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Classification and Nomenclature of Congenital Heart Defects*

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In the previous edition of this book, we emphasized that one of the first tasks given by God to Adam was to name the animals. Thus began natural science. The population was studied, and the observation of similarities and differences between individuals permitted groups to be identified. These groups formed the basis of a classification and, once this had been established, nomenclature followed. A fundamental purpose of nomenclature, therefore, in addition to providing names, is to recognize similarities and differences, and to give them weight in proportion to their importance. Superficially, a caterpillar resembles a centipede more than it does a butterfly. As far as classification is concerned, however, the fact that caterpillars turn into butterflies, whereas centipedes do not, is much more important. Caterpillars, therefore, are classified with butterflies. In a subject that developed as rapidly as the surgical correction of congenital cardiac malformations, changes of emphasis as to what is or is not important were inevitable. It followed inexorably that nomenclature would also change, albeit that recent years have seen more consensus amongst differing ''schools of thought''. In this respect, what is fundamental to an embryologist may be of very limited importance to a surgeon. Should each choose to ignore the other's point of view, nonetheless, two different nomenclatures will inevitably emerge, and the already existing failure of communication will be exacerbated. Happily, advances over the period since the appearance of the previous edition have seen welcome agreement in the approach to nomenclature. It is now agreed that, when the overall spectrum of congenital cardiac abnormalities is surveyed, the heart is made up of atrial, ventricular and great arterial segments, each of which can vary independently of the others. Thus, it is identification of the chambers and great arterial trunks that is the foundation of nomenclature of congenital cardiac disease. That fact has been accepted for many years. It was at the next step that divergent opinions tended to appear. The essence of the original segmental approach was to take the relations between and within the segments as fundamental (Van Praagh, 1972). When we sought to improve the segmental approach, we took the stance that relationships were relatively unimportant when compared to the way the segments were joined together (Macartney *et al*., 1976; Shinebourne *et al*., 1976; Tynan *et al*., 1979; Anderson and Ho, 1997). Relations describe the way that cardiac structures are arranged in space. These do not necessarily reflect that fashion in which two structures are joined one to the other, this latter feature being described by us as a ''connection''. Abnormalities of relations may modify the surgical approach, but they do not require surgical treatment. Abnormalities of connections, in contrast, need surgical correction, unless the effects of two abnormal connections cancel each other out. For this reason, the approach emphasized in this chapter is in spirit identical to that put forward by Kirklin *et al*. (1973), although Kirklin and his colleagues did not distinguish specifically between connections and relations. It is now noteworthy that Van Praagh has recognized the way the segments join to each other, albeit that he chooses to describe this feature as ''alignments'' (Van Praagh, 2000). We continue to place our emphasis on the union between the cardiac segments rather than their relations, since this approach

* Editors' note: The editors have decided to use Latin anatomical terminology to maintain uniformity throughout this book. This was done with the agreement of Professors Anderson and Macartney. They wish to emphasize, however, their preference for usage of English rather than Latin words.

† It was the Reverend Professor Fergus Macartney who produced this chapter for the initial editions of this book, and who collaborated with RHA on the revision of the chapter for this edition. Sadly, Fergus died before this edition could be printed. The editors of the book join with the surviving author of this chapter in dedicating these pages to his eternal memory.

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is relevant not only to surgeons but also to embryologists, pathologists, cardiologists and radiologists. In short, it is pertinent to all those involved in the nomenclature of cardiac disease. Once the heart is divided into segments and the junctions between the segments are properly analysed, then most problems previously perceived to be ''complex'' turn out to be rather simple. It is probably fair to say that the surgical correction of complex cardiac abnormalities was held up no more by imperfections in surgical technique than by imprecision in preoperative diagnosis. This, in turn, may have resulted from inadequate application of sequential segmental analysis.

IDENTIFICATION OF THE CARDIAC SEGMENTS

Atrial Situs (Atrial Arrangement) and Morphology

Sequential segmental analysis can only work if structures are defined morphologically (Van Praagh, 1972; Anderson and Ho, 1997). This presents a problem only in relation to nomenclature of ventricles and the atrial chambers. Strict clarity demands that the words morphologically ''left'', or ''right'', ''ventricle'' or ''atrium'' should be used whenever these chambers are discussed and described, but this is somewhat cumbersome. For this reason, throughout this book the term ''left ventricle'' is used to indicate the morphologically left ventricle, and so on. In all other circumstances, left and right are used to indicate position, not morphology. It is the appendages of the atrial chambers, and their relationship to the remaining atrial components, that serve as the guide to the basic viscero-atrial situs. The right atrial appendage is broad and blunt, extending from the orifice of the superior vena cava almost to the orifice of the inferior vena cava. It contains parallel trabeculations, the musculi pectinati, which run all the way round the atrio-ventricular junction, and insert at right angles into a rather thicker muscular band, the crista terminalis, inserting in the fashion of the teeth of a comb (**Figure 1.1**). The left atrial appendage, in contrast, is narrow and pointed, often looking like a crooked finger. The musculi pectinati are confined within the appendage, so that the left atrial vestibule is smooth (**Figure 1.1**). The presence of a morphologically left appendage on one side, and a morphologically right appendage on the other, constitutes lateralization. This can be seen in the situs solitus, or situs inversus. In other situations, however, there can be isomeric situs with duplication of morphologically right or morphologically left structures on each side of the body. Because the spleen is the only organ in the body that is left-sided from its inception, right isomerism tends to be associated with asplenia, while left isomerism is usually accompanied by polysplenia, in which the spleens arise on either side of the dorsal mesogastrium (Van Mierop *et al*., 1972). In right isomerism, both lungs tend to have three lobes, and to be fed by eparterial bronchi, whereas in left isomerism, both lungs tend to be bilobed, and to have hyparterial bronchi. The remainder of the organs is haphazardly distributed; an arrangement often termed visceral heterotaxy, albeit that, taken literally, ''heterotaxy'' describes any departure from the normal (Macartney *et al*., 1978). It is the appendages, nonetheless, that provide the best guide to isomerism within the heart. Thus, when musculi pectinati encircle both atrio-ventricular junctions, there is right isomerism, whereas bilaterally smooth vestibules are indicative of left isomerism (Uemura *et al*., 1995b). The method of distinction between lateralized and isomeric situs should not depend on autopsy. The traditional division of complex malformation with jumbled-up bodily organs into the subsets of ''polysplenia'' or ''asplenia'' is unsatisfactory for a number of reasons. First, there is no real consensus on the distinction between polysplenia and one large spleen plus multiple small spleens (Moller *et al*., 1967; Landing *et al*., 1971; Van Mierop *et al*., 1972). Second, the so-called ''asplenia syndrome'' has been described in the presence of a spleen, albeit rudimentary (Layman *et al*., 1967). Most importantly, there are marked discrepancies in classification when features are analysed in the post mortem room on the basis of splenic morphology, as opposed to the morphology of the atrial appendages (Uemura *et al*., 1995a). If such uncertainties exist at autopsy, they are hardly likely to be clarified by investigation in a living patient. In the living patient, it is simplest to infer the atrial situs having visualized the situs of the tracheobronchial tree by plain chest radiography or bronchial tomography (Van Mierop *et al*., 1970; Partridge *et al*., 1975; Macartney *et al*., 1978). A ratio between the lengths of the two main bronchi that is less than 1.6 indicates isomerism, whereas a

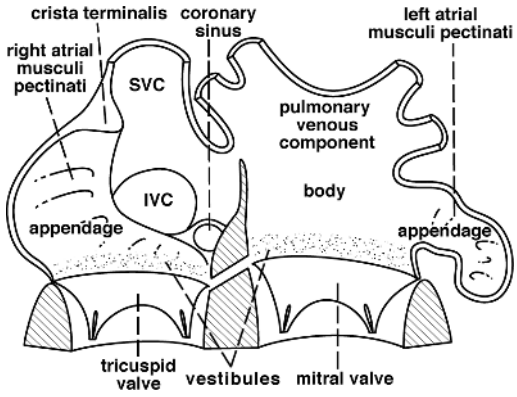

Figure 1.1

ratio higher than 1.6 indicates lateralization, with the morphologically left bronchus being longer. Although discordance between bronchial and atrial situs is also recognized (Caruso and Becker, 1979), the rule of broncho-atrial concordance is accepted as one of the most reliable to be found in paediatric cardiology, given that all rules have exceptions (Macartney *et al*., 1980). Routine cross-sectional echocardiographic scanning of the descending aorta and inferior caval vein also permits accurate inference of atrial arrangement, with the usual situation found when they lie apart with the aorta to the left, mirror imagery when they lie apart but with the aorta to the right, and right isomerism when they run together (Huhta *et al*., 1982). Interruption of the inferior vena cava is suggestive, but not diagnostic, of left isomerism. Because of the lack of complete harmony between the situs of the atrial appendages and the rest of the organs (Uemura *et al*., 1995a), the term ''viscero-atrial situs'' has limited meaning. It is the arrangement of the appendages that is the key to the appropriate description of the atrial segment (Uemura *et al*., 1995b). When in doubt, all systems should be described in independent fashion.

Ventricular Morphology

Because the ventricles develop in series rather than in parallel, there is very limited opportunity for development of ventricular isomerism, although a solitary case has been described with two right ventricles (Rinne *et al*., 2000). The nomenclature of the heart characterized by defective septation, however, has been bedevilled by failure to define precisely what is meant by a ventricle, specifically, how much of a ventricle may be missing before it no longer merits the name ''ventricle'' (Anderson and Ho, 1998). In the past, we argued that chambers lacking an atrial input should be denied ventricular status (Wilkinson *et al*., 1979). We now recognize the futility of this approach (Anderson and Cook, 2004). The key to appropriate analysis is to recognize that normal ventricles have three major components (**Figures 1.2 and 1.3**):

- The inlet component, upstream to the origin of the tensor apparatus.
- The outlet component, or the outflows to the two great arteries.
- The apical trabecular component, found toward the apex of the ventricles.

The septum normally separating the ventricles may be identified by its characteristic tendency to run inferoposteriorly towards the crux of the heart. This external landmark is marked by the intersection of one line formed by the coronary sinus with another formed by the interatrial groove superiorly and the inferior interventricular coronary artery. The septum can also run to the crux, however, when both atrio-ventricular junctions are connected to the morphologically right ventricle (**Figure 1.4**). In some situations, the ventricular septum is malaligned relative to the atrial septum, as in double-inlet left ventricle, and then the ventricular septum runs toward the acute margin of the heart (**Figure 1.4**). A similar situation, with antero-cephalad malalignment of the ventricular septum, is seen in the setting of straddling of the tricuspid valve (Milo *et al*., 1979). In this setting, the tension apparatus of the atrio-ventricular valve is attached to both sides of the

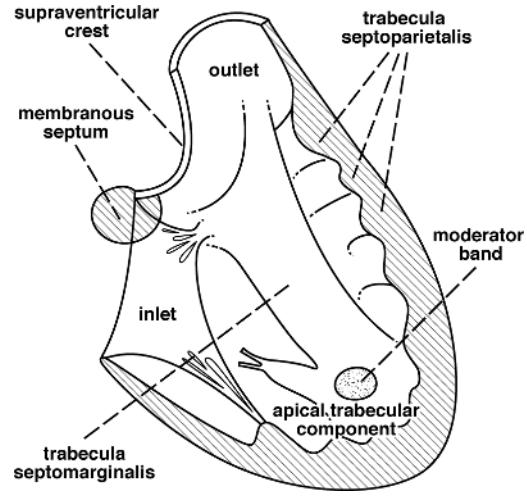

Figure 1.2

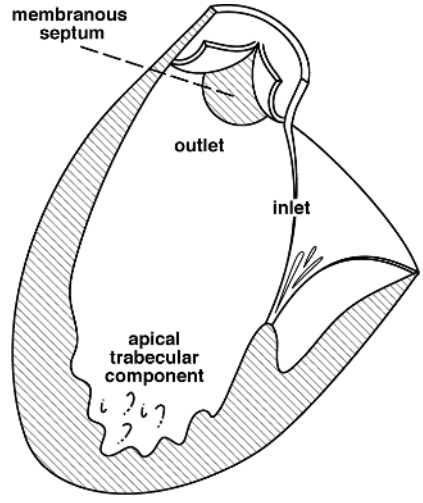

Figure 1.3

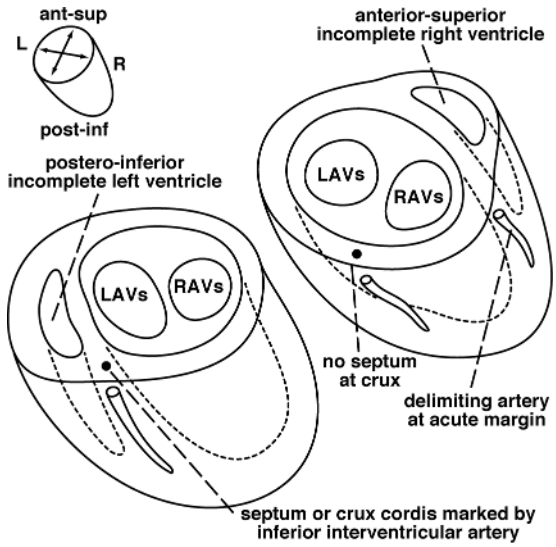

Figure 1.4

septum. The mitral valve can also straddle in this fashion, and straddling is usually associated with overriding of the valvar orifice. On occasion, straddling occurs without overriding, or overriding without straddling (Milo *et al*., 1979). Pathological observations indicate a continuous spectrum of the degree of straddling, from the situation in which one cord merely crosses the septum, to one in which tensor apparatus originates equally from either side. There is a similar spectrum of overriding. In order to determine the atrio-ventricular junctional connections, the valve must be arbitrarily assigned to one ventricle or the other, just as will be done for overriding arterial valves. The valve is assigned to the ventricle that supports the majority of the circumference of the atrio-ventricular junction. Irrespective of these junctional arrangements, however, the presence of valvar overriding has no influence of the apical trabeculations within the ventricles, albeit that the chambers tend to be hypoplastic when they receive only part of an inlet or outlet. In the past, these situations demanded that we change the name of ventricles and rudimentary chambers according to whether or not they possessed an inlet component. Now, we recognize that all chambers within the ventricular mass possessing apical components can be named as ventricles (Anderson and Cook, 2004). They can be of right, left, or indeterminate morphology. Right and left ventricles coexist, with the apical parts separated by the apical septum. This septum carries the conduction system, and is perfused by the septal perforating arteries. Ventricles of indeterminate morphology lack an apical septum, and are the only examples of anatomically single or solitary ventricles.

The apical septum is further characterized by having a morphologically right side that is heavily trabeculated, and a morphologically left side that is finely trabeculated and usually completely smooth in its superior portion. The other important right ventricular characteristic is the presence of a well-developed infundibulum, which interposes between the leaflets of the atrio-ventricular and arterial valves, these leaflets being typically in fibrous continuity in the roof of the left ventricle. This rule, however, has some well-known exceptions. When the atrio-ventricular valves are normally incorporated into the ventricles, a mitral valve, with a solitary zone of apposition between two leaflets, and two papillary muscles not attached to the septum, reliably identifies the left ventricle. A tricuspid valve, with three leaflets, three zones of apposition, and a septal leaflet attached by short cords and multiple discrete papillary muscles to the septum, indicates the right ventricle.

Muscle Bundles Within the Right Ventricle

In the normal heart, the ‘‘supraventricular crest’’ (crista supraventricularis), separates the attachments of the leaflets of the tricuspid and pulmonary valves (**Figure 1.2**). At the point where this crest fuses with the rest of the septum, a small part of muscle can be identified by dissection that interposes between the subaortic and subpulmonary outflow tracts. Without dissection, however, it is not possible to distinguish between these two components. In hearts with anomalies involving the outflow tracts, the two component parts are able to achieve their own identity. Instead of one muscular structure in the ventricular roof, there are two. One, the ventriculo-infundibular fold, separates the attachments of the leaflets of one or both atrio-ventricular valves from the leaflets of one or both arterial valves. The other, the muscular outlet, or infundibular, septum separates the subarterial ventricular outflows to the two great arteries. It is confusing, therefore, to use the term ‘‘crista supraventricularis’’ in an attempt to describe these muscle bundles when the outflow tracts are themselves malformed (Anderson *et al*., 1977). The terms ‘‘parietal band’’ and ‘‘septal band’’, still popular in certain parts of North America, are even more confusing. This is because most surgeons use ‘‘septal band’’ in malformed hearts to describe the septal attachment of the muscular outlet septum. In the normal heart, however, the term is also used to denote an entirely different structure. This is the prominent Y-shaped muscular strap that reinforces the right ventricular septal surface. This strap is best described as the trabecula septomarginalis (Vricella *et al*., 2004). It has two limbs, the postero-inferior, which supports the medial papillary muscle complex or the papillary muscle of the conus, and the

anterosuperior limb, which runs in cephalad direction to support the leaflets of the pulmonary valve. In tetralogy of Fallot, for example, these limbs form a Y-shaped configuration and constitute the inferior rim of the ventricular septal defect. The stem of the Y is the body, which gives off a series of trabeculae septomarginales, one of which, the moderator band, extends to join the anterior papillary muscle of the tricuspid valve (**Figure 1.2**). These considerations are particularly pertinent to tetralogy of Fallot. Thus, in this setting, it is the septal and parietal extensions of the muscular outlet septum that are mobilized to relieve infundibular obstruction, not the ''septal'' and ''parietal'' bands. And the defect is certainly not ''infracristal'', since the components of the normal supraventricular crest have sprung apart as a result of the malalignment between the muscular outlet septum and the rest of the ventricular septum. Because of this septal malalignment, the aortic valve overrides the septal defect, with biventricular connection of the aortic valve (Becker *et al*., 1975).

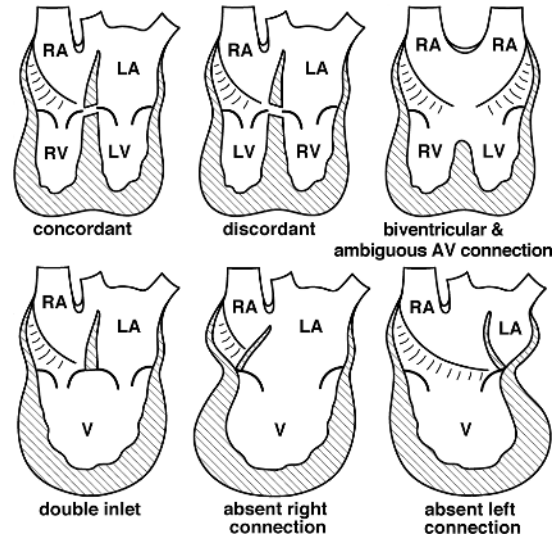

Figure 1.5

Great Arterial Morphology

There is in general little difficulty in distinguishing the two great arteries, in as much as the aorta gives the origin to the coronary and brachiocephalic arteries, while the pulmonary trunk divides into right and left pulmonary arteries. On occasion, one coronary artery or, more rarely, both coronary arteries, can originate from the pulmonary trunk. A brachiocephalic artery may appear to arise from the left or right pulmonary arteries, but in such cases, careful analysis will show that the communication is via a persistently patent ductus arteriosus. No case has thus far been described, to the best of our knowledge, in which there was real difficulty in distinguishing the two great arteries.

JUNCTIONS BETWEEN THE CARDIAC SEGMENTS

Types of Atrio-ventricular Connection

There are several ways in which the atrial chambers can be joined to the underlying ventricular mass (Tynan *et al*., 1979), as illustrated in **Figure 1.5**. We describe the way in which the atrial and ventricular muscles masses are joined together as the ''types of atrio-ventricular connection''. In this respect, it should be noted that the normal heart, and many malformed hearts, possess two atrio-ventricular junctions. Thus, connections are usually plural. Should there be overriding of an atrio-ventricular valve, or the more frequent common atrio-ventricular valve, or rarely both atrio-ventricular valves, the overriding valvar orifice is assigned to the ventricle supporting the greater part of its circumference. When the right atrium drains to the right ventricle, and the left atrium to the left ventricle, the atrio-ventricular connections are said to be *concordant*. When the connections are *discordant*, the right atrium is joined to the left ventricle, and the left atrium to the right ventricle. In the setting of isomeric atrial appendages, then, of necessity, if each atrium is joined to its own ventricle, the connections will be biventricular and *ambiguous*. When both atrial chambers are connected to only one ventricle, there is a *double-inlet* connection. This is the paradigm of the univentricular atrio-ventricular connection, but absence of one atrio-ventricular connection also fulfils the definitions for this final category (Anderson and Cook, 2004). This last condition, therefore, is the only one needing further explanation. It is still frequently not appreciated that the majority of patients with classical tricuspid atresia do not possess an imperforate tricuspid valve, but rather have no tricuspid valve at all, nor any muscular right atrio-ventricular connection (Anderson *et al*., 1979). There is a plane of cleavage containing extracardiac adipose tissue interposed between the right atrium and ventricular mass that can be revealed by blunt dissection in the right atrio-ventricular groove. This plane extends to the central fibrous body, where it achieves continuity with the left atrio-ventricular junction. In this instance, therefore, there is only one atrio-ventricular junction present. The same applies to the left atrio-ventricular groove when the left atrio-ventricular connection is absent. If one atrio-ventricular

connection is *absent*, of necessity there must be a solitary atrio-ventricular valve. In most instances, this solitary valve is exclusively connected to one ventricle, thus producing the second variant of the univentricular atrio-ventricular connection. Infrequently, however, the solitary valve can itself straddle and override. This then produces a uniatrial but biventricular connection, giving one form of so-called ''double-outlet atrium'' (Anderson and Cook, 2004). When describing hearts with absence of one atrio-ventricular connection, it is not always possible to use the adjectives ''tricuspid'' or ''mitral'' with confidence. It is much more accurate, and less confusing, simply to describe the side on which the connection is absent.

Modes of Atrio-ventricular Connection

The *modes* of connection are quite separate from the *types* of connection (Tynan *et al*., 1979). They are illustrated in **Figure 1.6**. The mode of connection is a way of describing the morphology of the atrio-ventricular valves. When both atrial chambers are joined to the ventricular mass, in other words, in the setting of two atrio-ventricular junctions, the mode of connection may be via *two perforate valves*, a *common atrio-ventricular valve*, or *one perforate and one imperforate valve*. Common atrio-ventricular valves normally, but not invariably, *straddle*. The common valve guards both atrio-ventricular junctions. In contrast, the single perforate valve in any of the combinations described earlier, or even the one that is associated with absence of one connection, guards only a solitary atrio-ventricular junction. Such a valve, nonetheless, may also straddle. An imperforate valve is recognized as a translucent membrane blocking the junction between an atrium and its underlying ventricle. Because this can occasionally be confused with the membranous atrio-ventricular septum, the presence of any kind of tension apparatus, be it only a single chorda tendinea inserting into the membrane from its ventricular aspect, assists greatly in recognition of an imperforate valve. Hitherto, the difficulty of distinguishing preoperatively between an absent connection and an imperforate valve has tended to blur the distinction between the two. The ability to make this distinction has now been greatly enhanced by techniques such as cross-sectional echocardiography and other tomographic modalities.

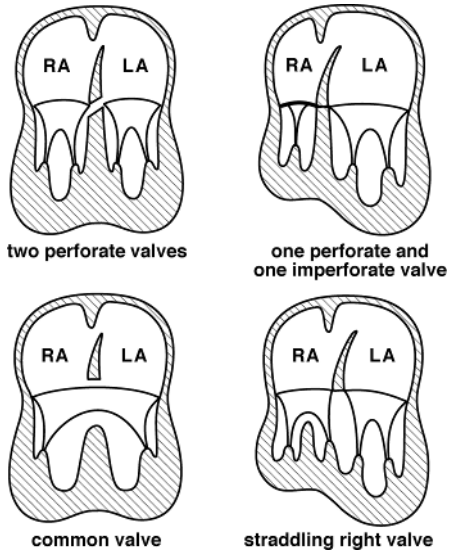

Figure 1.6

Types of Ventriculo-arterial Connection

These are analogous to the types of atrio-ventricular connection, except that there is no biventricular and ambiguous connection. Categorization may also necessitate assignment of an overriding arterial valve to the ventricle, supporting the greater part of its circumference (Kirklin *et al*., 1973; Shinebourne *et al*., 1976; Tynan *et al*., 1979). This assignment is somewhat more arbitrary than for atrio-ventricular valves because arterial overriding is far more common. When overriding approaches 50%, it often helps to assign each leaflet of the arterial valve in turn to a ventricle. The valve is then assigned to supporting the most leaflets or portions thereof. The situation, in which the aorta arises from the left ventricle or its rudiment, and the pulmonary trunk from the right ventricle or its rudiment, is described in terms of concordant ventriculo-arterial connections. With discordant connections, often called ''transposition'', these sites of origin are reversed. Exactly analogous to the double-inlet atrio-ventricular connection is double-outlet ventriculo-arterial connection. When only one great artery is connected to the heart, there is a single outlet from the heart (**Figure 1.7**). This can be via a common arterial trunk, an aorta with pulmonary atresia, a pulmonary trunk with aortic atresia, or via a solitary arterial trunk when there is complete absence of the intrapericardial pulmonary arteries. Should it be possible to trace the atretic trunk to its ventricle of origin, however, it is justifiable to describe the connection accordingly, such as double-outlet right ventricle with pulmonary atresia. It is necessary also to describe a solitary arterial trunk, since it is not possible to determine, had intrapericardial pulmonary arteries been present, whether they would have taken origin from the pulmonary trunk or from the ventricular mass.

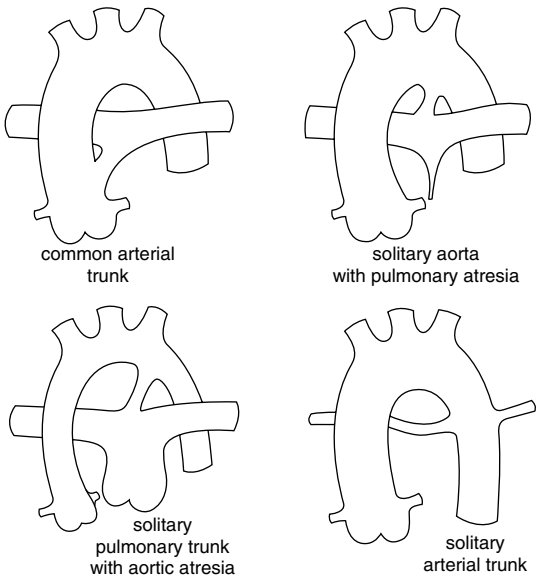

Figure 1.7

Modes of Arterial Connection

The mode of ventriculo-arterial connection includes common, overriding and imperforate valves. Because potential connections of the atrial chambers or great arteries to the ventricular mass by imperforate valves may be difficult to discern in living patients, the absence of the right or left atrio-ventricular connections, and single outlet from the heart, are the designations most likely to be changed by post mortem examination. One of the strengths of this classification, nonetheless, is that even if, for example, a concordant ventriculo-arterial connection with aortic atresia is technically misclassified as single outlet via the pulmonary trunk, a great deal of useful information will have been given.

RELATIONS AND CARDIAC POSITION

These may need to be specified, particularly if they are unpredictable from knowledge of atrial situs and segmental connections. Although an unpredictable relation or position may affect the surgical approach, it does not alter the nature of the operation in the way that abnormal connections do. For example, if the atrio-ventricular connections are discordant, whilst the ventriculo-arterial connections are concordant, it is fundamental to appreciate that the pulmonary and systemic circulations require reversal. Whether the heart is in the right or left side of the chest, whether the ventricles are side by side or one is above the other, and whether the aorta is anterior or posterior to the pulmonary trunk all constitute important pieces of information, but none in itself is fundamental. It is particularly confusing to use a relationship to indicate a connection. Above all, the term ''d-transposition'' should not be used to imply concordant atrio-ventricular connections, neither should the term ''l-transposition'' be used to imply discordant atrio-ventricular connections.

Implications for Reporting Surgical Results

In our opinion, although often held up to ridicule, the so-called ''50% rule'' as applied to determination of atrio-ventricular and ventriculo-arterial connections has solved many more problems than it has created. The rule does have implications, nonetheless, as used currently, for reporting of results of surgery on conditions that form part of a spectrum of anomalies, such as tetralogy of Fallot and double-outlet right ventricle with subaortic ventricular septal defect and pulmonary stenosis. Results for both conditions should best be presented simultaneously. Indeed, it could be argued that the term ''double-outlet'' should be used only to account for the ventriculo-arterial connection, and not to describe a phenotypic entity.

ADDITIONAL CARDIOVASCULAR ANOMALIES

These are most helpfully listed in an order, starting with the great veins and passing through the heart to the great arteries. In the majority of cases, these abnormalities are the only ones present. Consequently, most conditions continue to be described by extremely familiar terms.

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