This particular example came from a patient with usual atrial arrangement.

indeterminate morphology are usually found in association with isomerism of the right atrial appendages, but can exist with usual atrial arrangement (Fig. 8). They should be anticipated to exist in the setting of mirror-imaged atrial arrangement. Most of the examples described thus far have had double inlet, but they can also exist with absence of one or other atrioventricular connection.

Analysis of the atrioventricular junctions

The essence of the normal heart is that each atrial vestibule is connected to its own ventricle, albeit that the muscular masses, other than at the site of the atrioventricular bundle, are separated by the fibrofatty insulating planes of the atrioventricular grooves. The atrioventricular junctions thus formed, in most instances, are guarded by separate right and left

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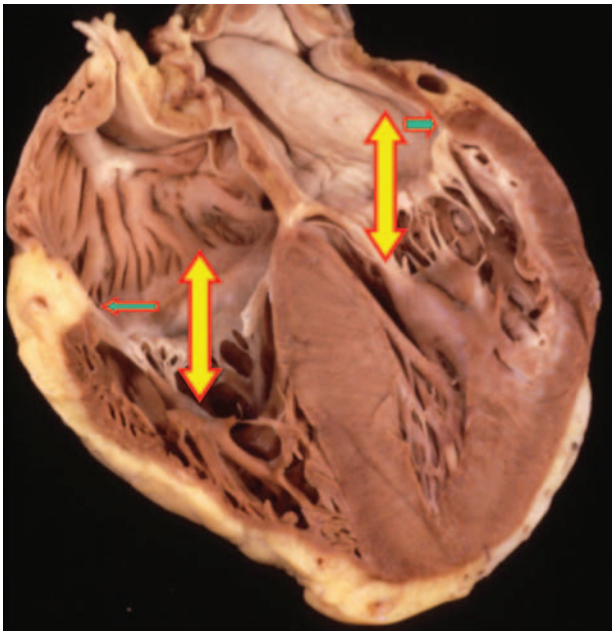


Figure 9.
This normal heart has been sectioned in "four-chamber" plane to show the concordant atrioventricular junctions (double headed arrows), with the fibro-fatty parietal junctions (green and red arrows) producing electrical insulation between the atrial and ventricular musculatures at all sites except the penetration of the atrioventricular conduction axis.

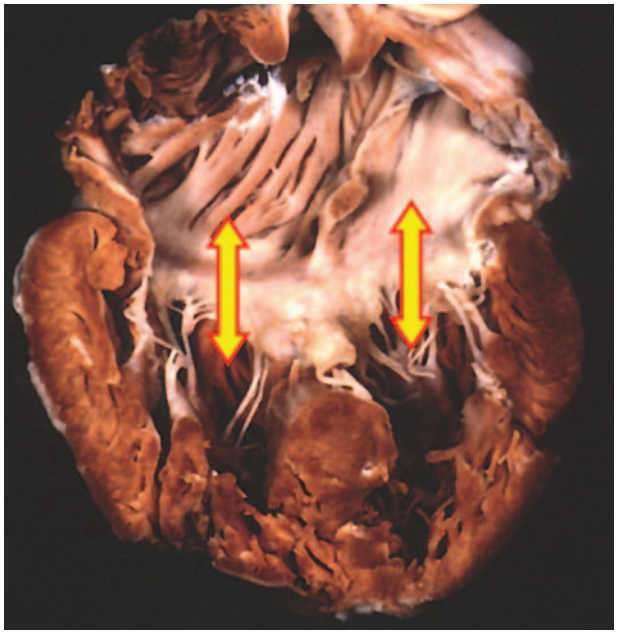


Figure 10.
This heart has an atrioventricular septal defect with common atrioventricular junction. The presence of a common atrioventricular valve does not disguise the fact that, like in the normal heart (see Fig. 9) there are concordant atrioventricular connections (double headed arrows).

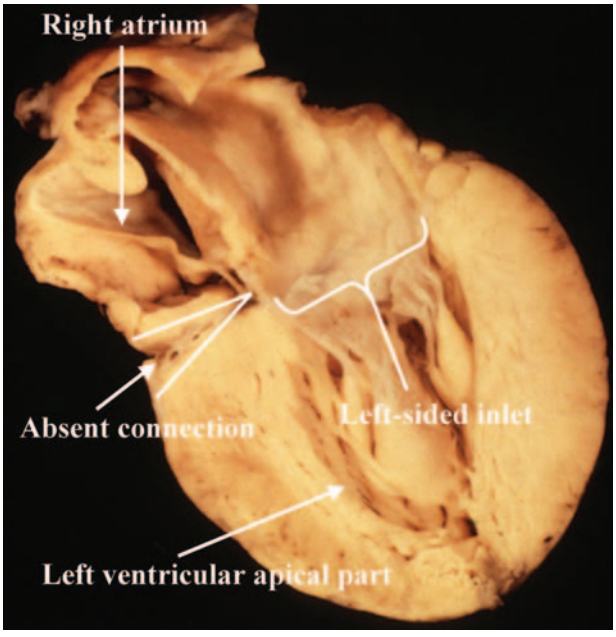


Figure 11.
This "four-chamber" section shows the morphology of absence of the right atrioventricular connection, seen here in "classical" tricuspid atresia.

atrioventricular valves (Fig. 9). Such biventricular connections, however, can also be found in the setting of a common atrioventricular valve (Fig. 10). In this setting, despite the presence of the common valve, it is still possible to recognise that the two atrial vestibules are each connected to their own morphologically discrete ventricles. Biventricular atrioventricular connections can be concordant, discordant, or ambiguous, the latter arrangement being found in the presence of isomeric atrial arrangements.

There are then three other possible arrangements at the atrioventricular junctions, each of which produces an anatomically univentricular connection. The first of these is double inlet ventricle, where both segments of atrial myocardium are connected to the same ventricle, which can be of left (Fig. 4), right (Fig. 6), or solitary and indeterminate (Fig. 8) morphology. The other anatomic situations producing univentricular connections are seen when one or other atrioventricular connection is absent. The absent connection can be right- (Fig. 11) or left- (Fig. 12) sided. Absence of one atrioventricular connection produces one variant of atrioventricular valvar atresia. These variants must be distinguished from imperforate atrioventricular valves. The imperforate valves are rarer than absence of one connection, and can be found with biventricular (Fig. 13) or double inlet (Fig. 14) atrioventricular connections. When found in the setting of biventricular atrioventricular connections, nonetheless, they produce functionally univentricular hearts (Fig. 13).

There is then one further variant of atrioventricular valvar atresia that produces a functionally univentricular heart. This is when one atrioventricular

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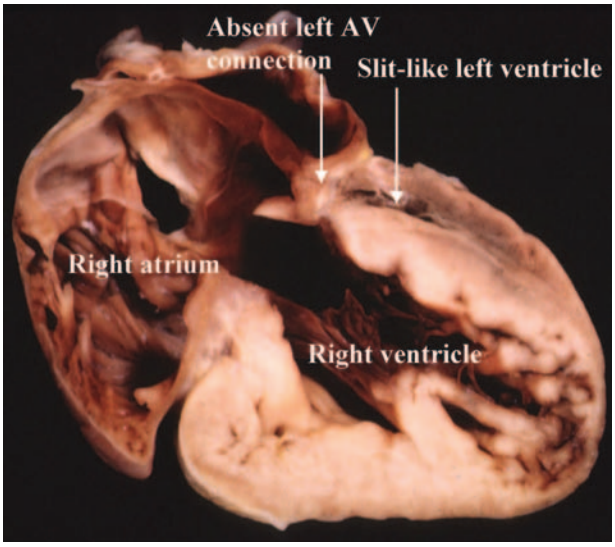


Figure 12.
In this heart, also shown in "four-chamber" section, there is absence of the left atrioventricular connection in the setting of hypoplasia of the left heart.

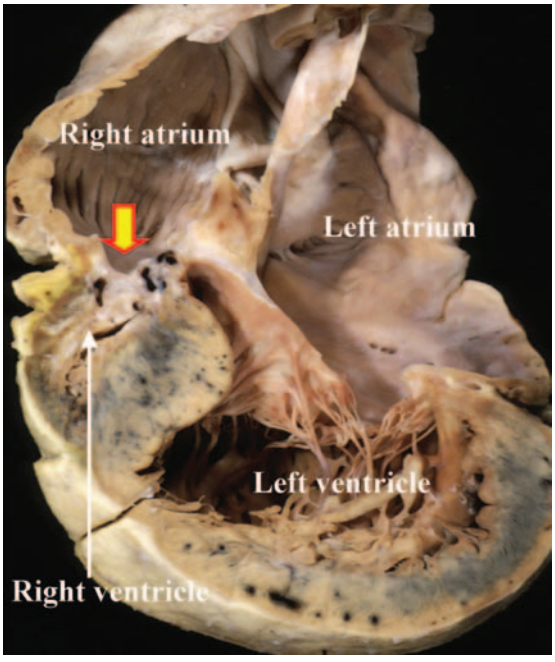


Figure 13.
The heart from a patient with pulmonary atresia with intact ventricular septum has been sectioned in "four-chamber" plane, showing an imperforate right atrioventricular valve (arrow) in the setting of concordant atrioventricular connections. Even though there are biventricular atrioventricular connections, the heart is functionally univentricular.

connection is absent, but the solitary atrioventricular valve is shared between the right and left ventricles. In this setting, the solitary atrioventricular valve will be of mitral or tricuspid (Fig. 15) morphology.

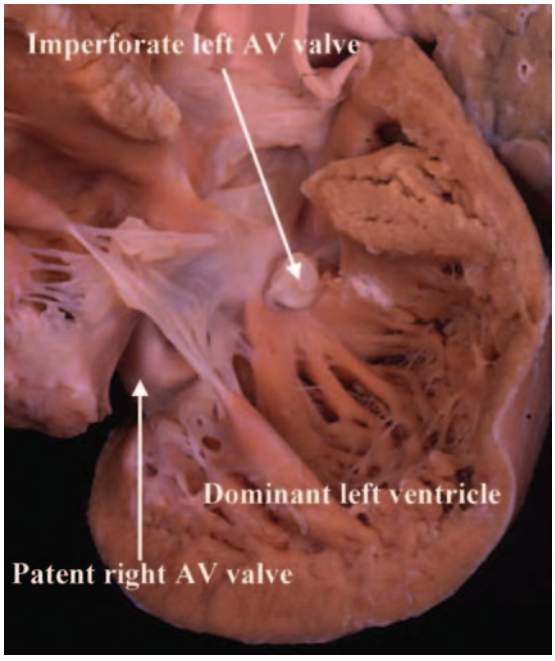


Figure 14.
There is an imperforate left atrioventricular (AV) valve in this heart, but in the setting of double inlet left ventricle.

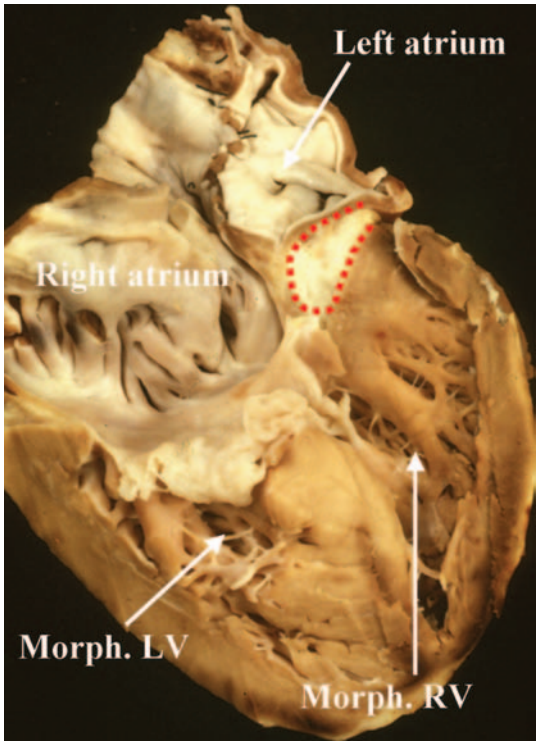


Figure 15.
In the heart illustrated by this "four-chamber" section, there is absence of the left atrioventricular connection (dotted red line). The right atrium, however, is connected to both ventricles by a straddling right valve. There is left-hand ventricular topology, so the straddling valve is of mitral morphology. This arrangement produces a uniatrial but biventricular atrioventricular connection. Morph. LV, RV – morphologically left and right ventricles.

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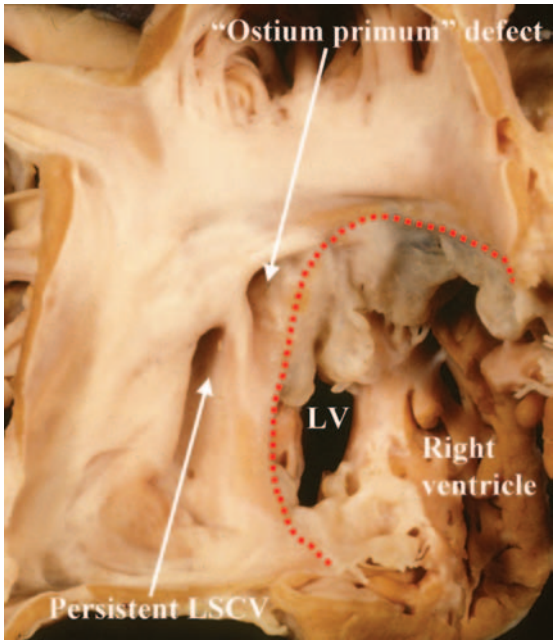


Figure 16.
This heart also has a uniatrial but biventricular atrioventricular connection, but this time because of gross malposition of the atrial septum in the setting of a common atrioventricular junction. The heart has been opened through the right side of the common junction (red dotted line) which drains the right atrium to both ventricles (LV – left ventricle). The left atrium exits through the "ostium primum" defect. Note also the presence of a persistent left superior caval vein (LSCV).

This uniatrial and biventricular connection of a solitary valve must be distinguished from common atrioventricular valve co-existing with malposition of the atrial septum (Fig. 16). In the latter situation, it is possible surgically to remove the atrial septum, and to partition the common atrioventricular valve so as to restore biventricular circulations. This is unlikely to be feasible with the uniatrial and biventricular connection, since it will not be possible adequately to share the straddling mitral or tricuspid valve between the ventricles.

Anatomic substrates of the functionally univentricular heart

The functionally univentricular heart is the arrangement in which the ventricular mass cannot be separated surgically so as to support separate pulmonary and systemic circulations. Anatomically univentricular hearts will self-evidently produce a functionally univentricular arrangement. These lesions are exceedingly rare. The majority of malformations giving the functionally univentricular arrangement will have one big and one small ventricle. It is not possible in the limited space available to discuss all these anomalies in detail, but we will summarise the salient

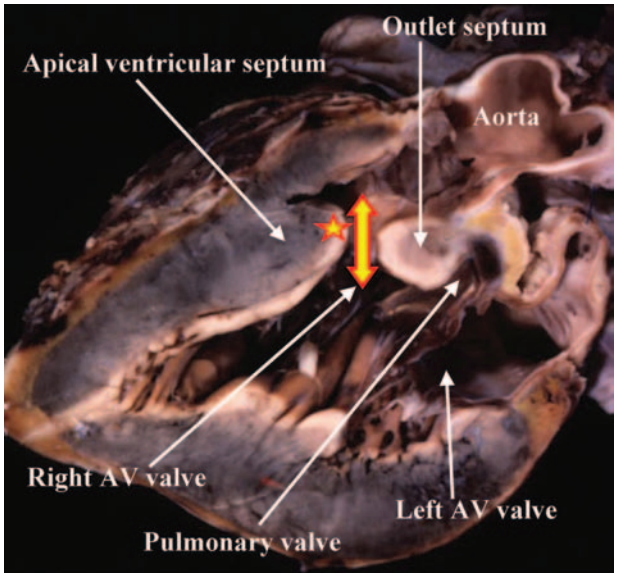


Figure 17.
This oblique long axis section shows the structure of double inlet left ventricle. Both atrioventricular (AV) valves enter the dominant left ventricle, which gives rise also to the pulmonary trunk, posterior to the apical ventricular septum, which separates the apical parts of the dominant and rudimentary ventricles, and carries the conduction tissues on its crest (star). The outlet septum separates the subpulmonary and subaortic outflow tracts, with the ventricular septal defect between the two septal structures (double headed arrow).

anatomical features of the major groups making up this important category.

Double inlet ventricle

Double inlet, as already discussed, can be found with a dominant left ventricle, a dominant right ventricle, or with a solitary and indeterminate ventricle. Most will have a dominant left ventricle. There has been controversy concerning the second chamber in such hearts. Some continue to argue that the small chamber is no more than an infundibulum.[3] Were this the case, the chamber would be separated from the dominant ventricle by the infundibular or "conal" septum. In fact, the infundibular septum separates the subarterial components of the ventricular outlets, and forms the roof of the ventricular septal defect seen in double inlet left ventricle. The apical part of the septum is the true ventricular septum, being irrigated by the perforating branches of the left coronary artery,[16] and carrying the ventricular conduction tissues on its crest (Fig. 17). This confirms the identification of the small chamber as the hypoplastic and incomplete right ventricle. It can be to the left or the right of the dominant left ventricle, but is always positioned antero-superiorly within the ventricular mass. With double inlet right ventricle, in contrast, the hypoplastic and its complete left ventricle is usually positioned

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postero-inferiorly and to the left, although it can be right-sided. Unlike the incomplete right ventricle, it is unusual for the hypoplastic left ventricle to support one of the arterial trunks. More usually it is no more than a blind-ending pouch, but one of unequivocally left morphology (Fig. 1).

Atrioventricular valvar atresia

As already explained, in most instances atrioventricular valvar atresia is the consequence of absence of one or other atrioventricular connection, as typically seen in "classical" tricuspid atresia (Fig. 11). In this setting, the dominant ventricle is of left morphology, while the right ventricle is incomplete and hypoplastic because of complete absent of the inlet component. Typically the hypoplastic right ventricle supports the pulmonary trunk (Fig. 18), but it can support the aorta, both arterial trunks, or rarely neither arterial trunk. Absence of the left atrioventricular connection most usually produces a variant of left heart hypoplasia (see below), but can also be found with a dominant left ventricle, when the incomplete and hypoplastic right ventricle is usually left-sided (Fig. 19). Atrioventricular valvar atresia due to an imperforate valve is much rarer, usually being seen as part of hypoplasia of the left heart, or else with pulmonary atresia and intact ventricular septum (Fig. 13), or Ebstein's malformation.

Straddling atrioventricular valves

Depending on the degree of override of the atrioventricular junction, hearts with straddling atrioventricular valves can produce either functionally univentricular or biventricular hearts. When we used to disqualify chambers from ventricular status according to whether or not they possessed an inlet component, these hearts produced great problems in classification. Now that we describe all chambers as ventricles according to the morphology of their apical parts, this no longer poses a problem. And the atrioventricular connection does, indeed, become functionally univentricular when all of one junction, along with more than half of the overriding junction, is supported by the dominant ventricle. Even

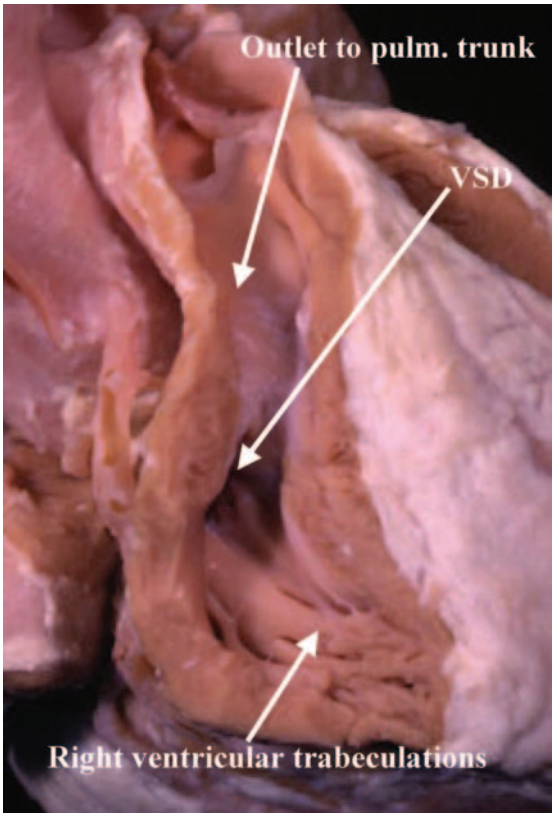


Figure 18.
The anterior rudimentary and incomplete right ventricle from a case of "classical" tricuspid atresia. Because of absence of the right atrioventricular connection, the ventricle lacks totally its inlet component, possessing an apical part with right ventricular trabeculations, and an infundibulum supporting, in the heart, the pulmonary trunk.

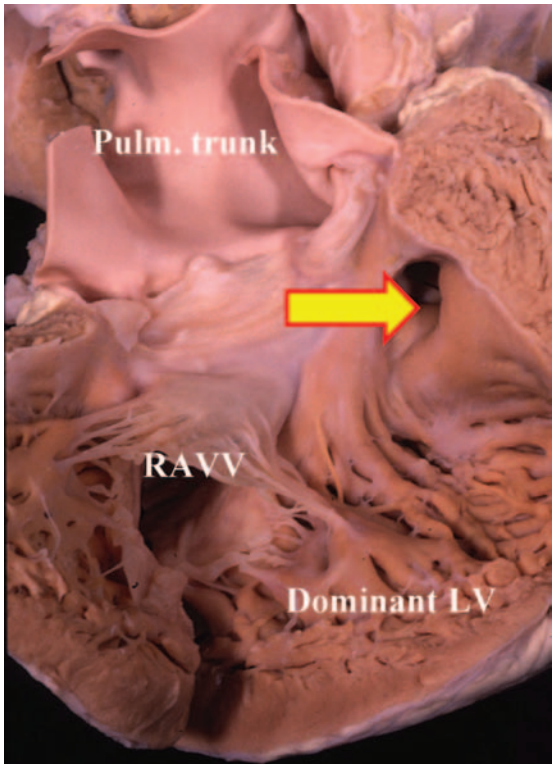


Figure 19.
The dominant left ventricle from a heart with usual atrial arrangement and absence of the left atrioventricular connection. The right-sided atrioventricular valve (RAVV) is of mitral morphology, and the ventricular septal defect (arrow) leads to the left-sided rudimentary and incomplete right ventricle.

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in situations where the connection is deemed to be biventricular, nonetheless, many centres nowadays will opt for functionally univentricular repair.

Hypoplasia of the left heart

All of the hearts falling into this category produce a functionally univentricular arrangement. They are the topic of the greater part of this supplement, and their morphology is discussed in detail elsewhere (see pp 13–21).

Hypoplasia of the right heart

Pulmonary atresia with an intact ventricular septum is the prototype of the malformation that possesses an anatomically biventricular heart that may or may not be functionally univentricular, depending on the degree of hypoplasia of the complete but abnormal right ventricle. The extent of hypoplasia itself depends on the degree and location of mural hypertrophy.[17] If the mural hypertrophy is minimal, and the pulmonary valve is imperforate, then the right ventricle is usually capable of supporting the pulmonary circulation (Fig. 20). If mural hypertrophy is extreme, and obliterates the cavities of both the apical parts and outlet of the right ventricle, then the heart will be functionally univentricular (Fig. 21). In this setting, fistulous communications from the right ventricle to the coronary arteries can also compromise the performance of the dominant left ventricle.[18] Between these extremes are those examples with so-called "bipartite" right ventricular cavities. According to the extent of mural overgrowth of the apical part, these right ventricles may or may not be capable of supporting the pulmonary circulation.

Ventricular imbalance with deficient septation

Completing the heterogeneous made-up of hearts with a potentially functional univentricular circulation are those hearts with either an atrioventricular or ventricular septal defect. When the left ventricle is grossly hypoplastic in this setting, some would consider these lesions as representing part of the spectrum of hypoplasia of the left heart. As explained elsewhere in our supplement, however, the "classical" variants of left heart hypoplasia are found in the setting of an intact ventricular septum. It is moot, therefore, whether hearts with ventricular or atrioventricular septal deficiencies should be included in the "syndrome". It is an undeniable fact, nonetheless, that the left ventricle in this situation can be sufficiently hypoplastic to dictate the need for functionally univentricular repair.

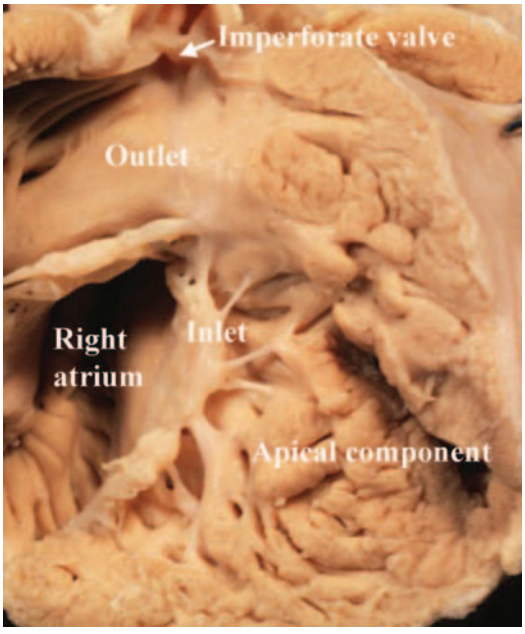


Figure 20.
In this heart from a patient with pulmonary atresia and intact ventricular septum, there is minimal mural hypertrophy, so that the ventricular cavity has three patent and well-formed components. Patients of this kind, if seen during their life, are usually suitable for biventricular repair.

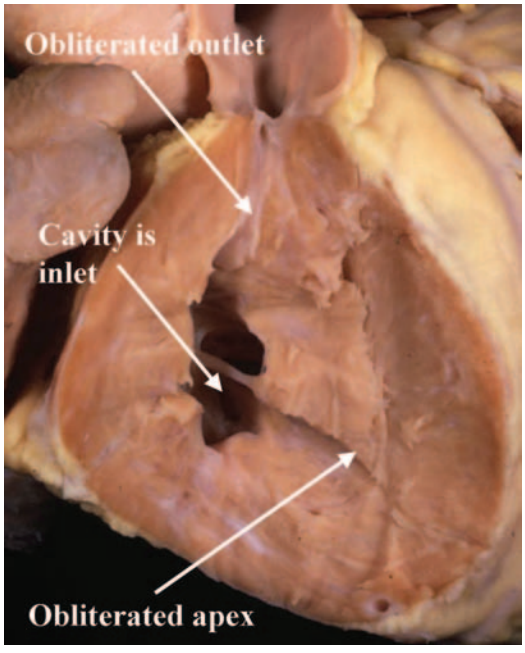


Figure 21.
This heart is also from a patient with pulmonary atresia and intact ventricular septum (compare with Fig. 20). In this specimen, however, gross mural hypertrophy has obliterated the cavities of the ventricular apical and outlet components. The cavity is, in effect, represented only by the inlet component. The heart has biventricular atrioventricular connections, but had it been seen during life, the patient would almost certainly have required a functionally univentricular repair.

https://doi.org/10.1017/S1047951104006237 Published online by Cambridge University Press

Conclusions

The arguments that continue concerning the suitability of the terms "univentricular heart", or "single ventricle", to describe a subset of congenitally malformed hearts can be defused by the application of simple logic. In anatomical terms, there are very few patients that have a solitary chamber within their ventricular mass. Most of those currently described in this fashion have one big and one small ventricle, the small ventricle usually lacking one or more of its normal components. In these hearts, it is the atrioventricular connection that is truly univentricular, either because of double inlet, or because one or other of the normal atrioventricular junctions is absent.[10,11] Other hearts with one big and one small ventricle achieve this arrangement because of atresia or critical stenosis of an arterial valve, almost always in the setting of an intact ventricular septum. Still others have straddling and overriding of one or other atrioventricular valve. Patients with these various lesions are now unified because of their likely surgical treatment, the endpoint being univentricular rather than biventricular support of the circulation. The nomenclature, therefore, is solved by describing them for what they are, functionally univentricular hearts.

References

1. Edwards JE. Discussion. In: Davila JC (ed.). 2nd Henry Ford Hospital International Symposium on Cardiac Surgery. New York, Appleton-Century-Crofts, 1977, p 242.
2. Anderson RH, Becker AE, Tynan M, Macartney FJ, Rigby ML, Wilkinson JL. The univentricular atrioventricular connection: getting to the root of a thorny problem. Am J Cardiol 1984; 54: 822–828.
3. Van Praagh R. Nomenclature and classification: Morphologic and segmental approach to diagnosis. In: Moller JH, Hoffman JIE (eds). Pediatric Cardiovascular Medicine. New York, Churchill Livingstone, 2000, pp 275–288.
4. Anderson RH, Becker AE, Macartney FJ, Shinebourne EA, Wilkinson JL, Tynan MJ. Is "tricuspid atresia" a univentricular heart? Pediatr Cardiol 1979; 1: 51–56.
5. Bharati S, Lev M. The concept of tricuspid atresia complex as distinct from that of the single ventricle complex. Pediatr Cardiol 1979; 1: 57–62.
6. Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax 1971; 26: 240–248.
7. de Leval MR, Kilner P, Gewillig M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. J Thorac Cardiovasc Surg 1988; 96: 682–695.
8. Barlow A, Pawade A, Wilkinson JL, Anderson RH. Cardiac anatomy in patients undergoing the Fontan procedure. Ann Thorac Surg 1995; 60: 1324–1330.
9. Van Praagh R, David I, Wright GB, Van Praagh S. Large RV plus small LV is not single RV. Circulation 1980; 61: 1057–1058.
10. Anderson RH, Ho SY. Continuing medical education. Sequential segmental analysis – description and categorization for the millenium. Cardiol Young 1997; 7: 98–116.
11. Anderson RH, Baker EJ, Redington AN. Can we describe structure as well as function when accounting for the arrangement of the ventricular mass. Cardiol Young 2000, 10: 247–260.
12. Keeton BR, Macartney FJ, Hunter S, Mortera C, Rees P, Shinebourne EA, Tynan M, Wilkinson JL, Anderson RH. Univentricular heart of right ventricular type with double or common inlet. Circulation 1979; 59: 403–411.
13. Brandt PWT. Cineangiography of atrioventricular and ventriculoarterial connexions. In: Godman MJ (ed.). Paediatric Cardiology Volume 4. Edinburgh, Churchill Livingstone, 1981, pp 199–220.
14. Bjork VO, Olin CL, Bjarke BB, Thoren LA. Right atrial-right ventricular anastomosis for correction of tricuspid atresia. J Thorac Cardiovasc Surg 1979; 77: 452–458.
15. Kurosawa H, Imai Y, Fukuchi S, Sawatari K, Koh Y, Nakazawa M, Takao A. Septation and Fontan repair of univentricular atrioventricular connection. J Thorac Cardiovasc Surg 1990; 99: 314–319.
16. Hosseinpour AR, Anderson RH, Ho SY. The anatomy of the septal perforating arteries in normal and congenitally malformed hearts. J Thorac Cardiovasc Surg 2001; 121: 1046–1052.
17. Anderson RH, Anderson C, Zuberbuhler JR. Further morphologic studies on hearts with pulmonary atresia and intact ventricular septum. Cardiol Young 1991; 1: 105–114.
18. Dyamenali U, Hanna B, Sharratt GP. Pulmonary atresia with intact ventricular septum: management of the coronary arterial anomalies. Cardiol Young 1997; 7: 105–113.

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