1 Terminology

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Introduction

It might reasonably be thought that those who diagnose and treat patients with congenitally malformed hearts would, by now, have reached consensus concerning the most appropriate way of describing the malformations with which they are confronted. It is certainly the case that nomenclature is far less contentious now than in the previous millennium. It would be a brave person, nonetheless, who stated that the field of description and categorization was now fully resolved. It is not our intention, in this chapter, to extend these polemics. Rather, we describe our own system for description, leaving the readers to decide whether this is satisfactory for their needs. By and large, there is no right or wrong way of describing hearts, simply different ways.[1,2] Even these different ways have been mitigated to considerable extent by the cross-mapping of existing systems.[3] The ongoing differences should now be resolved simply by describing the abnormal anatomy as it is observed.

The need for a standardized approach reflects the fact that the number of individual lesions that can coexist within malformed hearts is considerable. Add to this the possibilities for combinations of lesions, and the problem of providing "pigeon holes" for each entity becomes immense. Straightforward lesions, such as septal deficiencies or valvar stenoses, are typically encountered in hearts that are otherwise structurally normal. It is when the hearts containing the lesions are themselves built in grossly abnormal fashion that difficulties are produced. If these alleged complex lesions are approached in a simple and straightforward fashion, none need be difficult to understand and describe.

The simplicity is provided by recognizing that the heart has three basic building blocks, namely the atriums, ventricular mass, and arterial trunks (Fig. 1.1). The system for description and categorization based on recognition of the limited potential for variation in each of these cardiac segments was developed independently in the 1960s by two groups: one based in the United States and led by Richard Van Praagh,[1] and the other from Mexico City, headed by Maria Victoria de la Cruz.[4] Both of these systems concentrated on the different topologic arrangements of the components within each cardiac segment. When Van Praagh and colleagues[5,6] introduced the concept of concordance and discordance between atriums and ventricles, they were concerned primarily with the harmony or disharmony to be found between the topologic arrangements of the atrial and ventricular components. At this time, they placed less emphasis for description on the fashion in which the atrial and ventricular chambers were joined together across the atrioventricular junctions. A similar approach, concentrating on arterial relationships, had been taken by de la Cruz et al.[7] when they formulated their concept of arterioventricular concordance and discordance. These approaches were understandable because it was often difficult, at that time, to determine precisely how the adjacent structures were linked together.

All was changed by the advent of cross-sectional echocardiography. Since the mid-1970s, it has been possible with precision to determine how atriums are, or are not, joined to ventricles, and similarly to establish the precise morphology found at the ventriculoarterial junctions. Our preferred system evolved concomitantly with the development of echocardiography and concentrates on the variations possible across the atrioventricular and ventriculoarterial junctions. We call this system sequential segmental analysis (see Fig. 1.1). In making such analysis, we do not ignore the segments themselves. Indeed, junctional connections cannot be established without knowledge of segmental topology.

Our system, throughout its evolution,[8–12] has followed the same basic and simple rules. From the outset, we have formulated our categories on the basis of recognizable anatomic facts, avoiding any speculative embryologic assumptions. Again, from the start, we have emphasized the features of the morphology of the cardiac components, the way they are joined or not joined together, and the relations between them, as three different facets of the cardiac make-up. The clarity of the system depends upon its design. Some argue that brevity is an important feature and have constructed formidable codifications to achieve this aim.[13] However, in the final analysis, clarity is more important than brevity. Therefore we do not shy from using words to replace symbols, even if this requires several words. Wherever possible, we strive to use words that are as meaningful in their systematic role as in their everyday usage. In our desire to achieve optimal clarity, we have made changes in our descriptions over the years, most notably in our use of the term "univentricular heart."[14,15] We make no apologies for these changes because their formulation, in response to valid criticisms, has eradicated initially illogical points from our system to its advantage. It is our belief that the system now advocated is entirely logical, and we hope it is simple.

Basic Concepts of Sequential Segmental Analysis

The system we advocate depends first upon the establishment of the arrangement of the atrial chambers. Thereafter, attention is concentrated on the anatomic nature of the junctions

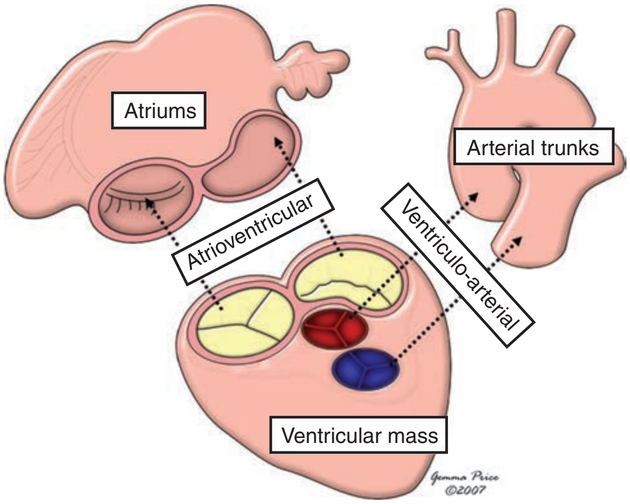

Fig. 1.1 The essence of sequential segmental analysis depends on recognition of the topologic arrangement of the three cardiac segments and combines this with analysis of the fashions in which the segments are joined, or are not joined, to each other.

between the atrial myocardium and the ventricular myocardial mass. This feature, which we describe as a type of connection, is separate from the additional feature of the morphology of the valve or valves that guard the junctions. There are two junctions in the normally constructed heart, and usually they are guarded by two separate valves. The two atrioventricular junctions can be guarded, on occasion, by a common valve. If we are to achieve this analysis of the atrioventricular junctions, we must also determine the structure, topology, and relationships of the chambers within the ventricular mass. Having dealt with the atrioventricular junctions, the ventriculoarterial junctions are also analyzed in terms of the way the arterial trunks are joined to the ventricular mass and the morphology of the arterial valves guarding their junctions. Separate attention is directed to the morphology of the outflow tracts and to the relationships of the arterial trunks. A catalog is made of all associated cardiac and, where pertinent, noncardiac, malformations. Included in this final category are such features as the location of the heart, the orientation of its apex, and the arrangement of the other thoracic and abdominal organs.

Implicit in the system is the ability to distinguish the morphology of the individual atriums and ventricles and to recognize the types of arterial trunk taking origin from the ventricles. This is not as straightforward as it may seem; often, in congenitally malformed hearts, these chambers or arterial trunks may lack some of the morphologic features that most obviously characterize them in the normal heart. The most obvious feature of the morphologically left atrium in the normal heart is the connection to it of the pulmonary veins. In hearts with totally anomalous pulmonary venous connection, these veins connect in extracardiac fashion. In spite of this, it is still possible to identify the left atrium. It is considerations of this type that prompted the concept we use for recognition of the cardiac chambers and great arteries. Dubbed by Van Praagh and his colleagues the "morphologic method"[16] and based on the initial work of Lev,[17] the principle states that structures should be recognized in terms of their own intrinsic morphology and that one part of the heart, which is itself variable, should not be defined on the basis of another variable structure. When this eminently sensible concept is applied to the atrial chambers, the connections of the great veins are obviously disqualified as markers of morphologic rightness or leftness because, as discussed previously, the veins do not always connect to their anticipated atriums. Fortunately, there is another component of the atrial chambers that, in our experience, has been almost universally present and that, on the basis of the morphology of its junction with the remainder of the chambers, has enabled us always to distinguish between morphologically right and left atriums. This is the appendage. The morphologically right appendage has the shape of a blunt triangle and joins over a broad junction with the remainder of the atrium. The junction is marked externally by the terminal groove and internally by the terminal crest. Its most significant feature is that the pectinate muscles lining the appendage extend around the parietal atrioventricular junction to reach the cardiac crux (Fig. 1.2A).

The morphologically left appendage, in contrast, is much narrower and tubular. It has a narrow junction with the remainder of the atrium, the junction being marked neither by a terminal groove nor by a muscular crest. The pectinate muscles are confined within the morphologically left appendage, with the walls of the remainder of the atrium being smooth as they extend to the cardiac crux (see Fig. 1.2B).

The morphologic method also shows its value when applied to the ventricular mass, which extends from the atrioventricular to the ventriculoarterial junctions. Within the ventricular mass as thus defined, there are almost always two ventricles. Description of ventricles, no matter how malformed they may be, is facilitated if they are analyzed as possessing three components. The first is the inlet, extending from the atrioventricular junction to the distal attachment of the atrioventricular valvar tension apparatus. The second part is the apical trabecular component. The third is the outlet component, supporting the leaflets of the arterial valve (Fig. 1.3).

Of these three components, the apical trabecular component is most universally present in normal, as well as in malformed and incomplete, ventricles. Furthermore, it is the pattern of the apical trabeculations that differentiates morphologically right from left ventricles (see Fig. 1.3). This is the case even when the apical components exist as incomplete ventricles, lacking either inlet or outlet components, or sometimes both of these components (Fig. 1.4).

When the morphology of individual ventricles is identified in this fashion, all hearts with two ventricles can be analyzed according to the way that the inlet and outlet components are shared between the apical trabecular components. To fully describe any ventricle, account must also be taken of its size. It is necessary further to describe the way that the two ventricles themselves are related within the ventricular mass. This feature is described in terms of ventricular topology because two basic patterns are found that cannot be changed without physically taking apart the ventricular components and reassembling them. The two patterns are mirror images of each other. They can be conceptualized in terms of the way that, figuratively speaking, the palmar surface of the hands can be placed upon the septal surface of the morphologically right ventricle. In the

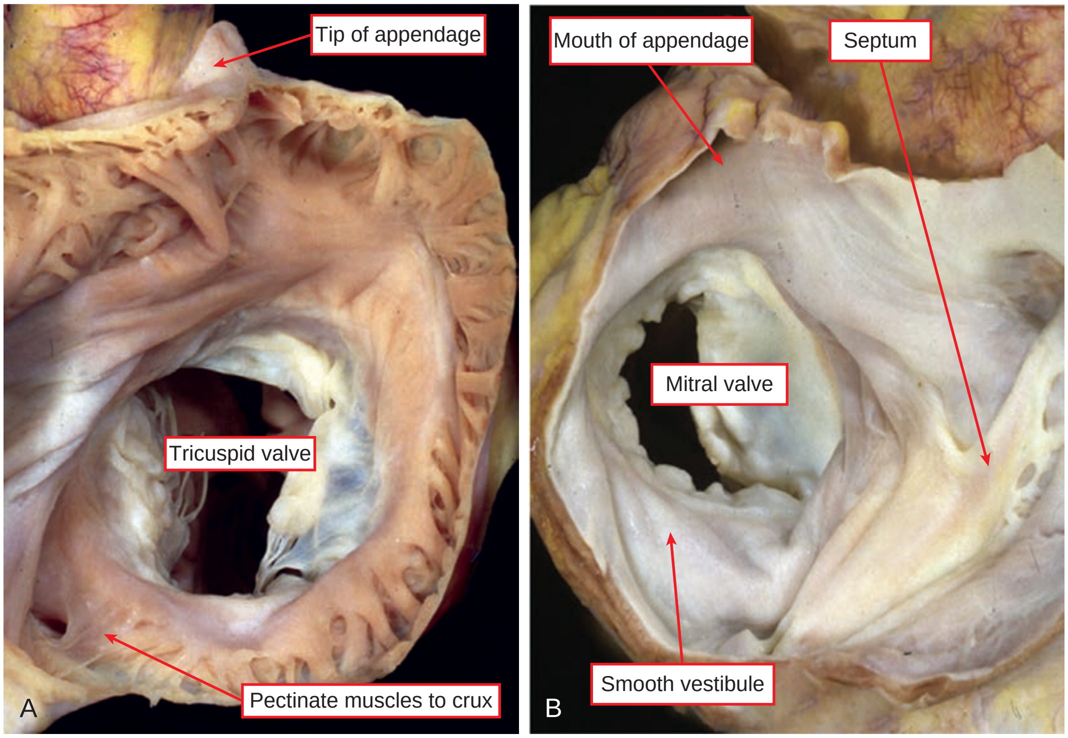

Fig. 1.2 (A) Short-axis view of the right atrioventricular junction from above, the atrium having been opened with a cut parallel to the atrioventricular junction, and with subsequent reflection of the wall of the appendage. Note that the pectinate muscles within the appendage extend all around the vestibule of the tricuspid valve. (B) Short-axis view of the left atrioventricular junction photographed from above from the same heart. The pectinate muscles are confined within the tubular appendage, so that the inferior wall of the atrium is smooth. This contains the coronary sinus within the morphologically left atrioventricular junction. Note also the typical appearance of the morphologically left side of the septum.

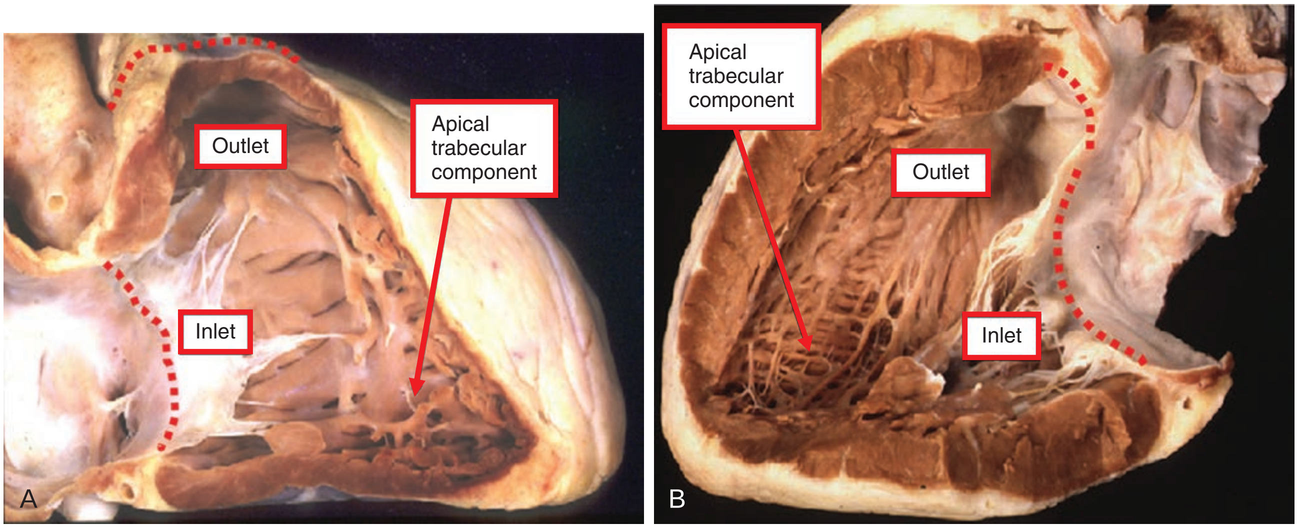

Fig. 1.3 (A) Three component parts of the morphologically right ventricle, which extends from the atrioventricular to the ventriculoarterial junctions *(dotted lines)*. The coarse apical trabeculations are the most constant of these features. (B) Three component parts of the morphologically left ventricle of the same heart. The ventricular cavity again extends from the atrioventricular to the ventriculoarterial junctions *(dotted lines)*. The fine apical trabeculations are its most constant feature.

morphologically right ventricle of the normal heart, irrespective of its position in space, only the palmar surface of the right hand can be placed on the septal surface such that the thumb occupies the inlet and the fingers fit into the outlet (Fig. 1.5).

Therefore the usual pattern can be described as right hand ventricular topology.[18] The other pattern, the mirror image of the right hand prototype, is described as left hand ventricular topology. In this left hand pattern, seen typically in the mirror-imaged normal heart, or in the variant of congenitally corrected transposition found with usual atrial arrangement, it is the palmar surface of the left hand that fits on the septal surface of the morphologically right ventricle with the thumb in the inlet and the fingers in

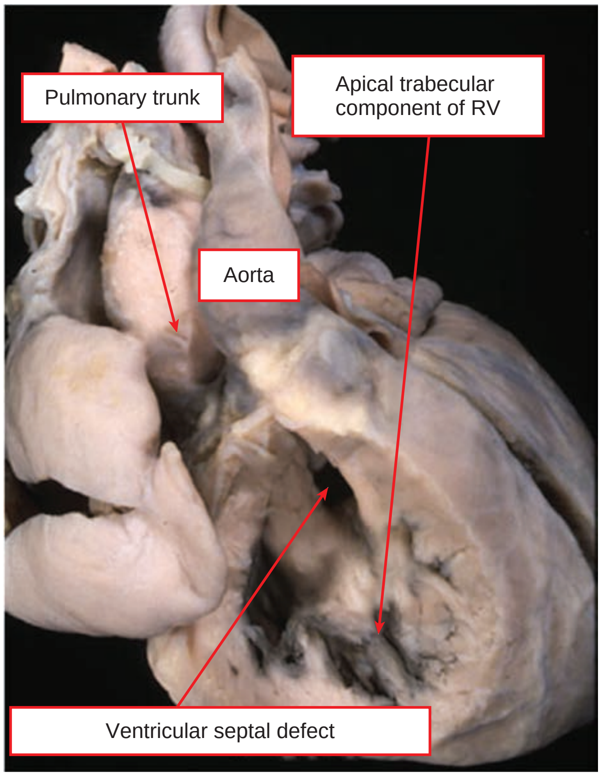

Fig. 1.4 Heart illustrating a double inlet to, and double outlet from, a dominant left ventricle. The aorta and pulmonary trunk are seen arising in parallel fashion from the left ventricle, with the aorta anterior and to the left. However, on the anterior and right-sided shoulder of the dominant left ventricle, there is still a second chamber to be seen, fed through a ventricular septal defect. This chamber is the apical trabecular component of the right ventricle (RV), identified because of its coarse trabeculations.

the outlet. This is the essence of left hand topology, or the "l-ventricular loop" (see Fig. 1.5). These two topologic patterns can always be distinguished irrespective of the location occupied in space by the ventricular mass itself. Therefore a left hand pattern of topology is readily distinguished from a ventricular mass with right hand topology in which the right ventricle has been rotated to occupy a left-sided position. Component make-up, trabecular pattern, topology, and size are independent features of the ventricles. On occasion, all may need separate description to remove any potential for confusion.

Only rarely will hearts be encountered with a solitary ventricle. Sometimes this may be because a right or left ventricle is so small that it cannot be recognized with usual clinical investigatory techniques. Nonetheless, there is a third pattern of apical ventricular morphology that is found in hearts possessing a truly single ventricle. This is when the apical component is of neither right nor left type but is very coarsely trabeculated and crossed by multiple large muscle bundles. Such a solitary ventricle has an indeterminate morphology (Fig. 1.6).

Analysis of ventricles on the basis of their apical trabeculations precludes the need to use illogically the terms "single ventricle" or "univentricular heart" for description of those hearts with one big and one small ventricle. These hearts may produce a functionally univentricular arrangement, but all chambers that possess apical trabecular components can be described as ventricles, be they big or small and be they incomplete or complete. Any attempt to disqualify such chambers from ventricular state must lead to a system that is artificial. Only hearts with a truly solitary ventricle need be described as univentricular, albeit that the connections of the atrioventricular junctions can be univentricular in many more hearts.

When determining the morphology of the great arteries, no intrinsic features enable an aorta to be distinguished from a pulmonary trunk or from a common or solitary arterial trunk. Nonetheless, the branching pattern of the trunks themselves is sufficiently characteristic to permit these distinctions (Fig. 1.7).

The aorta gives rise to at least one coronary artery and the bulk of the systemic arteries. The pulmonary trunk gives rise directly to both, or one or other, of the pulmonary arteries. A common trunk supplies directly the coronary, systemic, and pulmonary arteries. A solitary arterial trunk exists in the absence of the proximal portion of the pulmonary trunk. In such circumstances, it is impossible to state with certainty whether the persisting trunk is common or aortic. Even in the rare cases that have transgressed one of these rules, examination of the overall branching pattern has always permitted us to distinguish the nature of the arterial trunk.

Atrial Arrangement

The cornerstone of any system of sequential analysis must be accurate establishment of atrial arrangement because this is the starting point for subsequent analysis. When arrangement of the atriums is assessed according to the morphology of the junction of the appendages with the rest of the atriums,[19] There are only four possible patterns of arrangement (Fig. 1.8) because all hearts have two atrial appendages, each of which can only be morphologically left or right.

The most common is the usual arrangement, also called situs solitus, in which the morphologically right appendage is right-sided and the morphologically left appendage is left-sided. The second arrangement, which is very rare, is the mirror image of the usual. It is often called situs inversus, even though the atrial chambers are not upside down. In these two arrangements, the appendages are lateralized, with the morphologically right appendage being to one side, and the morphologically left appendage to the other. The two other arrangements do not show such lateralization. Instead, there is isomerism of the atrial appendages. In these patterns, the two appendages are mirror images of each other, with morphologic characteristics at their junctions with the rest of the atriums on both sides of either right type or left type.

Recognition of Atrial Arrangement

The arrangement of the appendages, ideally, is recognized by direct examination of the extent of the pectinate muscles

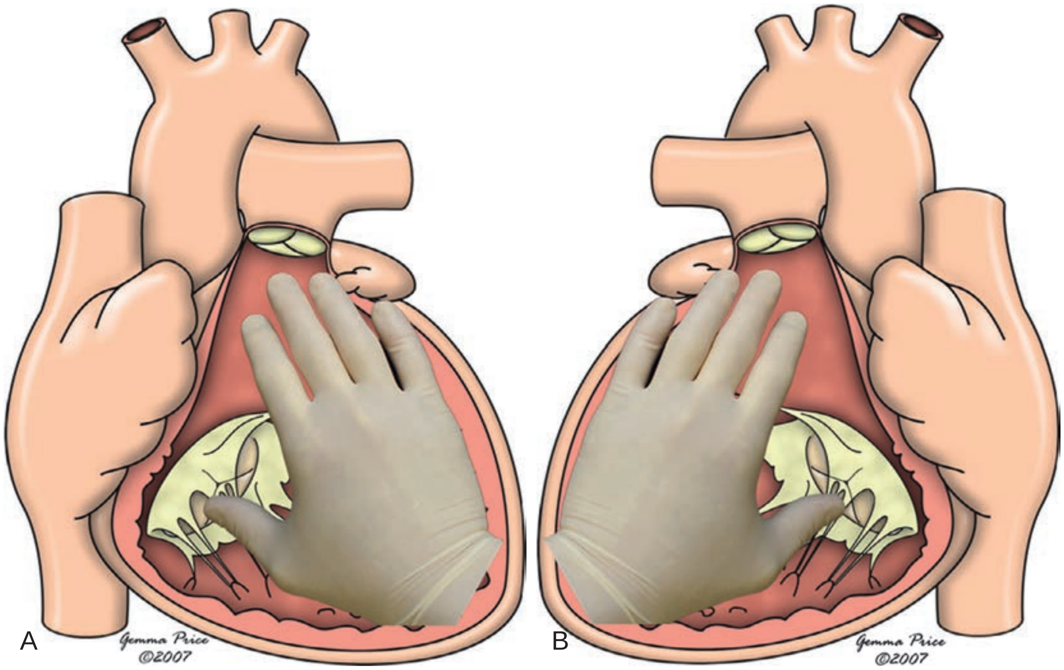

Fig. 1.5 Diagram showing how the palmar surface of the right hand can be placed on the septal surface of the normal morphologically right ventricle with the thumb in the inlet component and the fingers extending into the ventricular outlet. (A) The essence of right hand ventricular topology, also known as a d-ventricular loop. The palmar surface of the left hand fits in comparable fashion within the morphologically left ventricle, but the right hand is taken as the arbiter for the purposes of categorization. (B) The mirror-imaged normal heart. In this setting, the palmar surface of the left hand can be placed on the septal surface of the morphologically right ventricle with the thumb in the inlet and the fingers in the outlet.

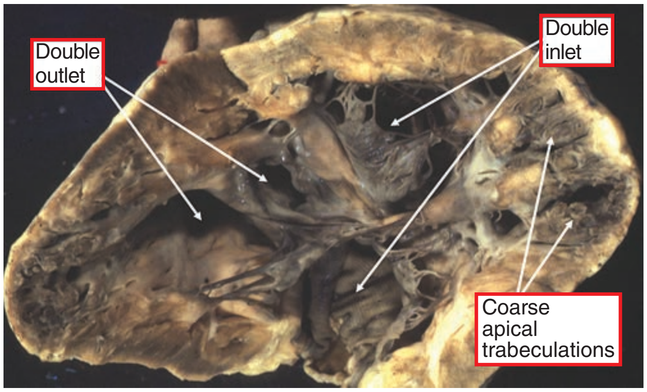

Fig. 1.6 Heart opened in clamshell fashion to show that both atrioventricular valves enter the same ventricular chamber, which also gives rise to both outflow tracts. We were unable to find a second ventricular chamber in this example. The exceedingly coarse apical trabeculations and the absence of the second chamber identify this heart as having a solitary ventricle of indeterminate morphology. This is the only true "single" ventricle.

round the vestibules (see Fig. 1.2). It has been questioned for some time as to whether these features can be distinguished in the clinical setting. With modern-day equipment, it is our belief that the arrangements should now be recognizable using cross-sectional echocardiography, particularly from the transesophageal window. The extent of the pectinate muscles can be demonstrated by using computed tomography. However, in most clinical situations, it is rarely necessary to rely only on direct identification. This is because the morphology of the appendages is almost always in harmony with the arrangements of the thoracic and abdominal organs. In patients with lateralized arrangements, that is, the usual and mirror-imaged patterns, it is exceedingly rare for there to be disharmony between the location of the organs (Fig. 1.9).

When the appendages are isomeric, in contrast, usually the abdominal organs are typically jumbled up, although the lungs and bronchuses are typically isomeric (Fig. 1.10).

Even when there is abdominal heterotaxy, the lungs and bronchial tree are almost always symmetric. It is rare for the bronchial arrangement to show disharmony with the morphology of the appendages. The presence of isomerism therefore can almost always be inferred from the bronchial anatomy. The morphologically left bronchus is long. It branches only after it has been crossed by its accompanying pulmonary artery, making the bronchus hyparterial. In contrast, the morphologically right bronchus is short and is crossed by its pulmonary artery after it has branched, giving an eparterial pattern of branching. The four patterns of bronchial branching are almost always in harmony with the arrangement of the atrial appendages. Similar inferences to those provided from bronchial arrangement can also usually be obtained noninvasively by using cross-sectional ultrasonography to image the abdominal great vessels. These vessels bear a distinct relation to each other, and to the spine, which generally reflects bodily arrangement, although not as accurately as does bronchial anatomy. The vessels can be distinguished ultrasonically according to their pattern of

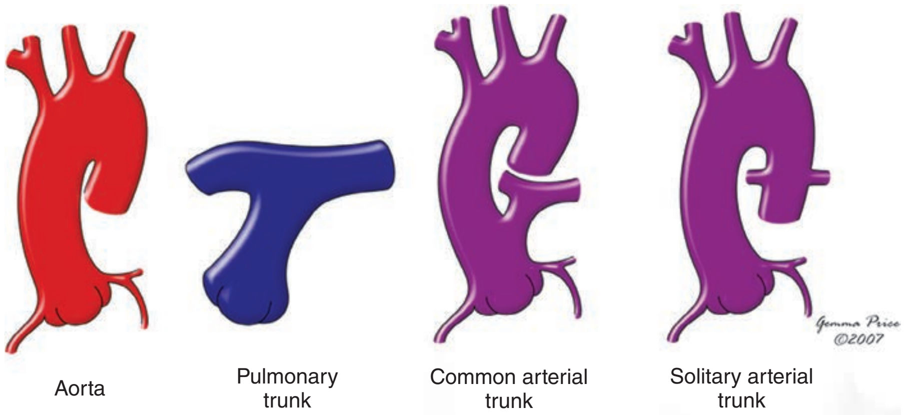

Fig. 1.7 The branching pattern of arterial trunks permits their distinction. The solitary arterial trunk is described when the intrapericardial pulmonary arteries are absent because in this setting it is impossible to determine, had they been present, whether they would have taken origin from the heart, making the trunk an aorta, or from the trunk itself, in which case there would have been a common arterial trunk with pulmonary atresia.

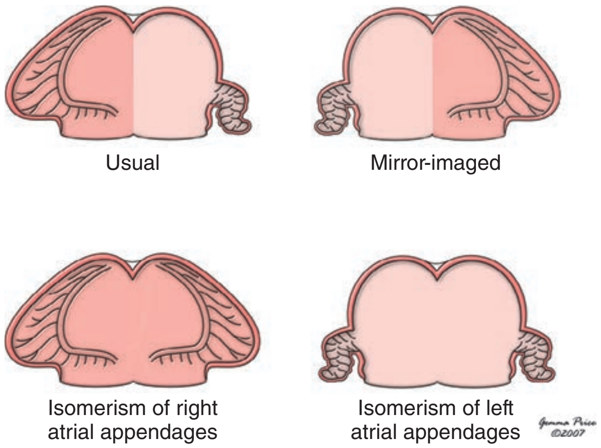

Fig. 1.8 When analyzed on the basis of the extent of the pectinate muscles relative to the atrioventricular vestibules (see Fig. 1.2), there are only four possible ways in which the two atrial appendages can be arranged. However, note that the venoatrial connections can show marked variation, particularly in the isomeric settings, also known collectively as visceral heterotaxy.

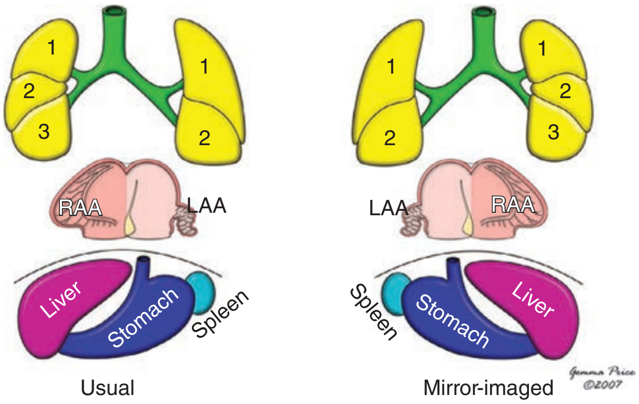

Fig. 1.9 Usual and mirror-imaged arrangements of the organs, which are lateralized. Almost always there is harmony between the arrangement of the right and left atrial appendages and the remaining thoracoabdominal organs. The numbers show the three lobes of the morphologically right and the two lobes of the morphologically left lungs. *LAA,* Left atrial appendage; *RAA,* right atrial appendage.

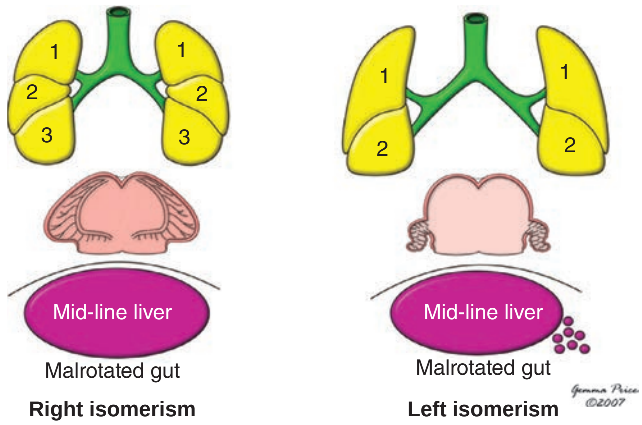

Fig. 1.10 Typical features of the thoracoabdominal organs in so-called visceral heterotaxy. The abdominal organs are jumbled up, but the lungs and atrial appendages are usually isomeric, having the same morphologic features on the right and left sides. It is usual for right isomerism to be associated with absence of the spleen and left isomerism with multiple spleens, but these associations are far from constant. Thus different pictures emerge when so-called heterotaxy is subdivided on the basis of isomerism as opposed to splenic morphology. However, cardiac assessment should start with analysis of atrial morphology based on the structure of the atrial appendages.

pulsation. When the atriums are lateralized, almost without exception the inferior caval vein and aorta lie to opposite sides of the spine, with the caval vein on the side of the morphologically right appendage. When there is isomerism, the great vessels usually lie to the same side of the spine, with the caval vein in anterior position in those with isomerism of the right atrial appendages, and posterior, or with the azygos vein posterior, in those having isomerism of the right atrial appendages.

In general, isomerism of the right atrial appendages is associated with absence of the spleen, whereas isomerism of the left atrial appendages is associated with multiple spleens. Patients with isomerism of the atrial appendages therefore are frequently grouped together, from the cardiac standpoint, under the banner of the "splenic syndromes." This approach is much less accurate than describing the syndromes directly in terms of isomerism of the atrial appendages because the correlation between isomerism of the right atrial appendages and absence of the spleen, and between isomerism of the left atrial appendages and multiple spleens, is far from perfect.[20]

Atrioventricular Junctions

In the normal heart, the atrial myocardium is contiguous with the ventricular mass around the orifices of the mitral and tricuspid valves. Electrical insulation is provided at these junctions by the fibrofatty atrioventricular grooves, other than at the site of the penetration of the bundle of His. To analyze accurately the morphology of the atrioventricular junctions in abnormal hearts, it is necessary to know the atrial arrangement. Equally, it is necessary to know the morphology of the ventricular mass to establish which atrium is connected to which ventricle. With this information at hand, it is possible to define the specific patterns of union or nonunion across the junctions and to determine the morphology of the valves guarding the atrioventricular junctions. In hearts with complex malformations, it is also necessary on occasion to describe the precise topology of the ventricular mass and to specify the relationships of the ventricles themselves.

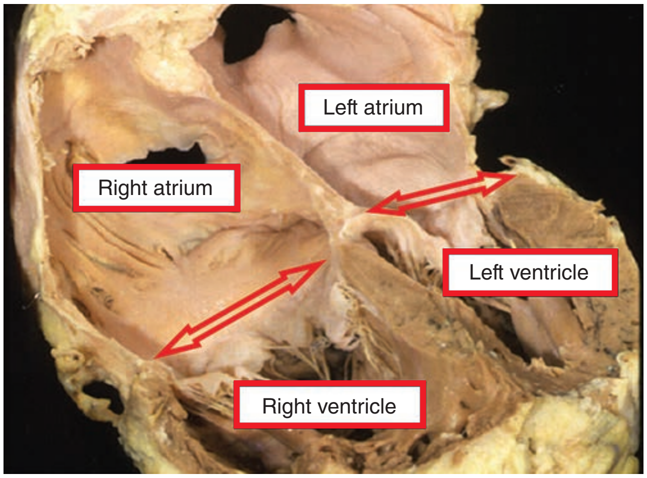

Fig. 1.11 Four-chamber section of the normal heart showing the paired atrioventricular junctions *(arrows)* across which the cavities of the atrial chambers are connected to their appropriate ventricles.

Patterns of Union or Nonunion of the Atrial and Ventricular Chambers

As already described, the patterns depend on the way that the myocardium of both atriums is joined to the ventricular myocardium around the entirety of the atrioventricular junctions, the atrial and ventricular muscle masses being separated from the electrical standpoint by the insulating fibrofatty tissues of the junctions other than at the site of the atrioventricular bundle. The cavities of the atrial chambers therefore are potentially connected to the underlying ventricular cavities via the atrioventricular orifices. In every heart, because there are always two atrial chambers, there is the possibility for two atrioventricular connections, which will be right sided and left sided (Fig. 1.11).

This is the case irrespective of whether the junctions themselves are guarded by two valves (see Fig. 1.11) or a common valve (Fig. 1.12).

One of the junctions may be blocked by an imperforate valvar membrane, but this does not alter the fact that, in such a setting, there are still two potential atrioventricular connections (Fig. 1.13).

In some hearts, this possibility is not fulfilled. This is because one of the connections is completely absent. In this setting the atrial myocardium on that side has no connection with the underlying ventricular myocardium, being separated from the ventricular mass by the fibrofatty tissues of the atrioventricular groove. This arrangement is the most common pattern producing atrioventricular valvar atresia (Fig. 1.14).

When atrioventricular connections are defined in this fashion, all hearts fit into one of three groups. In the first group, by far the most common, the cavity of each atrial chamber is joined actually or potentially, but separately, to that of an underlying ventricle. The feature of the second group is that only one of the ventricles, if indeed two are present, is in communication with the atrial cavities. There is an even rarer third group. This is seen when one atrioventricular connection is absent, and the solitary atrioventricular junction, via a straddling valve, is connected to two ventricles. This arrangement is uniatrial but biventricular.

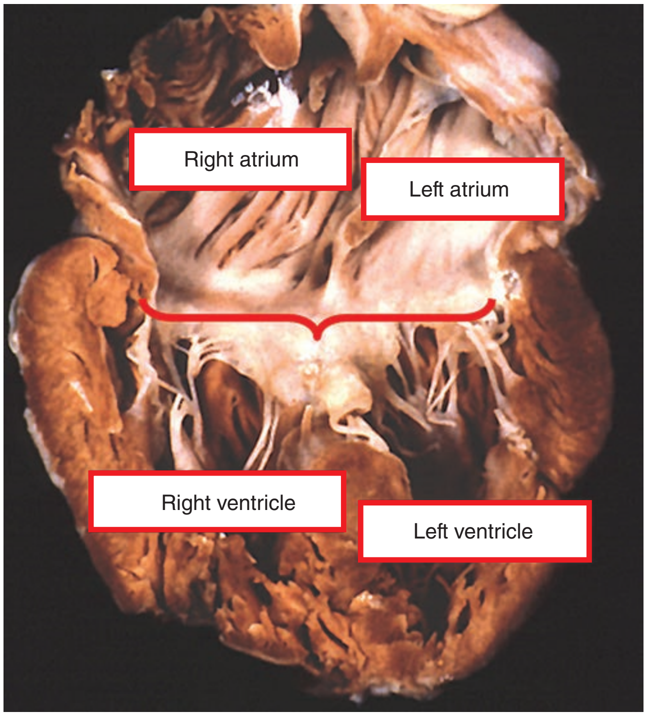

Fig. 1.12 Heart having an atrioventricular septal defect with common atrioventricular junction *(bracket)*. However, the presence of the common junction does not disguise the fact that each atrium is joined to its own ventricle across paired junctions, albeit now guarded by a common valve.

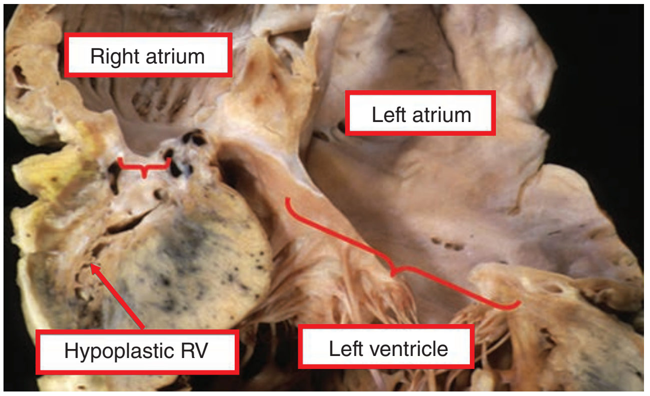

Fig. 1.13 Atrioventricular junctions sectioned in four-chamber fashion in a heart with combined tricuspid and pulmonary atresia. In this instance, unusually, the tricuspid atresia is the consequence of an imperforate right atrioventricular valve. The atrioventricular connections therefore are potentially concordant (compare with Fig. 1.14). *RV*, Right ventricle.

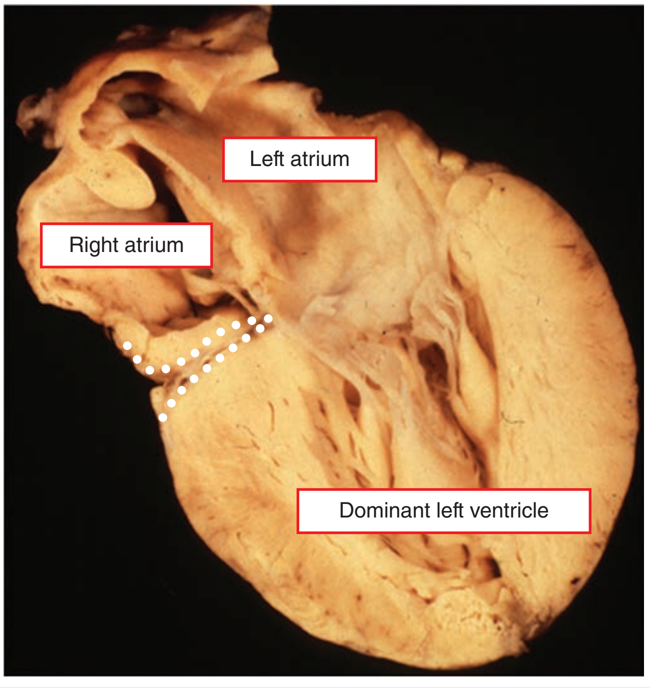

Fig. 1.14 A heart, with the usual form of tricuspid atresia, sectioned in four-chamber fashion. However, only three chambers are seen. This is because the essence of typical tricuspid atresia, and many patients with mitral atresia, is absence of an atrioventricular connection, in this instance the right atrioventricular connection *(dotted line)*.

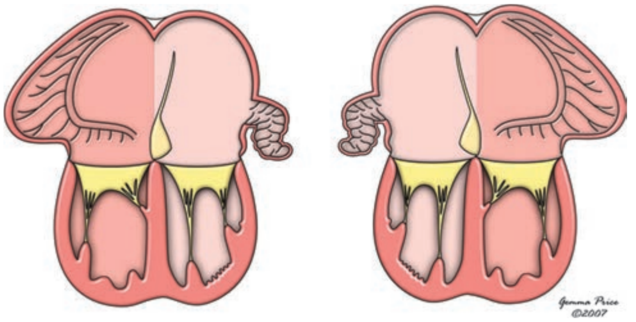

Fig. 1.15 Concordant atrioventricular connections can exist in usual and mirror-imaged patterns. Almost without exception, atriums with usually arranged appendages are joined to a ventricular mass with right hand topology, whereas atriums with mirror-imaged appendages are joined to a ventricular mass with left hand topology. Except when these associations are not present, it is not necessary also to state the topology of the ventricles.

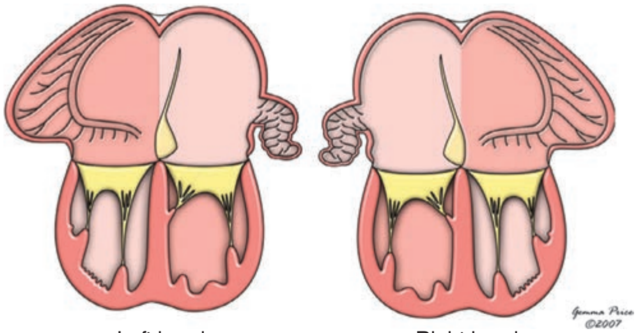

Fig. 1.16 Arrangements that, almost without exception, produce discordant atrioventricular connections.

There are three possible arrangements in those hearts with each atrium joined to its own ventricle; in other words, there are three types of biventricular atrioventricular connection. These depend on the morphology of the chambers connected together. The first pattern is seen when the atriums are joined to morphologically appropriate ventricles, irrespective of the topology or relationship of the ventricles or of the morphology of the valves guarding the junctions. This arrangement produces concordant atrioventricular connections. Such concordant connections can be found with either usually arranged atrial appendages or in the mirror-imaged arrangement (Fig. 1.15).

The second arrangement, which is the reverse of the first, is again independent of relationships or valvar morphology. It produces discordant atrioventricular connections and can again be found in the usual or mirror-imaged situations. When the atrial appendages are mirror imaged in patients with discordant atrioventricular connections, the ventricles are typically in their expected pattern and, in other words, show right hand topology (Fig. 1.16).

These first two arrangements (see Figs. 1.15 and 1.16) are found when the atrial appendages are lateralized. The other biventricular atrioventricular arrangement, in which each atrium is joined to a separate ventricle, is found in hearts with isomeric appendages, whether of right or left morphology. Because of the isomeric nature of the appendages, this third arrangement cannot accurately be described in terms of concordant or discordant connections. It is a discrete biventricular pattern in its own right, which is mixed (Fig. 1.17). It, too, is independent of ventricular relationships and atrioventricular valvar morphologies and requires specification of ventricular topology to make the description complete.

There are also three possible junctional arrangements that produce univentricular atrioventricular connections (Fig. 1.18). The first is when the cavities of right- and left-sided atrial chambers are connected directly to the same ventricle. This is called double-inlet atrioventricular connection, irrespective of whether the right- and left-sided atrioventricular junctions are guarded by two atrioventricular valves or a common valve. The other two arrangements exist when one atrioventricular connection is absent, giving absent right-sided and absent left-sided atrioventricular connection, respectively. The patterns across the junctions that produce univentricular atrioventricular connections are different from those found with biventricular connections. Not only are they independent of ventricular relationships and valvar morphology, but they are also independent of atrial and ventricular morphologies. Hearts with concordant or discordant atrioventricular connections can exist only when usually arranged or mirror-imaged atrial chambers are each joined to separate ventricles. A heart with biventricular mixed connection can only be found when each of two atrial chambers having isomeric appendages is joined to a separate ventricle. In contrast, double-inlet, absent right-sided, or absent left-sided atrioventricular connections can be found with usually arranged, mirror-imaged, or isomeric atrial appendages. Each type of univentricular atrioventricular connection can also be found with the atriums connected to a dominant right ventricle, dominant left ventricle, or morphologically indeterminate ventricle (see Fig. 1.18).

Therefore ventricular morphology must always be described separately in those hearts in which the atrial chambers are joined to only one ventricle. In these hearts, although only one ventricle is joined to the atriums, a second ventricle is present in most of them. This second ventricle, of necessity incomplete, will be of complementary trabecular pattern to the dominant ventricle. Most frequently, the dominant ventricle is a left ventricle. The incomplete ventricle possesses right ventricular apical trabeculations. More rarely, the

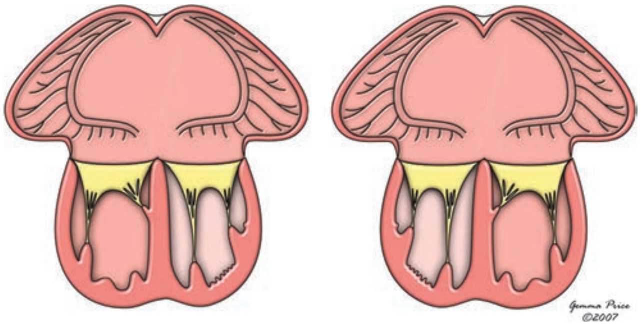

Fig. 1.17 In the setting of isomeric atrial appendages, with right isomerism as shown in the illustration, biventricular connections of necessity are mixed irrespective of ventricular topology. Therefore to fully describe these patterns, it is necessary to specify both the morphology of the atrial appendages and the ventricular topology.

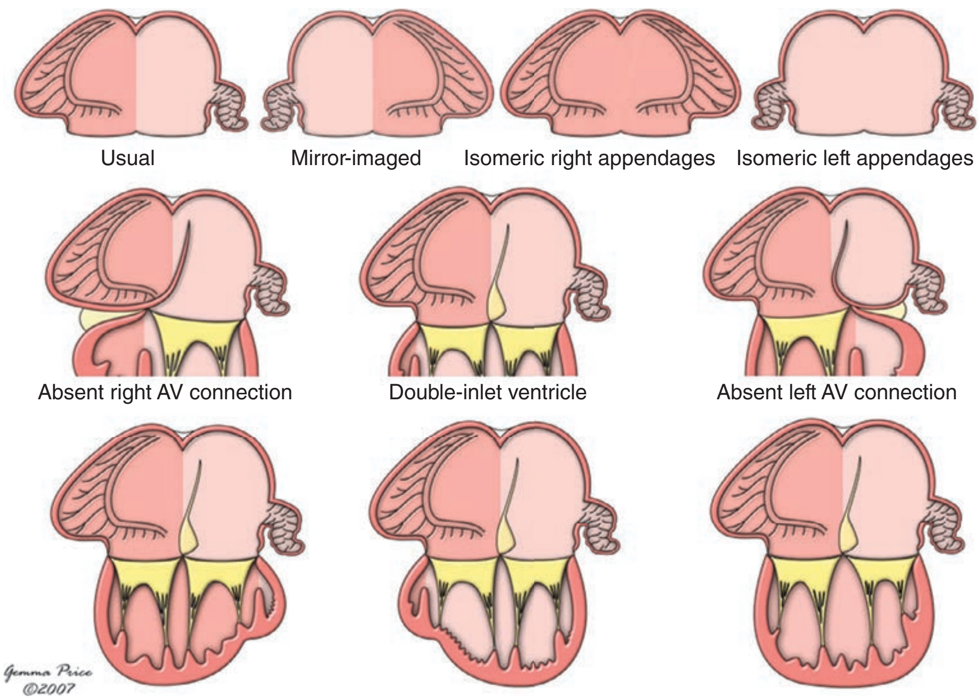

Fig. 1.18 Some of the potential univentricular atrioventricular connections. In reality, these can exist with any arrangement of the atrial appendages *(top)*, with double-inlet, absent right, or absent left atrioventricular *(AV)* connections *(middle)*, and with dominant left ventricle (LV) or right ventricle *(RV)*, or solitary and indeterminate ventricle *(bottom)*. The middle and bottom rows are illustrated with usual arrangement of the atrial appendages simply for convenience. There is further variability with regard to the position of the incomplete ventricle, and with ventriculoarterial connections, and so on. These hearts therefore exemplify the need for full sequential segmental analysis and description.

dominant ventricle is morphologically right, with the incomplete ventricle being morphologically left. Even more rarely, hearts will be found with a solitary ventricular chamber of indeterminate morphology (see Fig. 1.6). In clinical practice, seemingly solitary left or right ventricles may be encountered when the complementary incomplete ventricle is too small to be demonstrated.

Arrangements of the Atrioventricular Valves

Describing the fashion in which the atriums are joined to the ventricles across the atrioventricular junctions accounts only for the way in which the atrial musculature inserts into the base of the ventricular mass. The morphology of the valves guarding the overall atrioventricular junctional area is independent of this feature, within the constraints imposed by the pattern of the junctions itself. When the cavities of both atriums are joined directly to the ventricular mass, the right- and left-sided atrioventricular junctions may be guarded by two patent valves (see Fig. 1.11), by one patent valve and one imperforate valve (see Fig. 1.13), by a common valve (see Fig. 1.12), or by straddling and overriding valves (Fig. 1.19).

These arrangements of the valves can be found with concordant, discordant, biventricular and mixed, or double-inlet types of connection. Either the right- or left-sided valve may be imperforate, producing atresia but in the setting of a potential as opposed to an absent atrioventricular connection. A common valve guards both right- and left-sided atrioventricular junctions, irrespective of its morphology. A valve straddles when its tension apparatus is attached to both sides of a septum within the ventricular mass. It overrides when the atrioventricular junction is connected to ventricles on both sides of a septal structure. A right-sided valve, a left-sided valve, or a common valve can straddle, can override, or can straddle and override. Very rarely, both right- and left-sided valves may straddle and/or override in the same heart.

When one atrioventricular connection is absent, the possible modes of connection are greatly reduced. This is because there is a solitary right- or left-sided atrioventricular connection and hence a solitary atrioventricular valve. The single valve is usually committed in its entirety to one ventricle. More rarely, it may straddle, override, or straddle and override. These latter patterns produce the extremely rare group of uniatrial but biventricular connections (Fig. 1.20).

A valve that overrides has an additional influence on description. This is because the degree of commitment of the overriding atrioventricular junction to the ventricles on either side of the septum determines the precise fashion in which the atriums and ventricles are joined together. Hearts with two valves, in which one valve is overriding, are anatomically intermediate between those with, on the one hand, biventricular and, on the other hand, univentricular atrioventricular connections. There are two ways of describing such hearts. One is to consider the hearts as representing a special type of atrioventricular connection. The alternative is to recognize the intermediate nature of such hearts in a series of anomalies, and to split the series depending on the precise connection of the overriding junction. For the purposes of categorization, only the two ends of the series are labeled, with hearts in the middle being assigned to one or other of the end points. We prefer this second option (see Fig. 1.19). When most of an overriding junction is connected to a ventricle that is also joined to the other atrium, we designate the pattern as being double inlet. If the overriding junction is connected mostly to a ventricle not itself joined to the other atrium, each atrium is categorized as though joined to its own ventricle, giving the possibility of concordant, discordant, or mixed connections.

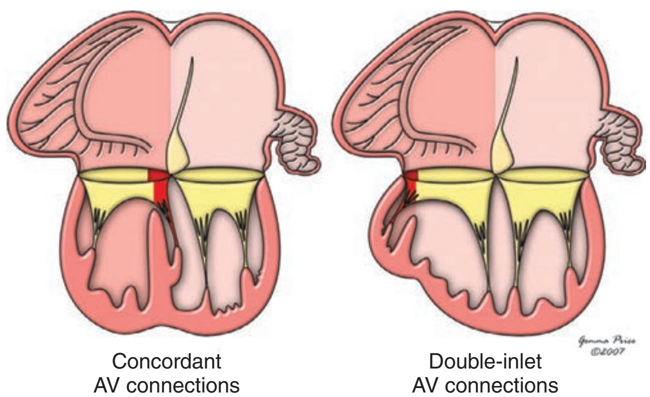

Fig. 1.19 Influence of an overriding atrioventricular (AV) junction on the precise arrangement of the connections. When the lesser part of the overriding junction is attached to the dominant ventricle, the connections are effectively biventricular and concordant in the example shown at *left*. In contrast, when the lesser part is committed to the incomplete ventricle, the connection is effectively double inlet and to the left ventricle in the illustration *(right)*. Any combination of atrial chambers and ventricles can be found with such straddling and overriding valves.

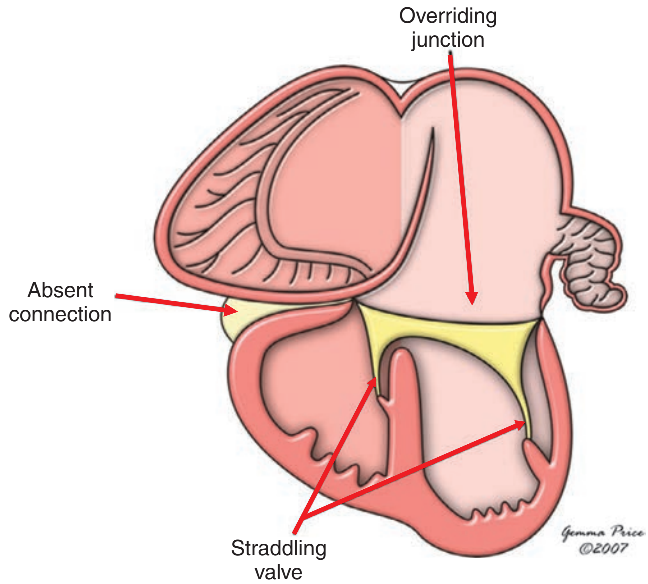

Fig. 1.20 Tricuspid atresia due to absence of the right atrioventricular connection associated with straddling and overriding of the left atrioventricular valve. This produces an atrioventricular connection that is uniatrial but biventricular. The connection can be found with any combination of atrial arrangement and ventricular topology.

When describing atrioventricular valves, it should also be noted that the adjectives “mitral” and “tricuspid” are strictly accurate only in hearts with biventricular atrioventricular connections having separate junctions, each guarded by its own valve. In this context, the tricuspid valve is always found in the morphologically right ventricle and the mitral valve in the morphologically left ventricle. In contrast, in hearts with biventricular atrioventricular connections but with a common junction, it is incorrect to consider the common valve as having mitral and tricuspid components, even when it is divided into right and left components. These right- and left-sided components, particularly on the left side, bear scant resemblance to the normal atrioventricular valves (see Chapter 36). In hearts with double inlet, the two valves are again better considered as right- and left-sided valves rather than as mitral or tricuspid. Similarly, although it is usually possible, when one connection is absent, to deduce the presumed nature of the remaining solitary valve from concepts of morphogenesis, this is not always practical or helpful. The valve can always accurately be described as being right or left sided. Potentially contentious arguments are thus defused when the right- or left-sided valve straddles in the absence of the other atrioventricular connection, giving the uniatrial but biventricular connections.

Ventricular Topology and Relationships

Even in the normal heart, the ventricular spatial relationships are complex. The inlet portions are more or less to the right and left, with the inferior part of the muscular ventricular septum lying in an approximately sagittal plane. The outlet portions are more or less anteroposteriorly related, with the septum between them in an approximately frontal plane. The apical portions extend between these two components, with the muscular septum spiraling between the inlet and outlet components. A shorthand term is needed to describe such complex spatial arrangements, and we use the concept of ventricular topology (see Fig. 1.5). In persons with usually arranged atriums and discordant atrioventricular connections, the ventricular mass almost always shows a left-handed topologic pattern, whereas right-handed ventricular topology is usually found with the combination of mirror-imaged atriums and discordant atrioventricular connections. Although these combinations are almost always present, exceptions can occur. When noting such unexpected ventricular relationships as a feature independent of the topology, we account for right-left, anterior-posterior, and superior-inferior coordinates. When necessary, we describe the position of the three ventricular components separately and relative to each other.

In hearts with disharmonious arrangements in the setting of usual atrial arrangement and discordant atrioventricular connections, the distal parts of the ventricles are usually rotated so that the morphologically right ventricular trabecular and outlet components are to the right of their morphologically left ventricular counterparts, giving the impression of “normal relationships.” In such “criss-cross” hearts seen with usual atrial arrangement and concordant atrioventricular connections, the ventricular rotation gives a spurious impression of left-handed topology. In cases with extreme rotation, the inlet of the morphologically right ventricle may also be right sided in association with discordant atrioventricular connections. Provided relationships are described accurately and separately from the connections and the ventricular topology, none of these unusual and apparently complex hearts will be difficult either to diagnose or to categorize. In addition to these problematic criss-cross hearts, we have already discussed how description of ventricular topology is essential when accounting for the combination of isomeric appendages with biventricular mixed atrioventricular connections. This is because, in this situation, the same terms would appropriately be used to describe the heart in which the left-sided atrium was connected to a morphologically right ventricle, as well as the heart in which the left-sided atrium was connected to a morphologically left ventricle. The arrangements are differentiated simply by describing also the ventricular topology.

Both the position and the relationships of incomplete ventricles need to be described in hearts with univentricular atrioventricular connections. Here the relationships are independent of both the connections and the ventricular morphology. The incomplete right ventricle is usually anterior and right sided in classical tricuspid atresia, but it can be anterior and left sided without in any way altering the clinical presentation and hemodynamic findings. Similarly, in hearts with a double-inlet ventricle, the position of the incomplete ventricle plays only a minor role in determining the clinical presentation. Although a case can be made for interpreting such hearts with univentricular atrioventricular connections on the basis of presumed morphogenesis in the setting of right- or left-handed topologies, there are sufficient exceptions to make this approach unsuitable in the clinical setting. Therefore when we describe the position of incomplete ventricles, we simply account for their location relative to the dominant ventricle, taking note again when necessary of right-left, anterior-posterior, and superior-inferior coordinates. On occasion, it may also be advantageous to describe separately the position of apical and outlet components of an incomplete ventricle.

Ventriculoarterial Junctions

Most polemics concerning the ventriculoarterial junctions devolved upon the failure to distinguish between the way the arterial trunks arose from the ventricular mass as opposed to their relations to each other, along with undue emphasis on the nature of the infundibulums supporting their arterial valves. When these features are described independently, following the precepts of the morphologic method, all potential for disagreement is removed.

ORIGIN OF THE ARTERIAL TRUNKS FROM THE VENTRICULAR MASS

As with analysis of the atrioventricular junctions, it is necessary to account separately for the way the arteries take origin and the nature of the valves guarding the ventriculoarterial junctions. There are four possible types of origin. Concordant ventriculoarterial connections exist when the aorta arises from a morphologically left ventricle, and the pulmonary trunk from a morphologically right ventricle, be the ventricles complete or incomplete. The arrangement where the aorta arises from a morphologically right ventricle or its rudiment, and the pulmonary trunk from a morphologically left ventricle or its rudiment, produces discordant ventriculoarterial connections. Double-outlet connection is found when both arteries are connected to the same ventricle, which may be of right ventricular, left ventricular, or indeterminate ventricular pattern. As with atrioventricular valves, overriding arterial valves (see later) are assigned to the ventricle supporting the greater parts of their circumference. The fourth ventriculoarterial connection is single outlet from the heart. This may take one of four forms. A common trunk exists when both ventricles are connected via a common arterial valve to one trunk that gives rise directly to the coronary arteries, at least one pulmonary artery, and the majority of the systemic circulation. A solitary arterial trunk exists when it is not possible to identify any remnant of an atretic pulmonary trunk within the pericardial cavity. The other forms of single outlet are single pulmonary trunk with aortic atresia or single aortic trunk with pulmonary atresia. These latter two categories describe only those arrangements in which, using clinical techniques, it is not possible to establish the precise connection of the atretic arterial trunk to a ventricular cavity. If its ventricular origin can be established but is found to be imperforate, then the connection is described, along with the presence of an imperforate valve (see later). It is also necessary in hearts with single outlet to describe the ventricular origin of the arterial trunk. This may be exclusively from a right or a left ventricle, but more usually the trunk overrides the septum, taking its origin from both ventricles.

There are fewer morphologies for the valves at the ventriculoarterial than at the atrioventricular junctions. A common arterial valve can exist only with a specific type of single outlet, namely common arterial trunk. Straddling of an arterial valve is impossible because it has no tension apparatus. Thus the possible patterns are two perforate valves, one or both of which may override, or one perforate and one imperforate valve. As with overriding atrioventricular valves, the degree of override of an arterial valve determines the precise origin of the arterial trunk from the ventricular mass, the overriding valve, or valves, being assigned to the ventricle supporting the greater part of its circumference. For example, if more than half of an overriding pulmonary valve was connected to a right ventricle, the aorta being connected to a left ventricle, we would code concordant connections. If more than half the overriding aortic valve was connected to the right ventricle in this situation, we would code double-outlet connections. In this way, we avoid the necessity for intermediate categories. Nonetheless, the precise degree of override is best stated whenever an overriding valve is found. This is done to the best of one's ability, using whichever techniques are available, and recognizing the subjective nature of the task. In this setting, as with atrioventricular connections, we err on the side of the more usually encountered pattern.

ARTERIAL RELATIONSHIPS

Relationships are usually described at valvar level, and many systems for nomenclature have been constructed on this basis. It remains a fact that "d-transposition" is used as though synonymous with all combinations of concordant atrioventricular and discordant ventriculoarterial connections, although this was not how the term was initially used. In the same way, "l-transposition" was used as a synonym for congenitally corrected transposition. In reality, we now know that the relationships of the arterial valves are a poor guide to ventricular topology. Describing arterial valvar position in terms of leftness and rightness also takes no cognizance of anteroposterior relationships, an omission particularly because, for many years, an anterior position of the aorta was used as the cornerstone for definitions of "transposition." We prefer to describe arterial valvar relationships in terms of both right-left and anterior-posterior coordinates. Such description can be accomplished with as great a degree of precision as is required. A good system is the one that describes aortic position in degrees of the arc of a circle constructed around the pulmonary valve.[18] Aortic valvar position is described relative to the pulmonary trunk in terms of eight positions of a compass, using the simple terms left, right, anterior, posterior, and side by side, in their various combinations. As long as we remember that these describe only arterial valvar relationships and convey no information about either the origin of the arterial trunks from the ventricular mass, or the morphology of the ventricular outflow tracts, we have no fear of producing confusion.

From the stance of positions of the arterial trunks, the possibilities are either for the pulmonary trunk to spiral round the aorta as it ascends from the base of the ventricles or for the two trunks to ascend in parallel fashion. Only rarely is it necessary specifically to describe these relationships. Spiraling trunks are associated most frequently with concordant ventriculoarterial connections, and parallel trunks with discordant or double-outlet connections, but again there is no predictive value in these relationships. In almost all hearts, the aortic arch crosses superiorly to the bifurcation of the pulmonary arteries.

An unexpected position of the aortic arch is a well-recognized associated anomaly of conditions such as tetralogy of Fallot (see Chapter 36) or common arterial trunk (see Chapter 41). In this respect, distinction should be made between the position of the arch and the side of the descending aorta, particularly when describing vascular rings (see Chapter 48). The side of the aortic arch depends on whether it passes to the right or left of the trachea. The position of the descending aorta is defined relative to the vertebral column.

INFUNDIBULAR MORPHOLOGY

The infundibular regions are no more and no less than the outlet components of the ventricular mass, but they have proven contentious in the realms of nomenclature. For example, in the past, the presence of bilateral conuses was considered an arbiter of the ventriculoarterial connection when associated with double-outlet right ventricle but ignored when each great artery with its complete muscular infundibulum was supported by its own ventricle. If the infundibular structures are recognized for what they are, and their morphology described as such, they provide no problems in recognition and description.[19] The morphology of the ventricular outlet portions is variable for any heart. Potentially, each ventricle can possess a complete muscular funnel as its outlet portion, and then each arterial valve can be said to have a complete infundibulum. When considered as a whole, the outlet portions of the ventricular mass in the setting of bilateral infundibulums have three discrete parts (Fig. 1.21).

Two of the parts form the anterior and posterior halves of the funnels of myocardium supporting the arterial valves. The anterior, parietal, part is the free anterior ventricular wall. The posterior part is the inner heart curvature, a structure that separates the leaflets of the arterial from those of the atrioventricular valves. We call this component the ventriculoinfundibular fold. The third part is the septum that separates the two subarterial outlets, which we designate the outlet, or infundibular, septum. The dimensions of the outlet septum are independent of the remainder of the infundibular musculature. Indeed, it is possible, albeit rarely, for both arterial valves to be separated from both atrioventricular valves by the ventriculoinfundibular fold but for the arterial valves to be in fibrous continuity with one another because of the absence of the outlet septum. However, in most hearts, some part of the infundibular musculature is effaced, so that fibrous continuity occurs between the leaflets of one of the arterial and the atrioventricular valves. Most frequently, it is the morphologically left ventricular part of the ventriculoinfundibular fold that is attenuated. As a result, there is fibrous continuity between the leaflets of the mitral valve and the arterial valve supported by the left ventricle. Whether the arterial valve is aortic or pulmonary will depend on the ventriculoarterial connections present. In the usual arrangement, the morphologically right ventricular part of the ventriculoinfundibular fold persists, so that there is tricuspid-arterial valvar discontinuity. Depending on the integrity of the outlet septum, there is usually a completely muscular outflow tract, or infundibulum, in the morphologically right ventricle. When both outlet portions are connected to the morphologically right ventricle, most frequently the ventriculoinfundibular fold persists in its entirety, and there is discontinuity bilaterally between the leaflets of the atrioventricular and arterial valves. However, many hearts in which both arterial valves are connected unequivocally to the right ventricle have fibrous continuity between at least one arterial valve and an atrioventricular valve. It makes little sense to deny the origin of both arterial trunks from the right ventricle in this setting. This situation is yet another example of the controversy generated when one feature of cardiac morphology is determined on the basis of a second, unrelated, feature. When both arterial trunks take their origin from the morphologically left ventricle, the tendency is for there to be continuity between the leaflets of both arterial valves and both atrioventricular valves. Even then, in some instances, the ventriculoinfundibular fold may persist in part or in its whole.

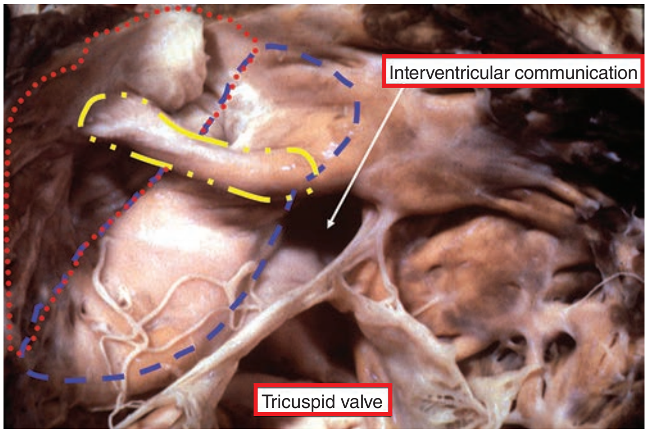

Fig. 1.21 Complete cone of musculature supporting both arterial valves in the setting of double-outlet right ventricle with bilateral infundibulums and subaortic interventricular communication. The cones have parietal parts, outlined in red, posterior parts adjacent to the atrioventricular junctions, outlined in blue, and a part that divides them, outlined in yellow. The part outlined in blue is the ventriculoinfundibular fold, separating the leaflets of the atrioventricular and arterial valves, whereas the dividing part, outlined in yellow, is the outlet septum, interposed between the leaflets of the arterial valves. The anterior part, outlined in red, is the parietal ventricular wall.

It is usually the state of the ventriculoinfundibular fold therefore that is the determining feature of infundibular morphology. Ignoring the rare situation of complete absence of the outlet septum and considering morphology from the standpoint of the arterial valves, there are four possible arrangements. First, there may be a complete subpulmonary infundibulum, with continuity between the leaflets of the aortic and atrioventricular valves. Second, there may be a complete subaortic infundibulum, with continuity between the pulmonary and the atrioventricular valves. Third, there may be bilateral infundibulums, with absence of continuity between the leaflets of the arterial and atrioventricular valves. Fourth, there may be bilaterally deficient infundibulums, with continuity bilaterally between the arterial and the atrioventricular valves. In themselves, these terms are not specific. For specificity, it is also necessary to know which arterial valve takes origin from which ventricle. This emphasizes the fact that infundibular morphology is independent of the ventriculoarterial connections.

Associated Malformations

The majority of patients seen with congenitally malformed hearts will have their cardiac segments joined together in usual fashion, together with normal morphology and relations. In such a setting, the associated malformation will be the anomaly. This textbook is concerned with describing the specific morphologic and clinical features of these anomalies. Nonetheless, it is also necessary to pay attention to the position of the heart within the chest and the orientation of the cardiac apex. It is also necessary to recognize that the heart may be positioned ectopically outside the thoracic cavity. An abnormal position of the heart within the chest is another associated malformation and should not be elevated to a prime diagnosis. This is not to decry the importance of an abnormal cardiac position, if only to aid in interpretation of the electrocardiogram. However, knowing that the heart is malpositioned gives no information concerning its internal architecture. Full sequential segmental analysis is needed to establish the cardiac structure, and not the other way around. The heart can be located mostly in the left hemithorax, mostly in the right hemithorax, or centrally positioned in the mediastinum. The cardiac apex can point to the left, to the right, or to the middle. The orientation of the apex is independent of cardiac position. Both of these are independent of the arrangement of the atrial appendages and of the thoracic and abdominal organs. Describing a right-sided heart, with leftward apex, should be understandable by all, even including the patient.

The complete reference list for this chapter is available at ExpertConsult.com.

Annotated References

Anderson RH, Becker AE, Freedom RM, et al. Sequential segmental analysis of congenital heart disease. *Pediatr Cardiol*. 1984;5:281–288.

Anderson RH, Becker AE, Tynan M, et al. The univentricular atrioventricular connection: getting to the root of a thorny problem. *Am J Cardiol*. 1984b;54:822–882.

In these two reviews, the European school, supported now also by the late Robert Freedom, recognized the wisdom of the "morphologic method." They pointed out that, in so-called hearts with "single ventricles," or "univentricular hearts," it was very rare for the ventricular mass to contain but one chamber. In fact, it was the atrioventricular connections that were univentricular in these settings. Since then, the European school has based its definitions exclusively on the "morphologic method," as explained at length in this chapter.

de la Cruz MV, Nadal-Ginard B. Rules for the diagnosis of visceral situs, truncoconal morphologies and ventricular inversions. *Am Heart J*. 1972;84:19–32.

This review summarized the thoughts of the Latin-American school headed by Maria Victoria de la Cruz, a splendid lady who based her concepts very much on her understanding of cardiac embryology. The system had much in common with the approach taken by Van Praagh and his colleagues and was equally important in guiding further innovations.

Ezon DS, Goldberg JF, Kyle WB. *Atlas of Congenital Heart Disease Nomenclature*. Houston: Ezon Educational Services; 2015.

This atlas provides a "dictionary" to demonstrate the similarities and differences between the Van Praaghian and the Andersonian approaches to description of the congenitally malformed heart. It is profusely illustrated with illustrations prepared by Gemma Price, the artist responsible for the illustrations in this book. The atlas discusses the anatomic features of all the major congenital cardiac lesions.

Jacobs ML, Anderson RH. Nomenclature of the functionally univentricular heart. *Cardiol Young*. 2006;16(Suppl 1):3–8.

This review showed how, by the addition of a simple adverb, namely "functionally," it was possible to defuse all the multiple arguments that continued to surround so-called hearts with "single ventricles." Most such hearts have one big and one small ventricle. The key point is that only the big ventricle is capable of supporting one or other of the circulations, or in most instances both circulations. Hence the arrangement, while not anatomically univentricular, is certainly functionally univentricular.

Shinebourne EA, Macartney FJ, Anderson RH. Sequential chamber localization: the logical approach to diagnosis in congenital heart disease. *Br Heart J*. 1976;38:327–340.

Tynan MJ, Becker AE, Macartney FJ, et al. Nomenclature and classification of congenital heart disease. *Br Heart J*. 1979;41:544–553.

These reviews represented the initial steps taken by the European school of nomenclaturists to refine the segmental approach to diagnosis. The Europeans shifted emphasis from the segments themselves, while still recognizing the importance of segmental morphology, but pointing out at the same time the need to assess the way the components of the segments were joined together, or in some instances not joined together.

Van Praagh R. The segmental approach to diagnosis in congenital heart disease. In: Bergsma D, ed. *Birth Defects Original Article Series*. vol. VIII, No. 5. The National Foundation – March of Dimes. Baltimore: Williams and Wilkins; 1972:4–23.

This chapter in a volume from a series devoted to congenital malformations in general summarized the "state of play" with segmental analysis following the two articles discussed above. The segmental approach, with its shorthand notations, has changed little since this work was published.

Van Praagh R, David I, Wright GB, Van Praagh S. Large RV plus small LV is not single LV. *Circulation*. 1980;61:1057–1058.

This crucial concept, stated in a letter to the Editor, identified a crucial flaw in the approach taken by the European school when analyzing patients with allegedly "single ventricles," or "univentricular hearts." The Bostonians pointed out that it was philosophically unsound to base definitions of a given structure on one of its parts that was variable. Instead, they established the crucial principle of the "morphologic method," stating that the structures be identified on the basis of their most constant components.

Van Praagh R, Ongley PA, Swan HJC. Anatomic types of single or common ventricle in man: morphologic and geometric aspects of sixty necropsied cases. *Am J Cardiol*. 1964;13:367–386.

Van Praagh R, Van Praagh S, Vlad P, Keith JD. Anatomic types of congenital dextrocardia. Diagnostic and embryologic implications. *Am J Cardiol*. 1964;13:510–531.

These two seminal works were the first to suggest that a logical approach be adopted to so-called complex congenital cardiac malformations. Prior to these innovative publications, the complicated malformations had usually been grouped together in a "miscellaneous" category. These important investigations showed that the lesions were amenable to logical analysis.

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