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Normal and abnormal structure of the ventriculo-arterial junctions

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THE NORMAL HEART POSSESSES TWO ventriculo-arterial junctions – one for the aortic valve, and the other for the pulmonary valve. In the normal heart, these pulmonary and aortic roots are discrete and separate structures, the subpulmonary infundibulum being a free-standing muscular sleeve, separated by extracardiac space from the outflow tract of the left ventricle. When considering the congenital malformations that can distort these outflow tracts, it is conventional to categorise them as existing at subvalvar, valvar, and supravalvar levels. Such an approach, however, ignores the fact the valvar leaflets themselves extend through a significant length of the outflow tracts, with so-called supravalvar stenosis almost always involving the sinutubular junction, this being an integral part of the mechanism that normally ensures competent valvar closure. When considering the lesions that involve abnormally structured outflow tracts, these are best analysed by recognising the different features of the ventriculo-arterial junctions, namely the way the arterial trunks are joined to their underlying ventricles, the interrelationships of the trunks one to the other, and the arrangement of the supporting ventricular structures. In this review, we will show how knowledge of the arrangement of the normal junctions provides the necessary scaffold for logical analysis of all the lesions that can afflict the outflow tracts, be they otherwise normally or abnormally structured.

The normal ventriculo-arterial junctions

One of the ongoing debates in the approach to the analysis of congenitally malformed hearts is whether the cardiac segments are joined[1] or aligned[2] to one another. This, in turn, depends on the particular definition used for the extent of the ventricular mass. In our opinion, the ventricular mass is the musculature structure that extends from the atrioventricular to the ventriculo-arterial junctions. It is the fibro-fatty insulating tissues of the atrioventricular grooves that separate the atrial and ventricular muscle masses, this locus corresponding for the larger part with the attachment of the leaflets of the atrioventricular valves. The situation is subtly different at the ventriculo-arterial junctions. The junctions themselves, when assessed from the stance of their anatomy, are the circular areas where the ventricular musculature supports the walls of the arterial valvar sinuses. This circular locus is best seen in the morphologically right ventricle, where a complete ring of ventricular muscle supports the pulmonary arterial root (Fig. 1). This muscular infundibulum supports the entirety of the pulmonary root, forming a free-standing muscular sleeve (Fig. 2). This can be removed in its entirety from the ventricular base without encroaching on the cavity of the left ventricle (Fig. 3). It is this anatomic arrangement that makes possible the removal of the pulmonary valve for use as an autograft in the Ross procedure.[3] In the morphologically left ventricle, the arrangement is different, since the ventricular muscle supports only part of the aortic root. The remainder of the aortic sinuses is supported by the fibrous continuity that almost always exists between the leaflets of the aortic and mitral valves (Fig. 4).

In both ventricles, nonetheless, because of this interdigitation of arterial valvar sinuses and ventricular support, each arterial root contains three fibrous triangles that extend to the sinutubular junctions, while crescents of ventricular wall, exclusively muscular in the right ventricle, but in part muscular and in part fibrous in the left ventricle, are incorporated at the base of each arterial sinus. The semilunar attachments of the leaflets then permits the valves to close together in competent

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fashion, the zones of apposition between the closed leaflets extending from the centre of the valvar orifice to the sinutubular junction (Fig. 5). Traditionally, it is the peripheral attachments of the leaflets at the sinutubular junction that are known as the "valvar commissures". The entire lengths of the zones of apposition must be competent, nonetheless, if the valve is to close in appropriate fashion. The fibrous triangles that separate the distal attachments of the valvar leaflets as they extend to the sinutubular junction are also of surgical significance, since incisions through these areas take the surgeon outside the heart. The triangle that separates the right and non-coronary leaflets of the aortic valve is more significant, since this triangle is confluent with the membranous septum, and the bundle of His penetrates from atrial to ventricular muscle masses through this important structure (Fig. 6).

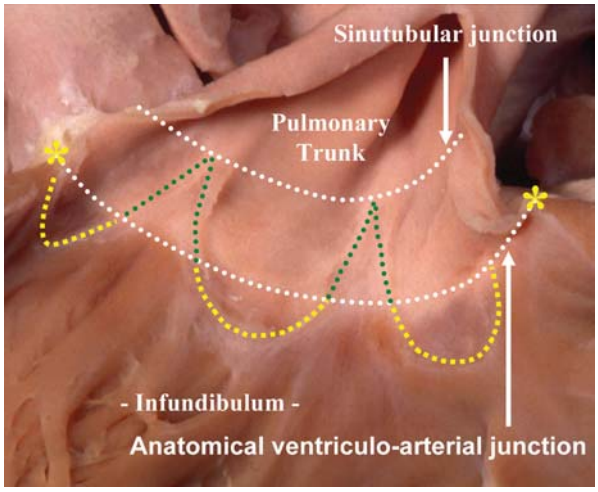


Figure 1.
The normal pulmonary root has been opened and photographed from the front. Note that the arterial walls of the pulmonary trunks are supported throughout their circumference by the muscular pulmonary infundibulum, the two joining across the anatomic ventriculo-anatomic junction, with ends of this junction marked by the yellow asterisks. This anatomic junction is then crossed by the semilunar attachments of the leaflets of the pulmonary valve, with these being attached to the sinutubular junction distally, but to the infundibulum proximally. Because of these semilunar attachments, parts of the leaflets are supported by fibrous triangles extending to the sinutubular junction (green dotted lines drawn for two of the three triangles), while the remaining parts are supported by the muscular infundibulum (yellow dotted crescent drawn for each sinus).

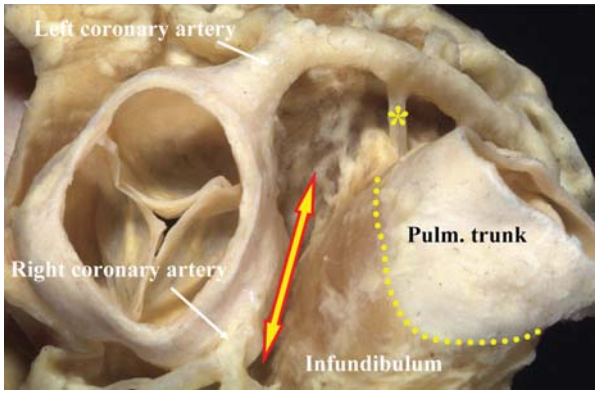


Figure 2.
The base of the ventricular mass has been cleaned, and the pulmonary root pulled forward relative to the aorta. Note the extracardiac tissue plane separating the two roots (double-headed arrow), the sleeve of infundibulum supporting the pulmonary trunk, the circular ventriculo-arterial junction (yellow dotted line), and the site of the first septal perforating artery (asterisk).

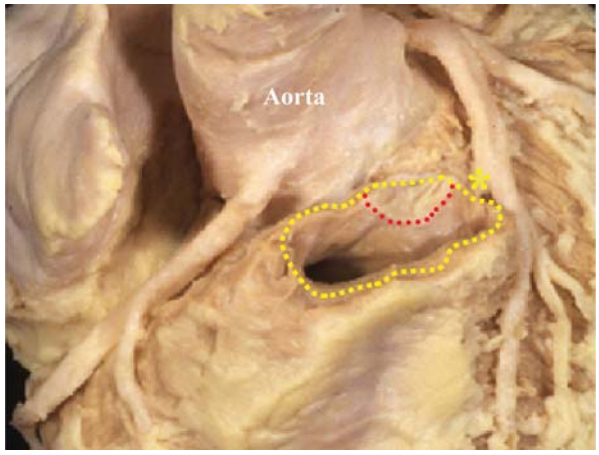


Figure 3.
In this normal heart, the subpulmonary infundibulum has been removed from the ventricular base (yellow dotted line) without encroaching on the cavity of the left ventricle. Note that the basal attachment remains of the right hand facing leaflet of the pulmonary valve (red dotted semicircle), and note also its relationship to the first septal perforating artery (asterisk).

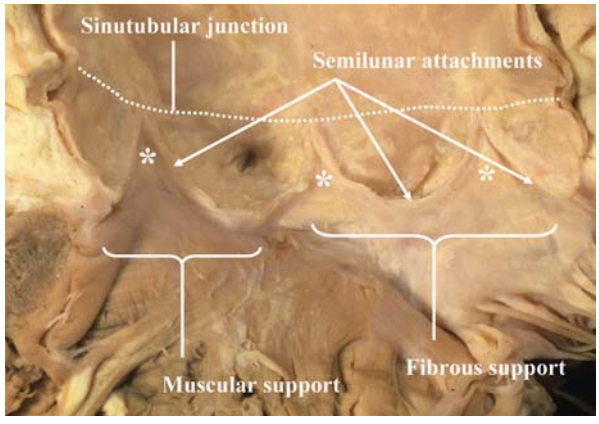


Figure 4.
The normal aortic root has been opened and photographed from the front, showing how the valvar leaflets are attached in semilunar fashion, incorporating triangles of fibrous tissue (asterisks) between the valvar sinuses that extend from the areas of ventricular support to the sinutubular junction. Note that the supporting structures are in part fibrous and in part muscular.

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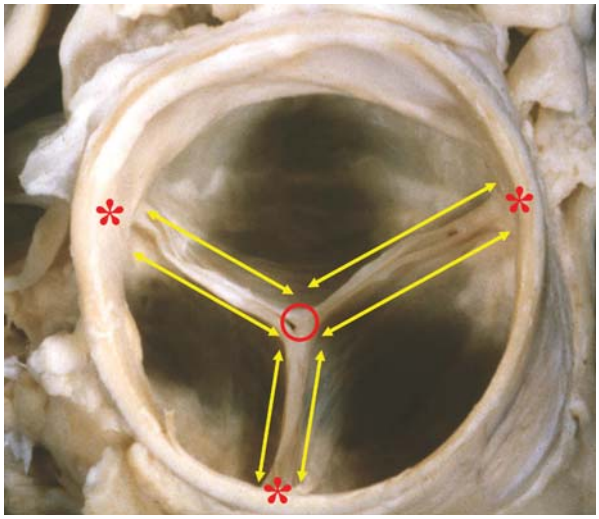


Figure 5.
The normal aortic valve is photographed from above in closed position. Note the three zones of apposition between the leaflets (yellow lines) that extend from the attachments of the leaflets at the sinutubular junction (asterisks) to the centre of the valvar orifice (red circle). Traditionally, it is only the peripheral attachments (asterisks) that are considered to represent the "commissure", but the entirety of the zones of apposition must be competent if the valve is to function properly.

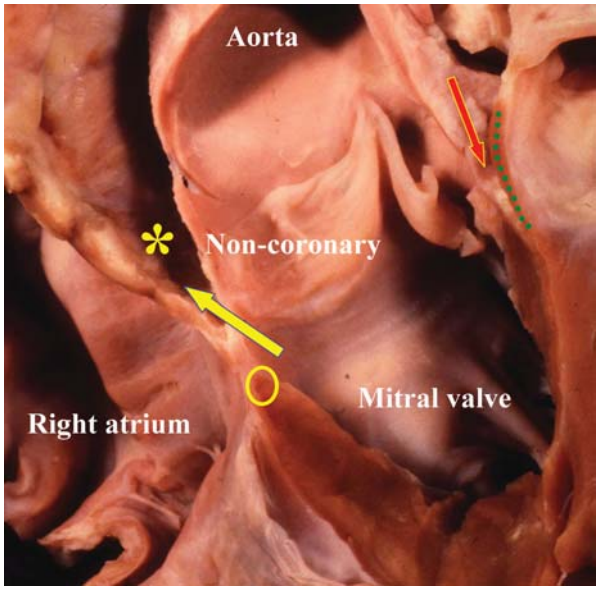


Figure 6.
The normal heart has been dissected from the front to reveal the triangles that separate the right and non-coronary leaflets of the aortic valve (yellow arrow), which points into the transverse sinus of the pericardium (asterisk), and the right and left coronary leaflets, which points into the space (red arrow) between the aortic root and the subpulmonary infundibulum (green dotted line). Note the position of the bundle of His (yellow oval) as it penetrates the atrioventricular component of the membranous septum.

Substrates for obstruction of the ventricular outflow tracts

As we have already intimated, it is usual to categorise obstructions of the ventricular outflow tracts as existing at subvalvar, valvar, and supravalvar levels. Although the "supravalvar" category usually involves also the valve itself, it is convenient to retain this categorisation, the more so since the mechanisms of obstruction are more-or-less comparable within the two ventricles. The differences that are seen in the nature of subvalvar obstruction reflect the different anatomic arrangements of the outflows from the morphologically right as opposed to the left ventricle. Significantly, these differences are maintained irrespective of the nature of the arterial trunk that is supported by the specific ventricular outflow tract. Thus, all the lesions that produce subaortic obstruction in the normally structured heart will produce subpulmonary obstruction when the left ventricular outflow tract gives rise to the pulmonary trunk (Fig. 7). These lesions can involve posterior deviation of the muscular outlet septum, or its fibrous remnant, when the ventricular septum is deficient; fibrous shelves or ridges, protrusion of tissue tags from the membranous septum or adjacent valvar leaflets; hypertrophy of the muscular ventricular septum; or anomalous attachment of the tension apparatus of the mitral valve. In the morphologically right ventricle, in view of the complete muscular infundibulum that supports the leaflets of the arterial valve, irrespective of whether that valve is aortic or pulmonary in identity, it is much commoner for muscular infundibular obstruction to be the substrate for narrowing of the outflow tract. This is then made appreciably worse when the muscular outlet septum is deviated in anterocephalad fashion, as in tetralogy of Fallot (Fig. 8).

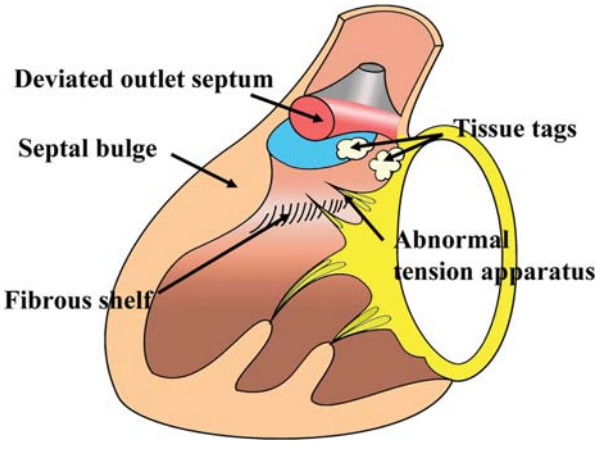


Figure 7.
The cartoon shows the potential substrates for obstruction of the morphologically left ventricular outflow tract, irrespective of whether it supports the aorta or the pulmonary trunk.

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On rare occasions, however, the morphologically right ventricle can also be obstructed by herniation of aneursymal tags, which can take their origin from the membranous septum,[4] can represent enlargement of an aortic sinus of Valsalva, or even the Eustachian valve when this atrial structure takes the form of a windsock.[5]

The essence of obstruction at valvar level is fusion along the zones of apposition between the valvar leaflets. When viewed from the ventricular aspect, this is associated with a loss of the normal semilunar attachments of the valvar leaflets, and failure of formation of one or more of the triangles of fibrous tissue that normally extend to the sinutubular junction.[6] So-called dome-shaped stenosis is seen in the setting of valvar obstruction of the outflow tract of the morphologically right ventricle (Fig. 9), reflecting the fact that the valvar leaflets are uniformly supported by infundibular musculature. The more severe forms of stenosis are associated with greater degrees of fusion along the zones of apposition between the leaflets towards the centre of the valvar orifice.

In the morphologically left ventricle, the typical arrangement producing critical stenosis is so-called unicuspid and unicommisural pattern (Fig. 10), and almost always the solitary persisting zone of apposition in the skirt of leaflet tissue points back towards

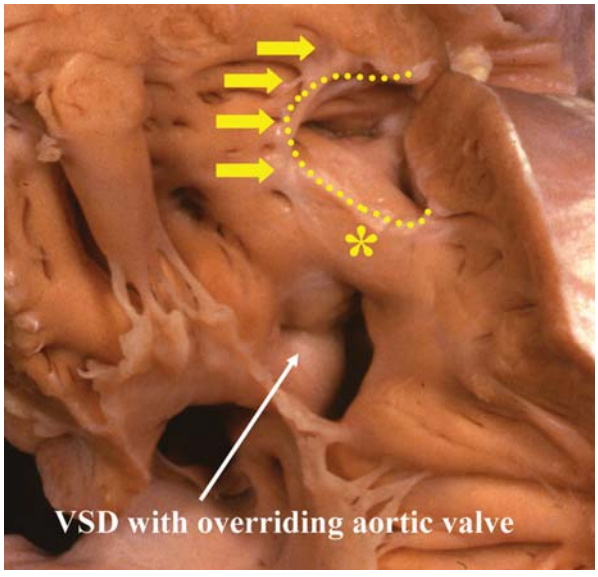


Figure 8.
Obstructions in the right ventricular outflow tract are usually muscular, as in this example of tetralogy of Fallot, with the tightness of obstruction exacerbated in this setting by the antero-cephalad attachment of the muscular outlet septum (asterisk) and fibrous accretions (arrows) around the muscular infundibulum (yellow dots).

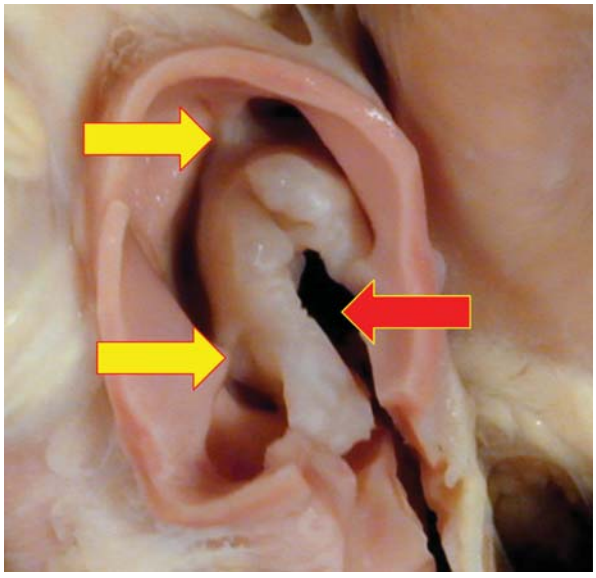


Figure 10.
An example of so-called unicuspid and unicommissural stenosis of the aortic valve. There is but one persisting zone of apposition in the skirt of leaflet tissue (red arrow), pointing back towards the area of fibrous continuity with the mitral valve. Note, however, the formation of two putative zones of apposition, with tethering to the aortic wall extending up to the sinutubular junction (yellow arrows).

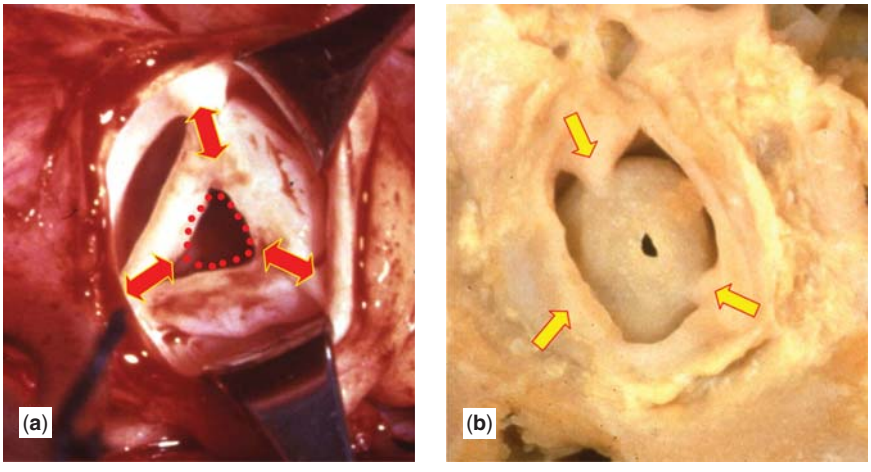


Figure 9.
*These two examples of pulmonary valvar stenosis shows how increasing fusion along the zones of apposition between the leaflets results in greater degrees of valvar stenosis. The arrows show the extent of fusion in (**a**), and the persisting attachments of the leaflets at the sinutubular junction in (**b**), the leaflets themselves having fused to form a dome with a centrally placed critically stenotic orifice.*

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the area of fibrous continuity with the arterial leaflet of the mitral valve. Assessment from the ventricular aspect again reveals that this arrangement is associated with failure of formation of two of the interleaflet triangles,[7] with the skirt of persisting valvar tissue becoming attached within the outflow tract in relatively annular fashion (Fig. 11). It is failure of formation of only one of the interleaflet triangles that produces the congenitally bifoliate valve, be the valve formed in aortic or pulmonary position.

So-called supravalvar stenosis is traditionally divided into the hourglass, diaphragmatic, and tubular forms. The hourglass variant is commonest, irrespective of whether it involves the aorta or the pulmonary trunk. This pattern is no more than stenosis at the level of the sinutubular junction, and involves the valvar leaflets as part of the abnormal arrangement.[8] Even the tubular form of stenosis commences at the sinutubular junction, and again involves the distal attachment of the valvar leaflets. The supravalvar abnormalities, therefore, are best assessed as malformations involving the sinutubular junction, extending variably to involve the tubular parts of the aorta and pulmonary trunk.

Abnormal junctions between the ventricles and the arterial trunks

The essence of the malformations discussed thus far has been that each ventricle supports its own arterial trunk, albeit that the morphological patterns described are unique to the ventricle itself, rather than the arterial trunk supported by the ventricle. In the remaining lesions to be discussed, the most important feature is that it is the junctions between the ventricles and the arterial trunks that are themselves abnormal. This can be because the arterial trunks arise from morphologically inappropriate ventricles, because both trunks arise from the same ventricle, or because there is a common or solitary arterial trunk. There is also an important, albeit rare, variant in which the essence of the abnormality is origin of the arterial trunks from morphologically appropriate ventricles, but in anatomically abnormal fashion. We have already alluded to the ongoing debate as to whether the arterial trunks are aligned with their supporting ventricles,[2] or are connected to them.[1] When considering the ventriculo-arterial junctions as we have described them, there is no question but that the trunks are indeed connected to their supporting ventricles. The supporting ventricular structures are an integral part of the ventricular mass, albeit usually exclusively muscular in the morphologically right ventricle, but fibrous in part in the morphologically left ventricle. As we will see, the specific anatomy of these supporting ventricular structures can vary, albeit without affecting the way that the blood entering the ventricle exits through the arterial trunk, or trunks, supported by the chamber. In similar fashion, the relationships between the arterial trunks themselves can also change without altering the basic haemodynamic picture. The key to logical analysis, therefore, is to begin by determining the fashion in which the arterial trunks are joined to the ventricles, and then subsequently to pay attention to the precise anatomic nature of the supporting ventricular structures, along with the relationships of the arterial trunks one to the other as they exit from the ventricular mass.

We describe these three mutually independent features as the ventriculo-arterial connections, infundibular morphology, and arterial relationships, respectively. It is the variations to be found in each of these features that we will emphasise in the remainder of our review, emphasising that it is the variations in ventriculo-arterial connections that determine the specific haemodynamic patterns.

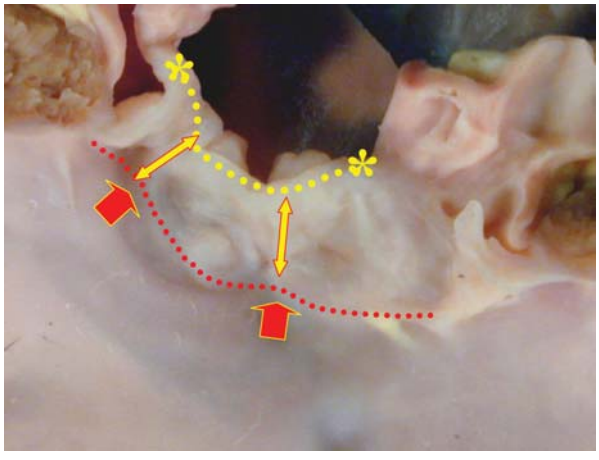


Figure 11.
The left ventricular outflow tract in a heart with critical aortic stenosis has been opened through the ends of the persisting zone of apposition in the skirt of leaflet tissue (asterisks and yellow dotted line). The leaflet tissue itself is attached in more annular fashion (red dotted line), with failure of formation of two interleaflet triangles (red arrows) and fusion along the putative zones of apposition (yellow arrows). Note the fibroelastosis of the left ventricular wall.

Normally structured outflow tracts

The essential feature of the normal heart is that the arterial trunks are connected in concordant fashion to their supporting ventricles. The arterial trunks, nonetheless, can also take origin from morphologically appropriate ventricles when the ventricular anatomy itself, and the relationship of the arterial trunks, are obviously abnormal. This means that, before assuming the presence of normality, it should be checked that, with right-hand topology, the aorta

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arises in right-sided and posterior position from the ventricular base, and that the intrapericardial part of the pulmonary trunk spirals round the aorta as it passes from the superior aspect of the cardiac silhouette before bifurcating into its right and left branches (Fig. 12). It is the presence of the free-standing subpulmonary infundibulum (Fig. 13a) that lifts the pulmonary valve away from the ventricular base. Almost without exception, however, the leaflets of the normally connected aortic valve are in fibrous continuity with one of the leaflets of the mitral valve. This feature stands out for the echocardiographer when assessing the parasternal long axis section (Fig. 13b). On very rare occasions, the aortic valvar leaflets can be separated from the leaflets of the mitral valve in the otherwise normally structured heart,[9] confirming again that it is the way that the arterial trunks arise from the ventricular mass that is the most distinctive anatomic feature, rather than the specific infundibular morphology.

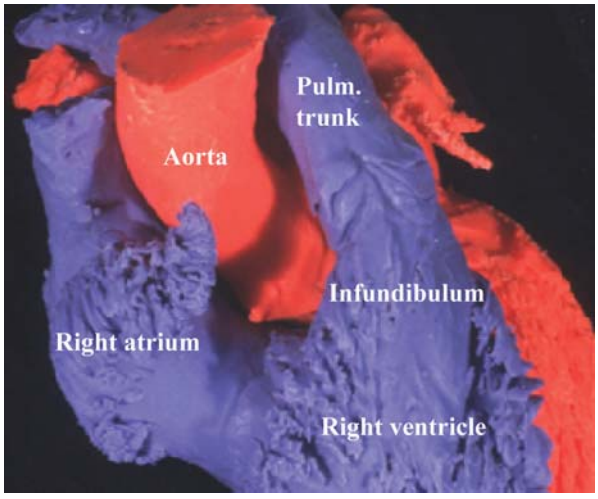


Figure 12.
The cast of the normal heart, photographed in anatomically appropriate orientation, shows the normal spiralling pattern of the intrapericardial parts of the arterial trunks. Note the infundibulum that lifts the pulmonary valve away from the ventricular base.

Discordant ventriculo-arterial connections

The essence of discordant ventriculo-arterial connections is that morphologically inappropriate ventricles support the arterial trunks. In the commonest variant of this arrangement, the atrioventricular connections are concordant, there is fibrous continuity between the leaflets of the pulmonary and mitral valve in the left ventricle, and the leaflets of the aortic valve are lifted away from the base of the morphologically right ventricle by a free-standing muscular infundibulum. In the majority of cases, the aortic valve is positioned anteriorly and to the right of the pulmonary valve (Fig. 14). This arrangement is most usually described simply as "transposition", and this variant is discussed in much greater depth elsewhere in this supplement (see pages 72–75). From the stance of the morphology of the ventriculo-arterial junctions, the clinical picture remains the same when the pulmonary valve arising from the left ventricle is supported by a complete muscular infundibulum (Fig. 15). As long as the ventriculo-arterial connections are discordant, the clinical picture is also undisturbed by different relationships of the arterial trunks. Thus, it is incorrect to describe the combination of concordant atrioventricular and discordant ventriculo-arterial connections as "d-transposition", since a right-sided and anterior aorta is the expected arrangement when congenitally corrected transposition is found in its mirror-imaged variant, while an anterior and

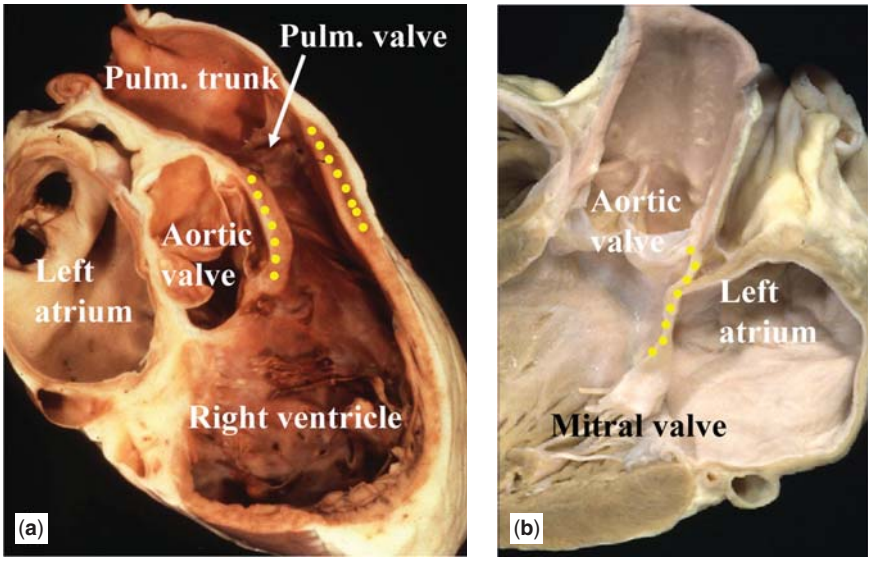


Figure 13.
*The normal heart shown in (**a**) has been sectioned in the oblique subcostal echocardiographic equivalent, revealing the free-standing muscular infundibulum (yellow dots) that lifts the pulmonary valve away from the ventricular base. (**b**), sectioned in parasternal long axis equivalent, shows the fibrous continuity between the leaflets of the aortic and mitral valve, in this particular section through the fibrous triangle separating the non-coronary and left coronary leaflets of the aortic valve (yellow dots).*

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left-sided aorta is the norm when regular transposition is found with mirror-imaged atrial chambers. The combination of usual atrial arrangement and left-sided transposed aorta is also far from rare when the atrioventricular connections are concordant.[10] Fully to describe patients with transposition, therefore, it is necessary to account not only for the discordant ventriculo-arterial connections, but also for abnormal infundibular morphology and unexpected arterial relationships. It is the discordant ventriculo-arterial connections, nonetheless, that are the essence of the abnormal circulatory patterns, this effect being itself modified, of course, by the potential further variations at the atrioventricular junctions.

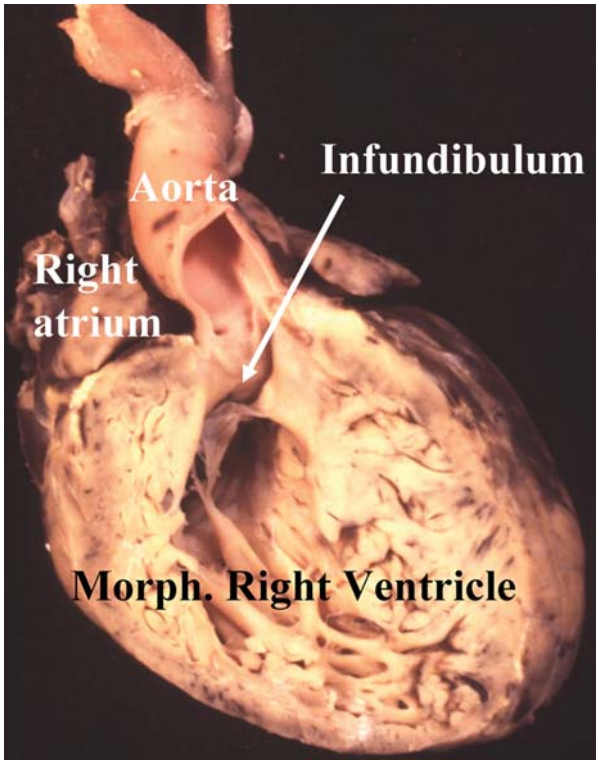


Figure 14.
The essence of transposition is that the atrial chambers are connected to morphologically appropriate ventricles, but the ventricles support morphologically inappropriate arterial trunks. This picture shows the typical arrangement, in which the aorta arises from the right ventricle anterior and to the right of the pulmonary trunk, and with its valve supported by a complete muscular infundibulum.

Double outlet ventriculo-arterial connection

Perhaps the arrangement that still produces most controversy concerning the criterion chosen for diagnosis is that in which, for their greater part, the leaflets of both arterial valves are supported above the cavity of the right ventricle. Some still insist that, so as to make this diagnosis, it is essential that the leaflets of both arterial valves be exclusively supported by muscular infundibular structures.[2] Patients do, of course, have both arterial trunks supported by the musculature of the right ventricle, and the diagnosis is, perhaps, made most easily in such settings, particularly when the interventricular communication is distant from both the subarterial outflow tracts, or in those uncommon situations when the ventricular septum is intact (Fig. 16). It is also the case, nonetheless, that the entirety of the circumference of both arterial valves can be supported by right ventricular structures when there is fibrous continuity between the leaflets of one of the arterial valves and an atrioventricular valve, sometimes the tricuspid valve but more frequently the mitral valve (Fig. 17). To our mind, it makes no sense to deny that, in this setting, the ventriculo-arterial connection is one of double outlet. No-one has suggested that those patients with discordant ventriculo-arterial connections, but with bilateral infundibular structures, as illustrated in Figure 15b, should be described as having double

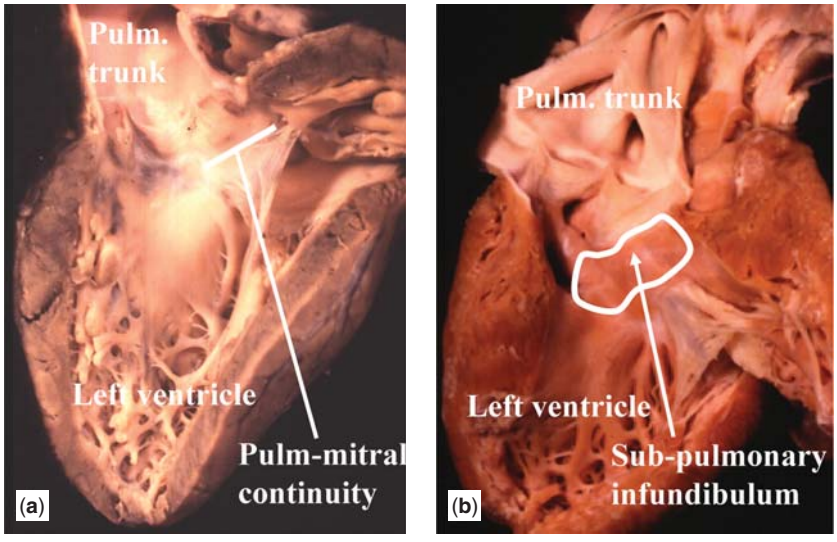


Figure 15.
*In patients with transposition, as shown in (**a**), the leaflets of the pulmonary valve are usually in fibrous continuity with the anterior leaflet of the mitral valve. As shown in (**b**), however, the presence of a subpulmonary infundibulum does not change the basic diagnosis.*

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outlet. Furthermore, defining the existence of double outlet on the basis of another variable feature, namely the state of the infundibular musculature, breaks one of the most important principles established by Van Praagh et al., namely the "morphological method".[11] In reality, "double outlet" is not a diagnosis in its own right, but rather describes one type of ventriculo-arterial connection. The double outlet connection can be found with various atrioventricular connections, with the interventricular communication in various positions relative to the outflow tracts,[12] with different and significant associated malformations, and with various relationships between the arterial trunks.[13] It is probably the location of the interventricular communication that influences to greatest extent the clinical presentation. It is convenient, therefore, to conceptualise the arrangements with double outlet connection as the end points of spectrums involving overriding of one or other, or both, arterial valves.

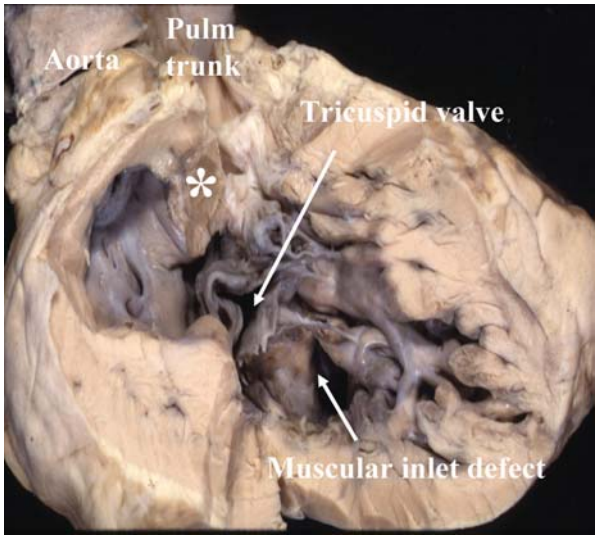


Figure 16.
In this heart, both arterial trunks are supported exclusively above the right ventricle, with the muscular outlet septum (asterisk) separating the outflow tracts. The tricuspid valvar interposes between the outflow tracts and the muscular interventricular communication, which is hence described as being non-committed.

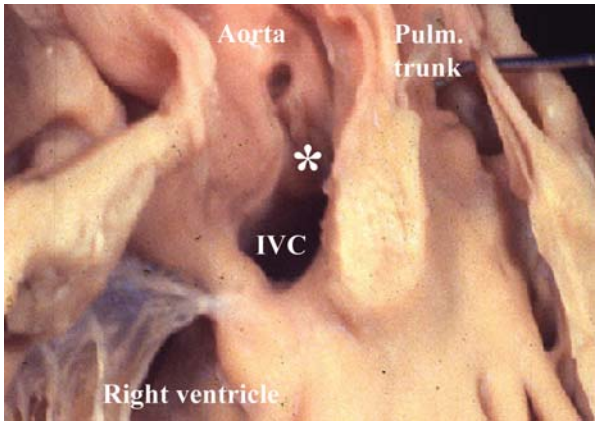


Figure 17.
In this heart, both arterial trunks are again supported by the morphologically right ventricle, as in the example shown in Figure 16, but in this specimen, the interventricular communication (IVC) is immediately below the aortic valve, with fibrous continuity between the leaflets of the aortic and mitral valve in the roof of the defect (asterisk).

Much more rarely, both the arterial valves can be supported over the greater parts of their circumference above the morphologically left ventricle. This arrangement is found most frequently when the leaflets of both arterial valves are in fibrous continuity with the anterior leaflet of the mitral valve, and usually with fibrous continuity also between the leaflets of the aortic and pulmonary valve. With this pattern, both arterial valves tend to override the ventricular septum, a finding that prompted Brandt et al.[14] to name the malformation "double outlet both ventricles". Experience has shown, nonetheless, that as with double outlet from the right ventricle, double outlet left ventricle can exist with different infundibular morphologies, and with various relationships between the arterial trunks.[15]

Single outlet from the heart

There is one other way in which the arterial structures can be attached to the ventricular mass, namely when the aortic and pulmonary pathways are supplied through only one patent arterial trunk. This trunk can be the aorta or the pulmonary trunk, the complimentary trunk then being atretic. Such arrangements are seen in pulmonary atresia with intact ventriclar septum, or classical hypoplasia of the left heart. In these settings, it is almost always possible to trace the atretic trunk to its supporting ventricle, and almost without exception, the lesions are produced in the basic setting of concordant ventriculo-arterial connections. On occasion, nonetheless, the patent aorta in the setting of pulmonary atresia, or the patent pulmonary trunk found with aortic atresia, can take its origin from the morphologically inappropriate ventricle. In such examples of transposition with aortic or pulmonary atresia, the ventricle supporting the atretic trunk then displays the characteristic features of the lesion as seen in the normally connected heart. Thus, in transposition with aortic atresia, it is the right ventricle that is grossly hypoplastic,[16] whereas the left ventricle is the hypoplastic chamber when transposition is associated with pulmonary atresia in the setting of an intact ventricular septum. As with discordant ventriculo-arterial connections in general, the situation can be further complicated by variations in the atrioventricular

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connections. Thus, aortic atresia or pulmonary atresia with intact ventricular septum can also complicate the segmental combinations producing congenitally corrected transposition,[17,18] and must be anticipated to co-exist in the setting of isomeric atrial appendages. Such variation emphasises the fact that differences seen at the ventriculo-arterial junctions are but a small part of the overall make-up of congenitally malformed hearts.

Solitary aorta with pulmonary atresia, or solitary pulmonary trunk with aortic atresia, can also be found in hearts with deficient ventricular septum. Such descriptions should only be used, however, when it is not possible to trace the ventricular origin of the atretic arterial trunk. When the ventricular origin can be identified, then the arrangement is best described as transposition with pulmonary atresia and ventricular septal defect, or double outlet right ventricle with pulmonary atresia and subaortic ventricular septal defect, and so on, according to the specific morphology identified.

There are then two other arrangements with single outlet of the heart that cannot adequately be described in this fashion. The first is when there is a common arterial trunk.[19] As the name suggests, the common arterial trunk supplies directly the pulmonary, aortic, and coronary arterial circulations (Fig. 18). The common trunk usually arises from the ventricular mass astride a directly juxta-arterial interventricular communication. In this setting, there is invariably fibrous continuity between the leaflets of the common truncal and the mitral valves (Fig. 18). In rarer situations, however, the common trunk can arise exclusively from the right ventricle, and then there can be a completely muscular subtruncal infundibulum, albeit that the truncal valve can still retain its continuity with the mitral valve when it arises exclusively from the right ventricle. The common trunk can also arise exclusively from the left ventricle, and then truncal to mitral valvar continuity would be anticipated, but cannot be guaranteed. Since the common trunk is, of necessity, a solitary structure, there is no possibility for variations in arterial relationships to be found at the level of the trunk itself. There is, of course, marked variation in the pattern of origin of the pulmonary arteries. This can be considered to be analogous to the variations in arterial positions as seen with discordant or double outlet ventriculo-arterial connections. Indeed, it was this variation in position of the pulmonary arteries that, for many years, was used to subclassify the hearts unified because they possess a common arterial trunk.[20] It is now accepted, however, that variation in structure of the aortic pathways, specifically the presence of interruption of the aortic arch, or severe aortic coarctation, is of far greater surgical significance.[21] Although some classify the variation as seen in the aortic pathways in alphanumeric fashion,[22] we find this approach unduly procrustean, since it takes no note of the fact that interruption of the aorta can co-exist with various patterns of the pulmonary arteries, nor does it account for such rare variations as when the pulmonary arteries arise from a confluent channel taking its origin from a truncal valvar sinus.

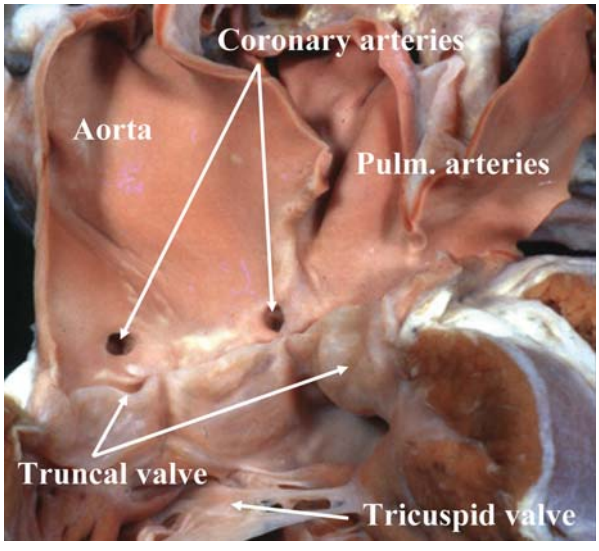


Figure 18.
A common arterial trunk is one that supplies directly the aortic, coronary arterial, and pulmonary circulations.

The final variation to be found when the arterial pathways are fed through a single arterial vessel arising from the ventricular mass is seen when there is complete absence of the intrapericardial pulmonary trunk. In this setting, the pulmonary arteries themselves are fed either exclusively through systemic-to-pulmonary arteries, through an arterial duct or ducts, or by a combination of one duct and systemic-to-pulmonary collateral arteries. In clinical terms, patients with this arrangement behave like, and are treated as, those who have tetralogy of Fallot with pulmonary atresia. Despite the similarities in treatment, it is inaccurate to describe the solitary arterial trunk as seen in this setting as an aorta. This is because, whilst the atretic pulmonary trunk seen in tetralogy of Fallot always takes its origin from the base of the morphologically right ventricle, hearts can also rarely be encountered where an atretic pulmonary trunk can be traced to take origin from a patent arterial trunk exiting from the base of the heart (Fig. 19). Thus, common arterial trunk can rarely co-exist with pulmonary atresia. In the absence of the intrapericardial pulmonary trunk, it is not possible to state with certainty that, had the trunk been present, it would have originated from the base of the right ventricle.

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The only logical description for the arrangement with absence of an intrapericardial arterial trunk, therefore, is to account for the trunk exiting from the base of the heart as being a solitary arterial structure.

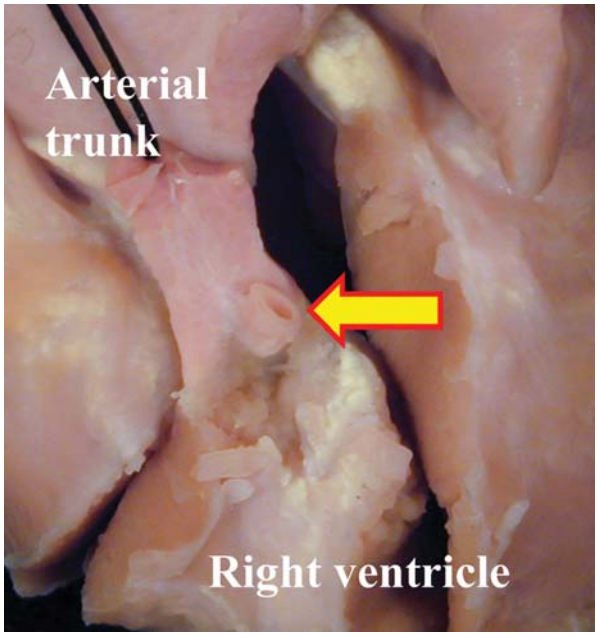


Figure 19.
In this patient, the pulmonary trunk was atretic, but as the dissection shows, the atretic remnant takes its origin from the arterial trunk exiting from the base of the ventricular mass. During development, therefore, the patient initially possessed a common arterial trunk, showing that, very rarely, the common trunk can co-exist with pulmonary atresia. Because of this, it is not justified to assume that, in absence of the intrapericardial pulmonary trunk, had the trunk existed it would always have taken origin from the ventricular mass.

Concordant ventriculo-arterial connections with abnormal outflow tracts

There is then another set of possibilities for abnormal arrangements at the ventriculo-arterial junctions that remains to be described. This group of anomalies are very rare. They have, nonetheless, produced problems for previous taxonomists seeking to provide a system of nomenclature that is both all-embracing and easy to understand. The problems reflect the fact that, as with all the other arrangements described thus far, the fashion in which the arterial trunks are abnormally joined to the ventricular mass is further compromised by the relationships of the arterial trunks one to the other, and by the differing nature of the supporting infundibular structures. The hearts in question are those in which the arterial trunks arise from morphologically appropriate ventricles, but with the arterial trunks leaving the ventricular mass in parallel rather than spiralling fashion, and with the aorta in anterior or side-by-side position rather than posterior to the pulmonary trunk.

In the past, because of the anterior position of the aorta, these hearts were often described as showing "transposition", and for long periods, their existence was questioned on embryological grounds. There is no question, however, but that the hearts do exist (Fig. 20). How, then, should we describe them? By using descriptive rather than nominative terminology! The hearts all have concordant ventriculo-arterial connections, but in the setting of parallel rather than spiralling arterial trunks. When the concordant ventriculo-arterial connections co-exist with concordant atrioventricular connections, it is important to distinguish the rare patients with such segmental

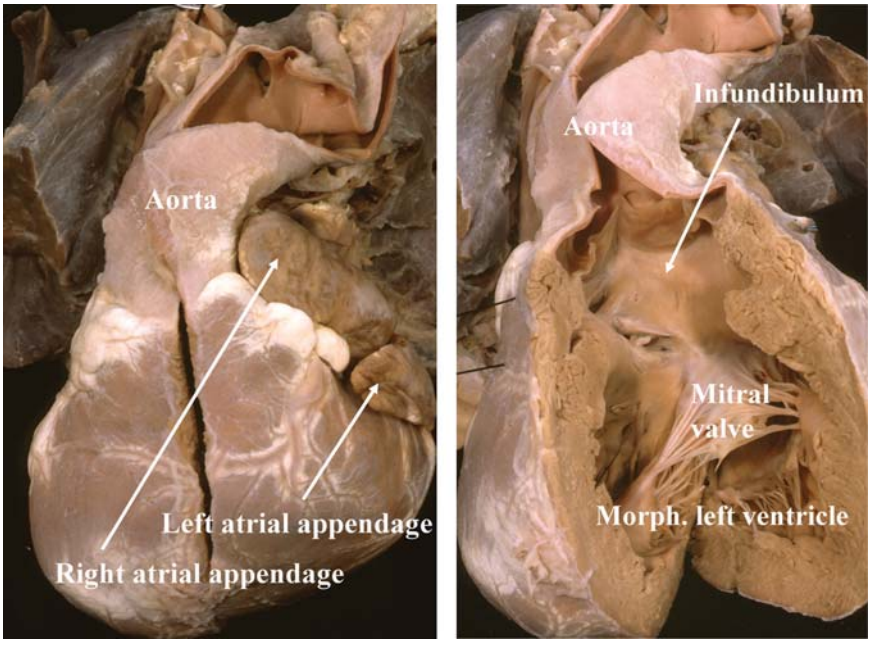


Figure 20.
In this heart, the aorta arise from the morphologically left ventricle, but unlike the usual situation, there is a subaortic muscular infundibulum, and the arterial trunks run parallel to one another as they exit from the ventricular base. The pulmonary trunk arises from the morphologically right ventricle, with its valvar leaflets supported by a complete muscular infundibulum. This particular arrangement of the ventriculo-arterial junctions is known as "anatomically corrected malposition".

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connections from those with congenitally corrected transposition, since both present with potentially normal circulatory patterns, and both most frequently exhibit anterior and left-sided aortas. In the patients with concordant ventriculo-arterial connections, however, it is the morphologically left ventricle that supports the anterior and left-sided aorta, rather than the morphologically inappropriate right ventricle as is the case in those with congenitally corrected transposition. Thus, should there be deficient ventricular septation in these settings, it is entirely appropriate simply to close the ventricular septal defect when the ventriculo-arterial connections are concordant. Nowadays, however, in those having discordant atrioventricular connections, because of worries about the capability of the morphologically right ventricle to support the systemic circulation over a complete lifetime,[23] considerations should be given to proceeding towards a double switch procedure. Note would need to be taken of the abnormal course of the conduction system in this setting.[24]

The potential presence of discordant atrioventricular connections has even greater resonance in the setting of these patients having concordant ventriculo-arterial connections with parallel arterial trunks. This is because this particular arrangement at the ventriculo-arterial junctions, while usually existing with concordant atrioventricular connections (Fig. 21 – left-hand panel), can also be found with discordant ventriculo-arterial connections (Fig. 21 – right-hand panel). The combination of discordant connections at the atrioventricular junctions with concordant ventriculo-arterial connections, of course, produces the clinical picture of transposition. The best surgical option for treatment of such patients is to perform an atrial redirection procedure, such as the Mustard or Senning procedure, since the morphologically left ventricle supports the aorta. Should it also be necessary to close a ventricular septal defect, it must then be remembered that, because of the discordant atrioventricular connections, the atrioventricular node will be abnormally positioned.

In the situations considered thus far, however, we have presumed the presence of complete muscular infundibulums in both ventricles. This is, indeed, the commonest pattern found in the patients with these rare segmental combinations. Even more rarely, however, such patients can be found when the leaflets of the aortic valve, supported by the right-sided morphologically left ventricle, achieve fibrous continuity with the leaflets of the mitral valve (Fig. 22 – left-hand panel). Because of this aortic-to-mitral continuity, some have suggested that the arterial trunks are "normally arranged".[2] These authorities therefore argue that, because the atrial chambers are also normally positioned in this variant, and the ventricular mass unequivocally exhibits left-hand topology (Fig. 22 – left-hand panel), it is appropriate to describe the combinations as "isolated ventricular inversion". And, should the same arrangement be seen with mirror-imaged atrial chambers, then the combination should be dubbed "isolated ventricular non-inversion" (Fig. 22 – right-hand panel).

Such analysis, however, ignores two crucially important facts. First, the "normal" situation for the arterial trunks arising from a ventricular mass exhibiting left-hand topology is as seen in the normally structured heart from a patient with mirror-imaged atrial arrangement, in other words with the aorta positioned to the left and posteriorly, with the pulmonary trunk spiralling round the aorta to the right as it ascends from the ventricular base (Fig. 23). Second, had the

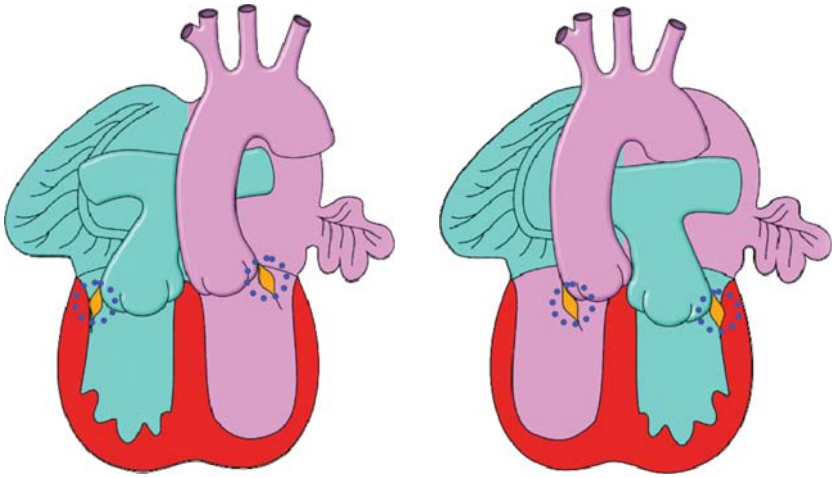


Figure 21.
The cartoons to the left hand show the situation in which arterial trunks arise from the ventricular mass in morphologically appropriate fashion, but in parallel rather than spiralling fashion. Most usually in this setting, there are complete muscular infundibulums supporting the arterial valves in both ventricles (blue dotted circles). The cartoon to the right hand shows exactly the same arrangements at the ventriculo-arterial junctions, but in association with discordant ventriculo-arterial connections. This segmental arrangement produces the haemodynamic situation of transposition, but with concordant ventriculo-arterial connections. Some would call both these arrangements "anatomically corrected malposition".[2] In view of the rarity of such arrangements, our preference is to describe in full the segmental combinations present as shown at the foot of each cartoon.

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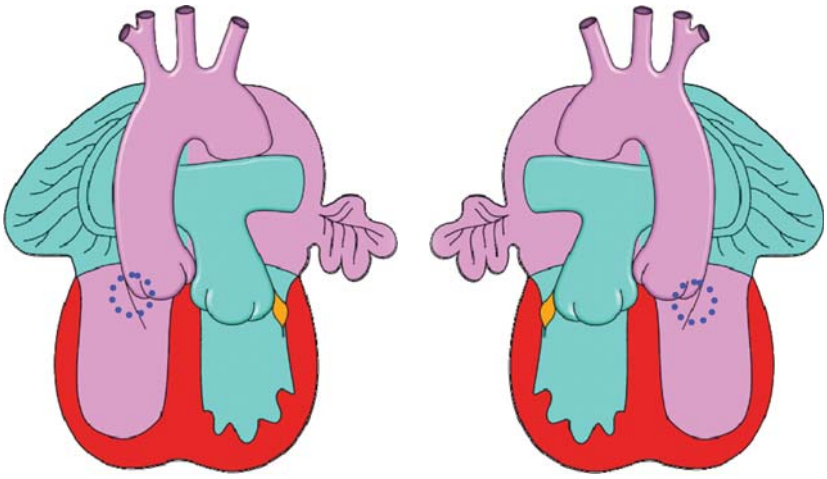


Figure 22.
The cartoon to the left hand shows a variation in the segmental combination of usual atrial arrangement, discordant atrioventricular connections, and concordant ventriculo-arterial connections. In the example shown, there is fibrous continuity between the leaflets of the aortic and mitral valve in the roof of the right-sided morphologically left ventricle (blue dotted circle). The same combination can also exist in the mirror-imaged (MI) format. Some have described the arrangement illustrated to the left hand as "isolated ventricular inversion", while the pattern shown to the right hand is dubbed "isolated ventricular non-inversion".[2] We prefer to describe each feature of the abnormal ventriculo-arterial junctions, as shown at the foot of each illustration.

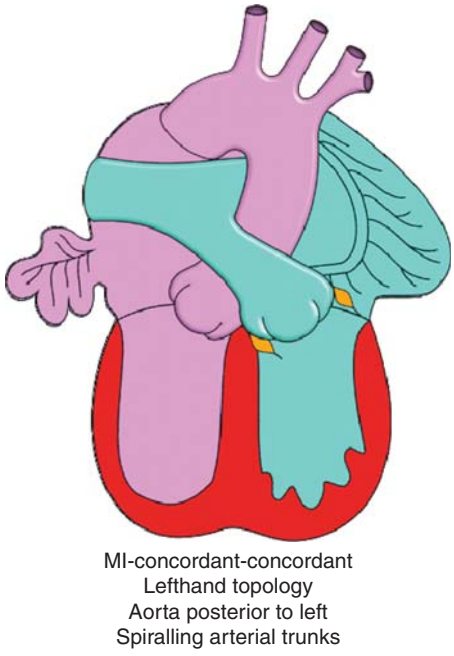


Figure 23.
The cartoon shows the usual situation in which the mirror-imaged ventricular mass, exhibiting left hand topology, gives rise to mirror-imaged but normally connected arterial trunks. The aortic valve is in fibrous continuity with the leaflets of the right-sided mitral valve, but is posterior and to the left of the pulmonary trunk, which spirals round the aorta to the right as it ascends from the ventricular mass. If there truly was "isolated ventricular inversion", then the arrangement would retain the infundibular pattern as shown in this cartoon. Note that, in this drawing, the aortic valve is shown to the right of the pulmonary valve, even though it should be left-sided.

patient with so-called "isolated ventricular inversion or non-inversion" possessed a bar of muscle interposed between the aortic and mitral valves, then at one stroke the description would be changed to "anatomically corrected malposition" (Fig. 24). Can the presence of one or two millimetres of infundibular musculature make so much difference to description, let alone diagnosis and treatment? We think not. For this reason, we prefer to account for these rare patients by analysing and describing all the variable features to be found at the ventriculo-arterial junctions, specifically the connections between the arterial trunks and the ventricles, the nature of the supporting ventricular structures, and the interrelationships of the arterial trunks themselves (Figs 21, 22 and 24).

Conclusions

In this review, we have started with the premise that all congenital malformations involving the ventricular outflow tracts can be described in simple and logical fashion, providing account is taken of the underlying anatomy. For the patient with normal segmental connections, the essential feature is the interdigitations of the arterial valvar sinuses and the supporting ventricular structures, coupled with the differences in ventricular support between the morphologically right and left ventricles, albeit that the arterial valvar leaflets are normally attached in semilunar fashion in both ventricles. For those with abnormal segmental connections, the key to analysis is to appreciate that the fashion in which the arterial trunks are joined to the ventricular mass is mutually independent of the other features of the junctions, these being the interrelationships of the arterial trunks themselves, and the anatomic nature of the ventricular structures supporting the arterial valvar leaflets. As has rightly been emphasised by Van Praagh et al., and dubbed by them the "morphological method",[11] it is philosophically unsound to attempt

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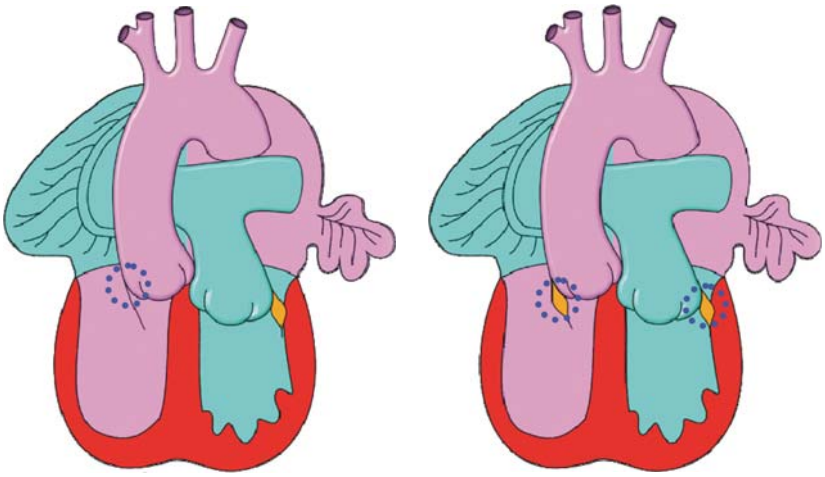


Figure 24.
The cartoons compare the arrangements in which there is usual atrial arrangement, discordant atrioventricular connections, but concordant ventriculo-arterial connections with the aorta arising in right-sided position from the morphologically right ventricle. The cartoon to the right hand shows the variant with both subaortic and subpulmonary infundibulums (red dotted circles), whilst in the example shown to the left hand, there is fibrous continuity between the leaflets of the aortic and mitral valves in the roof of the right-sided morphologically left ventricle (blue dotted circle). It makes little sense to us to argue that, because of this difference in infundibular morphology, the example shown in the right hand panel should be called "anatomically corrected malposition", whilst the arrangement shown to the left hand is called "isolated ventricular inversion". The malformations are encountered so infrequently that we believe it behoves those describing them to account for all features of the abnormal ventriculo-arterial junctions, as shown in the labels at the foot of each cartoon.

to classify one variable feature on the basis of another structure that is itself variable. This principle has become the foundation of our approach to diagnosis and description, since we are at pains to account separately for the variations found in each feature. It is the nature of the connections between the ventricles and the arterial trunks, nonetheless, which retains its primacy, since it is these variations that have the greatest influence on the clinical presentation.

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