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Double outlet right ventricle

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IT SEEMED THAT, OF LATE, THE ENTITY IN WHICH both great arterial trunks arise from the morphologically right ventricle had become less contentious.[1] For many years, arguments had raged with regard to whether the lesion was best defined according to the connections between the arterial trunks and their supporting ventricle,[2] or on the basis of the presence of infundibular musculature supporting exclusively the leaflets of both arterial valves.[3] Much of this controversy had stemmed from the premise that it was possible angiographically to distinguish between tetralogy of Fallot and double outlet right ventricle with pulmonary stenosis,[4] or had been related to the means of distinguishing the variant of double outlet right ventricle with subpulmonary ventricular septal defect, the so-called Taussig-Bing malformation, from similar hearts with discordant ventriculo-arterial connections.[5,6] Some of the problems in making these distinctions had also reflected difficulties in using the so-called "50% rule" in the presence of overriding arterial valves.[7] Advances in cross-sectional echocardiography,[8] coupled with acceptance of the value of the "morphological method", which stated that one morphological variable should not be defined in terms of another anatomic feature which was itself variable,[9] seemed to have resolved most, if not all, of these problems. More recently, however, it has been proposed that it may be better to dispense completely with the designation "double outlet right ventricle".[10] The evidence advanced to substantiate this radical proposal included the suggestion that, in hearts with different ventriculo-arterial connections, the key to diagnosis was the location of the "infundibular septum". As we will show in this review, this is, indeed, the case. But the interpretation of this anatomic evidence as proposed by Sidi and Lecompte[10] does not withstand rigorous analysis. Thus, the structure which they have named the "infundibular septum" itself differs in the anatomic lesions they seek to distinguish. In the setting they have illustrated with discordant ventriculo-arterial connections, their "infundibular septum" is shown as separating the ventricular outflow tracts (Fig. 1a). In contrast, in the lesions they seek to disbar from nomination as double outlet, their "septum" separates the leaflets of the arterial and atrioventricular valves. In fact, their diagram for this variant shows no muscular structure separating the sub-arterial outlets (Fig. 1b). The structure which they nominate as the "infundibular septum" in this situation is, in reality, part of the inner heart curvature. We describe this entity as the ventriculo-infundibular fold, and distinguish it from the infundibular, or outlet, septum.[11] As we will show in this review, therefore, Sidi and Lecompte[10] are correct when they focus attention on the septum which separates the subarterial outflow tracts one from the other. Recognition of the location of this structure, however, does not support the disqualification of double outlet right ventricle as a diagnostic entity. Instead, it is recognition of its location as either a right ventricular or interventricular structure which becomes the key to diagnosis of double outlet in contrast to the hearts having one-to-one ventriculo-arterial connections. It is this difference which we will emphasise in our review. We will also show how the relationship between the muscular outlet septum and the other muscular structures supporting and surrounding the subarterial outlets determines much of the anatomic heterogeneity to be found within the malformation properly described as double outlet right ventricle. We will commence, nonetheless, with a brief review of the evolution of understanding of this contentious lesion.

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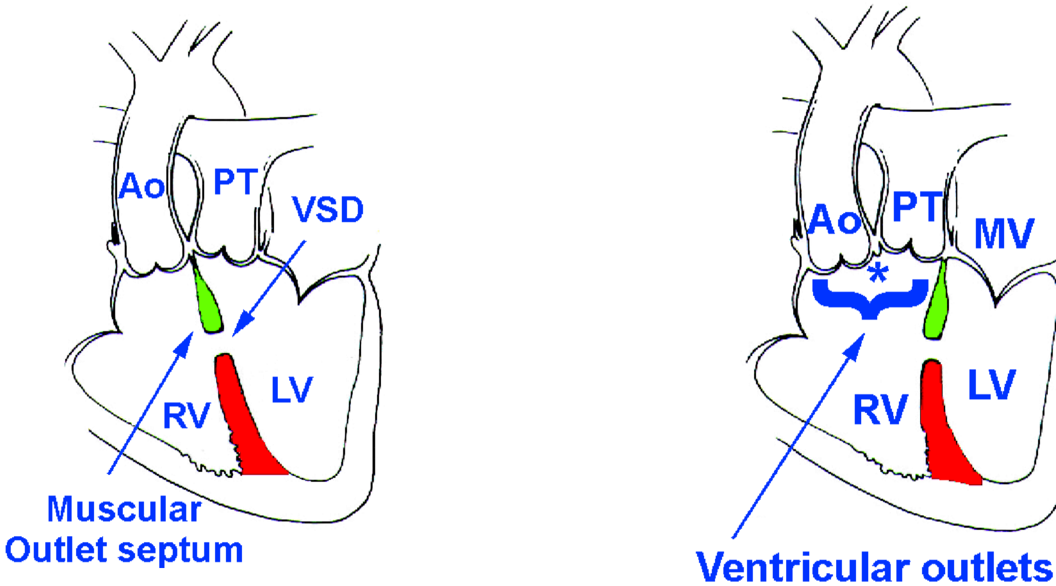

Figure 1.
Sidi and Lecompte[10] argue that the difference between hearts with discordant ventriculo-arterial connections (left hand panel) and the arrangement currently considered to be double outlet right ventricle (right hand panel) reflects the relationship of the arterial trunks to the "infundibular septum". This is certainly true, but from their Figure 26–5, we are not sure of which structure they identify in this fasion for the right hand panel. Thus, the structure we have coloured in green, in the left hand panel, is truly interposed between the sub-arterial outlets, this representing the arrangement with discordant ventriculo-arterial connections. The analogous structure they illustrate in the setting of double outlet, in contrast, is interposed between the leaflets of the pulmonary and mitral valves. In the situation illustrated by Sidi and Lecompte, the true infundibular septum, which would occupy the position shown by the asterisk, is absent. This reflects the arrangement seen in double outlet with doubly committed interventricular communication (see Fig. 15).
Abbreviations Ao – Aorta; PT – Pulmonary trunk; VSD – ventricular septal defect; MV – mitral valve; RV – right ventricle; LV – left ventricle.
Modified from Figure 26–5 from Reference # 10.

Historical considerations

As far as we are aware, it was Mr Abernethy, a surgeon at St Bartholomew's Hospital in London, who first described a heart with double outlet right ventricle.[12] According to him, "both ventricles, the left by means of an opening in the upper part of the septum ventriculorum, projected their blood into the aorta". As we will see, it is the nature and description of this hole in the upper part of the muscular ventricular septum which becomes a key feature in the ongoing debate concerning the existence of double outlet right ventricle. John Farre,[13] writing in 1814, regarded Abernethy's case as being similar in most respects to the entity we would now describe as Fallot's tetralogy, but significantly, Farre focussed attention on the right ventricular origin of the aorta in Abernethy's case. Similar anatomy was also illustrated in the exquisite monograph prepared by von Rokitansky,[14] albeit with neither of the hearts shown being nominated as "double outlet right ventricle" (Fig. 2), or the entity we now call Fallot's tetralogy (Fig. 3). Other earlier investigators, such as Birmingham in 1893,[15] also illustrated the entity, and both Spitzer and Pernkopf included hearts with both arteries arising from the right ventricle within their classification of "transposition".[16] It was Witham, nonetheless, who, to the best of our knowledge, introduced the term "double outlet right ventricle,[17] while Neufeld and his colleagues, in an important series of publications,[18–20] emphasised its clinico-pathological correlations. It was subsequent to these descriptions that the debate evolved concerning the relationship between double outlet and, on the one hand, tetralogy of Fallot and, on the other hand, the Taussig-Bing malformation.[1,2,4–6] This debate centres on one issue. Should double outlet right ventricle be defined on the basis of the connection between the ventricular mass and the arterial trunks, or does the entity exist only when the leaflets of both arterial valves are supported exclusively by infundibular musculature?

There can be no question but that hearts do exist in which both arterial trunks are exclusively supported by the right ventricle, and in which the sub-arterial outflow tracts are exclusively muscular

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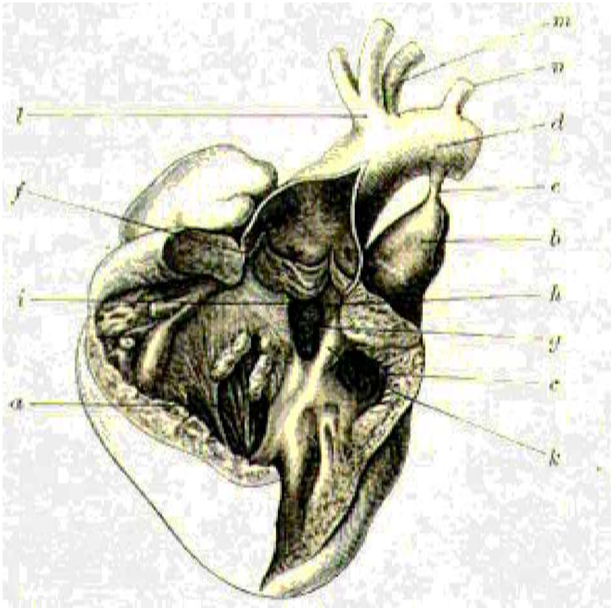

Figure 2.
Reproduction of Figure 6 from the atlas of von Rokitansky[14], showing an example of what we would now call double outlet right ventricle with subaortic interventricular communication. Compare with Figure 19.

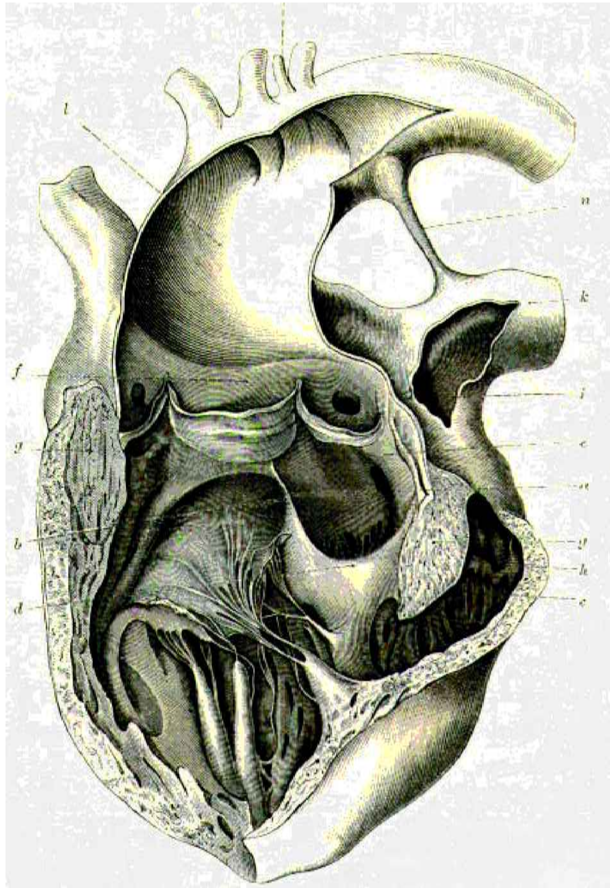

Figure 3.
Figure 7 from the atlas of von Rokitansky[14] is an example of the lesion we now describe as tetralogy of Fallot. Compare with Figure 12.

structures (Fig. 4). By the same token, hearts are also to be found in which both arterial trunks again arise exclusively from the right ventricle, and in which the leaflets of one or both of the arterial valves are in fibrous continuity with the atrioventricular valvar leaflets (Fig. 5). If these latter hearts are not to be described as having double outlet ventriculo-arterial connection, then how is the arrangement of the ventriculo-arterial junctions properly to be described? Furthermore, as has already been emphasised, the presence of bilateral infundibular structures is an inadmissible criterion for definition of ventriculo-arterial connections, or alignments, since use of this feature breaks the important "morphological method" established by Van Praagh and his colleagues[9]. The existence of bilateral infundibulums in hearts with the pulmonary trunk arising from the left ventricle above its own muscular infundibulum would not be used as an argument for nominating the lesion as "double outlet right ventricle". Why, then, should this criterion be used as a definition for double outlet? In fact, infundibular morphology is but one of the variable anatomic features to be found in hearts unified by exhibiting this particular ventriculo-arterial connection.[16] Once it is accepted that diagnosis of double outlet right ventricle depends only on the arrangement of the ventriculo-arterial junctions, it is easy to see why there is so much anatomic heterogeneity to be found in the overall group of hearts with this particular ventriculo-arterial connection. The answer to coping with this heterogeneity is not to dismiss the existence of this particular arrangement, as suggested by Sidi and Lecompte[10], but rather to account for all the variable features.[16]

Overriding arterial valves

Before describing the potentials for anatomic variability, it is worth spending time considering the problems posed by hearts having overriding of one or other of the arterial valves, since it is these entities which pose the major problems in diagnosis.[21] Overriding arterial valves form a subset of lesions between those hearts in which each arterial trunk is committed to its own ventricle, in other words those with one-to-one ventriculo-arterial connections (Fig. 6), and those in which both subarterial outlets arise exclusively from the same ventricle (Fig. 7). The differences between these two groups, however, also impinge on the terms to be discussed subsequently in this review. When

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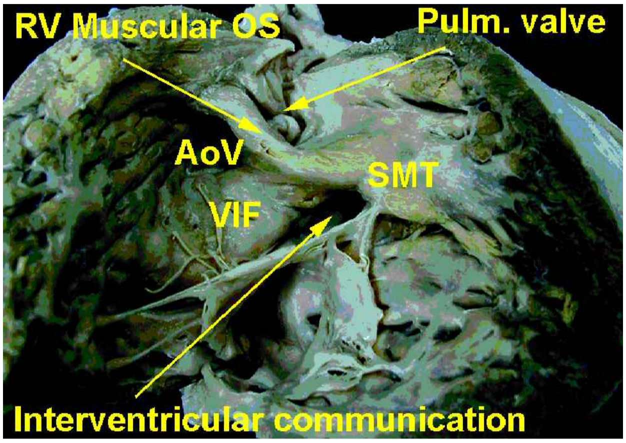

Figure 4.
This heart is a classical example of double outlet right ventricle with bilateral infundibulums. The aortic valve (AoV) is separated from the pulmonary (pulm.) valve by the right ventricular outlet septum (RVOS), which is a muscular structure. The aortic valve is itself supported by a long infundibulum, with the ventriculo-infundibular fold (VIF) interposed between the leaflets of the aortic and tricuspid valves. The interventricular communication is cradled within the limbs of the septomarginal trabeculation (SMT).

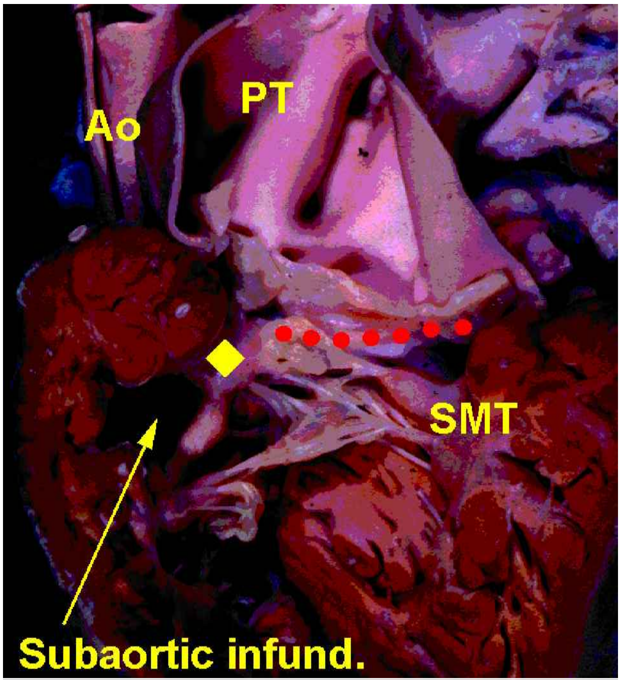

Figure 5.
In this heart, as in the specimen shown in Fig. 4, both arterial valves are supported completely within the right ventricle. The aortic valve again has a compete muscular infundibulum, with the muscular outlet septum (yellow diamond) being a right ventricular structure. The interventricular communication is again between the limbs of the septomarginal trabeculation, but in this heart, the roof of the defect is formed by fibrous continuity between the leaflets of the tricuspid, mitral, and pulmonary valves (red dotted line).

each arterial trunk is committed exclusively to its own ventricle, then the outlet septum, defined as the structure interposed between the sub-arterial outlets, is itself an interventricular structure. In presence of a septal defect between this outlet septum and the remainder of the muscular ventricular septum, the defect itself can also properly be described as the interventricular communication. At the same time, it is the hole closed by the surgeon to restore septal integrity (Fig. 6). The situation is quite different in the setting of double outlet ventricle (Fig. 7). When both arterial trunks arise in their entirety from the right ventricle, the muscular septum which interposes between the sub-arterial outlets is no longer an interventricular structure. Of necessity, it is a right ventricular outlet septum, a feature illustrated clearly in the hearts shown in von Rokitansky's atlas (Figs 2 & 3). The hole between the ventricles, or the interventricular communication, is then bounded superiorly by the inner heart curvature, the structure which we call the ventriculo-infundibular fold, and inferiorly by the crest of the muscular ventricular septum (Fig. 7).

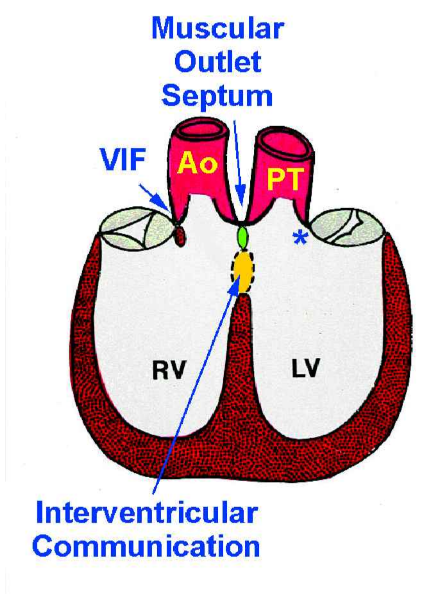

Figure 6.
This diagram shows, as also illustrated in the left hand panel of the cartoon of Sidi and Lecompte (Fig. 1), how the muscular outlet septum (green) is interposed between the sub-arterial outlets when the ventriculo-arterial connections are discordant, and is in line with the apical part of the muscular ventricular septum. The interventricular communication can be closed to restore septal integrity. The inner heart curve, or ventriculo-infundibular fold, is shown interposing between the aortic and tricuspid valves, while fibrous continuity is shown between the pulmonary and mitral valves(*), this being the typical infundibular morphology when the ventriculo-aretrial connections are discordant.

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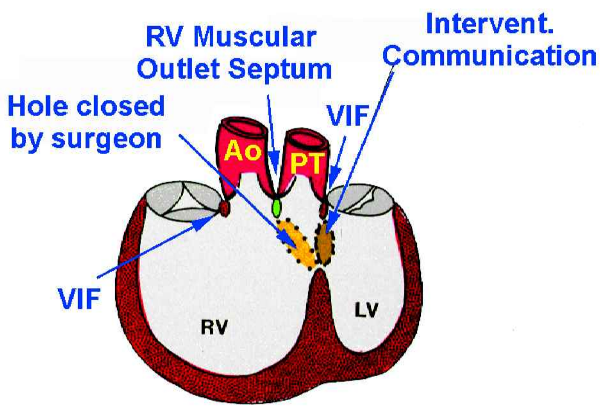

Figure 7.
This diagram contrasts the relationship of the muscular outlet septum (green) relative to the apical ventricular septum when both arterial trunks arise from the right ventricle. The interventricular communication is now the outflow tract from the left ventricle, being bounded by the crest of the apical ventricular septum and the ventriculo-infundibular fold between the leaflets of the mitral and pulmonary valves. The muscular outlet septum (green) is an exclusively right ventricular structure. In this situation, the surgeon would need to create a tunnel from this outlet septum to the crest of the ventricular septum so as to re-connect the pulmonary trunk with the left ventricle, combining this maneuver most usually with an arterial switch procedure.

With this arrangement of the ventriculo-arterial junctions, the surgeon can no longer close this hole to restore septal integrity without walling off one arterial trunk from the left ventricle. Instead, so as to create the haemodynamic situation in which each ventricle can eject to its own arterial trunk, the interventricular communication must be tunnelled to one or other sub-arterial outlet (Fig. 7).

In hearts which have overriding arterial valves, the situation is intermediate between these two arrangements. A cone of space exists between the circumference of the overriding arterial trunk and the crest of the muscular ventricular septum (Fig. 8). According to the precise connections of the leaflets of the overriding arterial valve, the plane of the interventricular communication is somewhere within this cone. The cone itself, however, also has right and left ventricular margins (Fig. 9). Should the degree of override be such that the left ventricular opening to this cone of space is judged to be the interventricular communication, then it follows inexorably that the ventriculo-arterial connection has become double outlet. Thus, in typical tetralogy of Fallot with concordant ventriculo-arterial connections, it is the right ventricular margin of the sub-arterial cone which is usually nominated as the ventricular septal defect (Fig. 10). Hearts can also be found, nonetheless, in which the morphology of the outflow tracts is typical for tetralogy, but the aorta arises exclusively from the right ventricle, either with bilateral infundibular structures (Fig. 11), or with fibrous continuity between the leaflets of the aortic and mitral valves (Fig. 12). In these hearts, as with the situation illustrated in Fig. 7, it is the interventricular communication which is usually considered to represent the "ventricular septal defect". Yet, in these settings, the surgeon would close the plane of space limited superiorly by the right ventricular

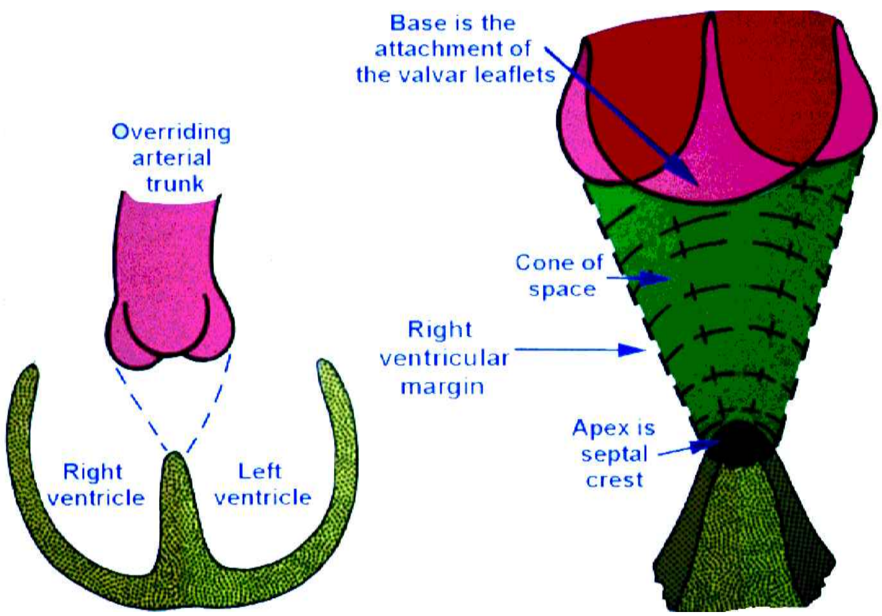

Figure 8.
When an arterial valve overrides the crest of the ventricular septum (left hand panel), a cone of space is created between the circumference of the valvar leaflets and the crest of the apical ventricular septum (right hand panel). Any plane within this cone can, with justification, be nominated as the "ventricular septal defect".

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outlet septum so as to re-connect the aorta with the left ventricle. In the heart illustrated in Figure 12, it may also be necessary to enlarge the interventricular communication, which forms the outlet from the left ventricle. This fact reinforces the importance of recognising the ventriculo-arterial connection as double outlet.

Overriding of an arterial valve exists in various patterns when the potential anatomic diagnosis is double outlet right ventricle. Each pattern represents a spectrum of malformation between two extremes. The commonest spectrum is found in the setting of overriding of the aortic valve, namely the spectrum between tetralogy of Fallot with concordant (Fig. 10) and double outlet ventriculo-arterial connections (Figs 11 & 12). The other well-recognised spectrum is found when the pulmonary valve overrides the crest of the muscular ventricular septum. One end of this spectrum is double outlet right ventricle with sub-pulmonary interventricular communication. The other end is represented by discordant ventriculo-arterial connections with ventricular septal defect. We consider this overall spectrum as encompassing the variants of the Taussig-Bing malformation.[22] There is then a third spectrum, to be found in absence of the muscular outlet septum when both arterial trunks override the muscular ventricular septal crest. This arrangement has been dubbed "double outlet both ventricles".[23] Our preference when dealing with this spectrum, as with all the others, is to consider the hearts with overriding valves as intermediates within the spectrum. In terms of the ventriculo-arterial connections, we assign them to one or other end of the spectrum, determining the arrangement in each individual case with the degree of precision made possible by the diagnostic techniques available. The use of this approach then permits us to make a pragmatic definition of double outlet ventriculo-arterial connection, namely the situation in which the greater part of the circumferences of both arterial valves are supported within the same ventricle.

Variability within double outlet ventriculo-arterial connection

Since our chosen definition for double outlet right ventricle includes hearts only according to the arrangement of their ventriculo-arterial junctions, it follows that there is marked variability to be found elsewhere within the hearts thus grouped

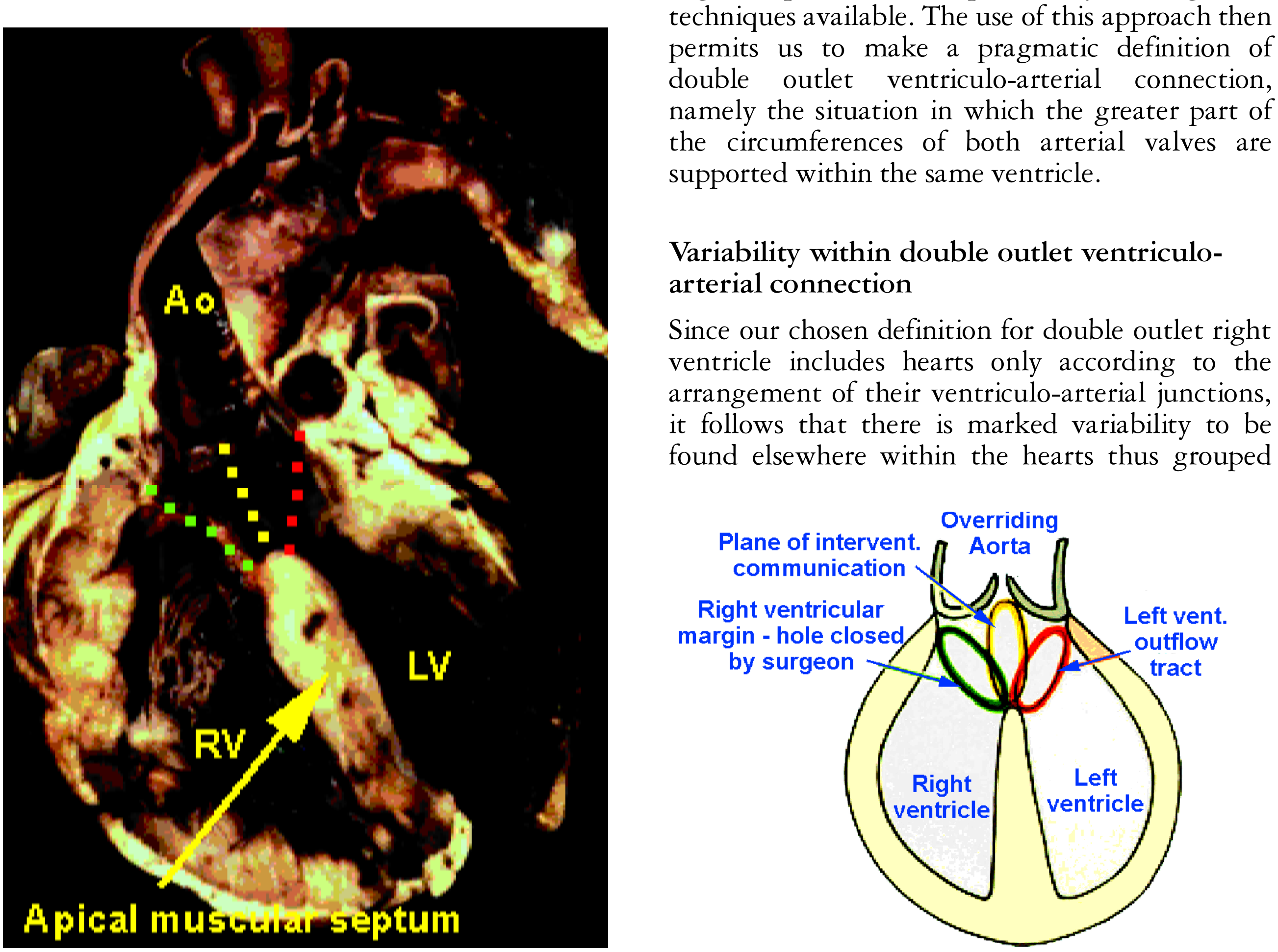

Figure 9.
The left hand panel shows a simulated "four-chamber" section through the aortic root in a heart with tetralogy of Fallot. Three planes are shown, and labelled in the right hand panel. The interventricular communication (yellow) is the direct upward continuation of the plane of the apical ventricular septum. It extends to the undersurface of the leaflets of the aortic valve. The left ventricular boundary (red) is the outflow from the left ventricle. The right ventricular boundary (green) is the plane closed by the surgeon so as to re-connect the aorta with the left ventricle. In this particular heart, the right border is muscular, whilst the other two planes have fibrous components. See text for further discussion.

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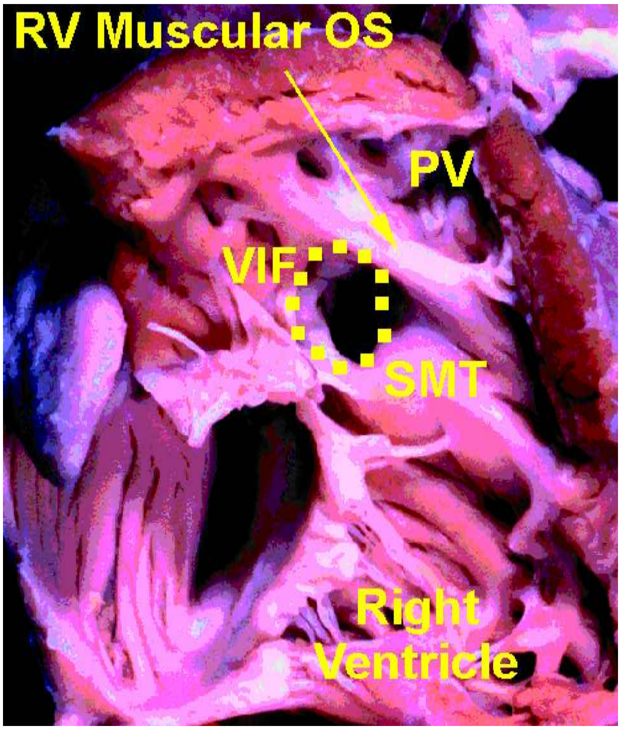

Figure 10.
This specimen exhibits the typical morphology of tetralogy of Fallot. The "ventricular septal defect" is usually considered to represent the locus shown by the yellow dots. This is the right ventricular boundary of the cone of space subtended from the attachments of the leaflets of the overriding aortic valve.

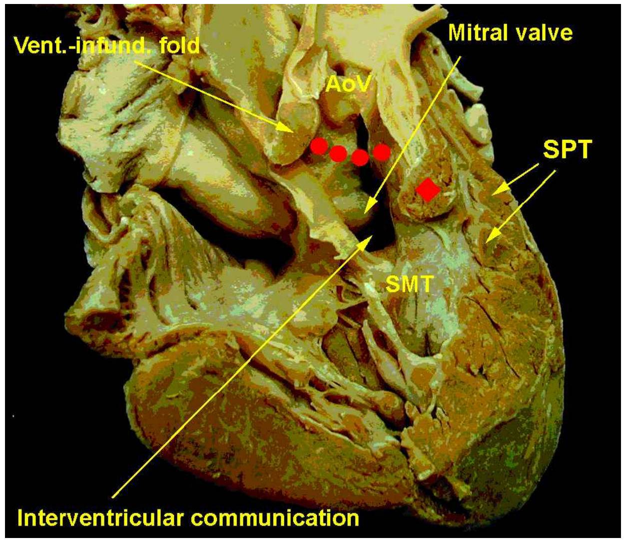

Figure 11.
This specimen also shows the typical morphology of the sub-pulmonary outflow tract as seen in tetralogy of Fallot, with obstruction produced between the right ventricular outlet septum (red diamond) and the hypertrophied septoparietal trabeculations (SPT). In this heart, however, the aorta arises exclusively from the right ventricle. The interventricular communication, between the limbs of the septomarginal trabeculation, is bounded superiorly by the ventriculo-infundibular (vent.-infund.) fold, which separates the leaflets of the aortic and mitral valves (red dotted line), producing a completely muscular subaortic infundibulum.

together. Most frequently, there will be usual arrangement of the atrial appendages ("solitus"), but the mirror-imaged variant ("inversus") may be found, or there can be isomerism of either the morphologically right or left atrial appendages ("visceral heterotaxy"). The atrioventricular connections typically are concordant, but they can be discordant, ambiguous and biventricular, double inlet, or one atrioventricular connection can be absent ("atrioventricular valvar atresia"). Associated malformations can be found in any part of the heart. There can be marked variability in the relationship of the arterial trunks (Fig. 13). As already discussed, and as emphasised by Lev and his colleagues,[24] the infundibular morphology can vary from bilateral muscular support of both arterial valves (Figs 4,11), to different patterns of fibrous continuity between the arterial and atrioventricular valves (Figs 5,12). All of these features need to be investigated and described so as to provide a complete diagnosis. Without question, however, it is the arrangement of the ventricular septum, and the relationship of the interventricular communication to the sub-arterial outlets, which is of greatest clinical significance, the feature also brought to attention by Lev et al.[24] It is these variations on which we will concentrate in the remainder of our review. The understanding of the anatomic variations can be facilitated by making clear distinctions between the various muscular structures which bound the ventricular outflow tracts. We have already seen how variations in such definitions can lead to controversy.[10,11]

Definition of components of the sub-arterial outlets

Traditionally, the building blocks of the sub-arterial outflow tracts were considered in terms of the "crista supraventricularis", which translates as the supraventricular crest. In the normal heart, this muscular structure separates the leaflets of the tricuspid and pulmonary valves in the roof of the right ventricle, inserting between the limbs of a prominent muscular strap which reinforces the septal surface of the right ventricle. The septal component is still nominated by some as the septal band of the "crista supraventricularis",[5] despite the fact that it does not occupy a supraventricular position. We prefer to consider this septal strap as the septomarginal trabeculation. We then

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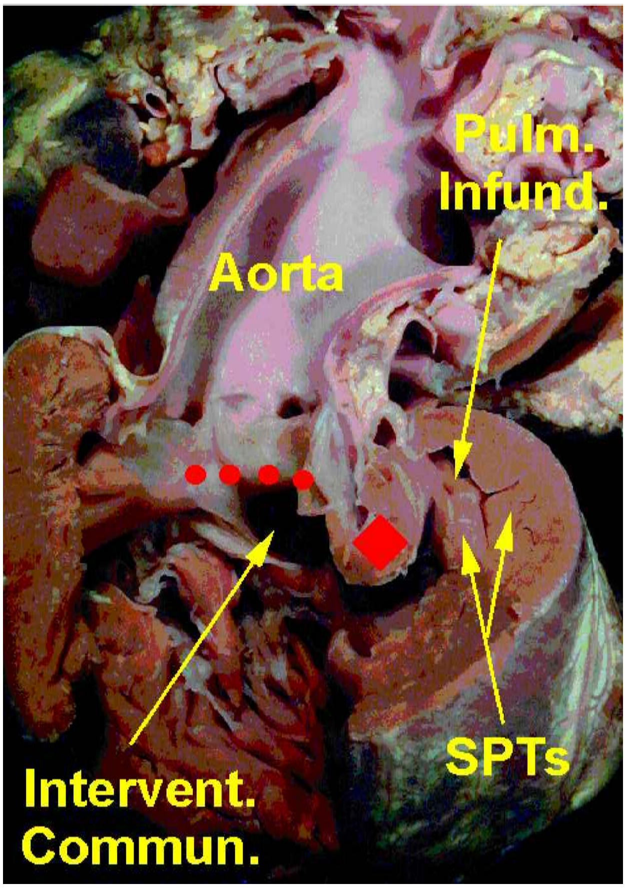

Figure 12.
As with the heart shown in Figure 11, both arterial trunks in this specimen arise entirely from the right ventricle, and the sub-pulmonary outflow tract shows the typical morphology for tetralogy of Fallot. In this heart, however, there is fibrous continuity between the leaflets of the aortic and mitral valves (red dotted line), forming the roof of the sub-aortic interventricular communication. The red diamond shows the right ventricular muscular outlet septum. Other abbreviations as before.

recognise that it has a body, anterior and posterior limbs, and ramifies at the apex of the right ventricle into the typical coarse trabeculations of this chamber (Fig. 14). A series of additional trabeculations are then found extending from the anterior surface of the septomarginal trabeculation to the parietal ventricular wall. These bundles were named the septoparietal trabeculations by Goor and Lillehei.[25] In the presence of a ventricular septal defect, and particularly when both arterial trunks arise from the right ventricle, the components of the supraventricular crest itself achieve their own identity. Some call these structures the "parietal band", and then distinguish different parts of this band.[5,24] This can lead to confusion, with distinguished authors, on occasion, describing different structures as the "parietal band".[26] To circumvent

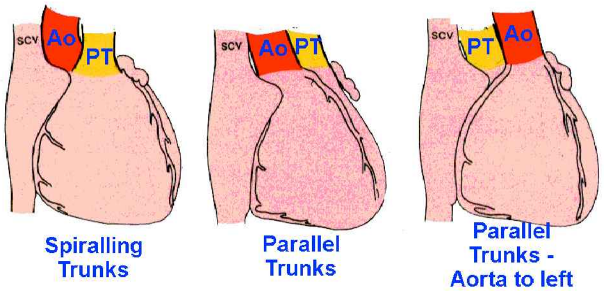

Figure 13.
This cartoon shows the three typical arrangements of the arterial trunks found when both arterial trunks arise exclusively from the right ventricle. The arrangement with the aorta (Ao) to the left of the pulmonary trunk (PT) is by far the rarest variant. SCV – superior caval vein.

this confusion, we proposed many years ago that it is better to define precisely the different components of the supraventricular crest, and give each its own name.[11] Thus, part of the supraventricular crest is the inner heart curvature, separating the leaflets of the atrioventricular and arterial valves. We call this structure the ventriculo-infundibular fold. The fold can be found in the roof of either the right or the left ventricle. The other part of the crest is much more prominent in the presence of an interventricular communication. This is the part which separates the leaflets of the aortic and pulmonary valves, and, by extension, also separates the sub-pulmonary from the sub-aortic outflow tract. Usually this structure is made of muscle, and is then described as the muscular outlet, or infundibular, septum. As we have already seen, this

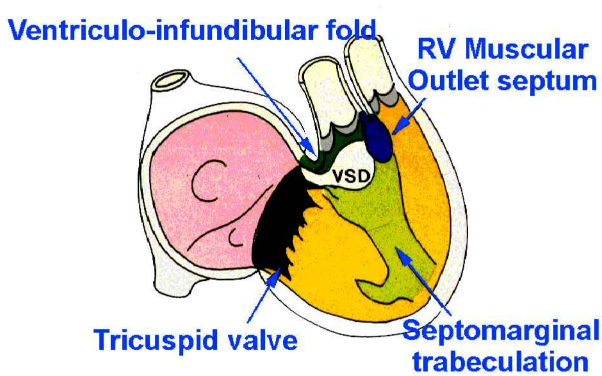

Figure 14.
This cartoon shows the three main "building blocks" of the sub-arterial outflow tracts, namely the muscular outlet septum, the ventriculo-infundibular fold, and the septomarginal trabeculation. Also of importance are the septoparietal trabeculations and the free-standing infundibular sleeves, albeit not shown in this diagram.

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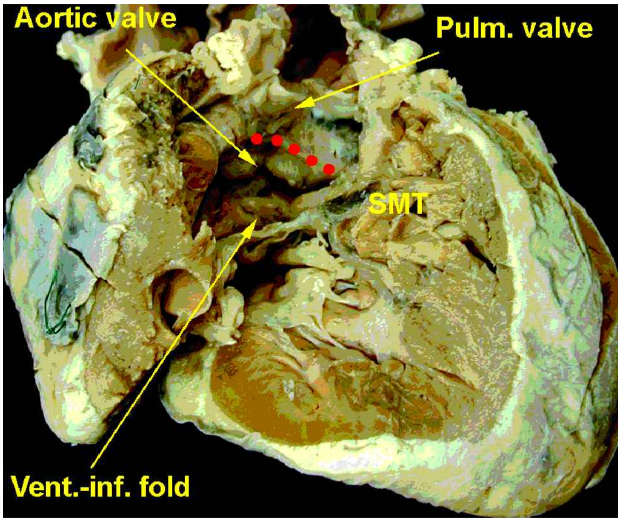

Figure 15.
In this specimen, in which the interventricular communication opens directly beneath both arterial valves, there is absence of the muscular outlet septum. Instead, the leaflets of the arterial valves are in direct fibrous continuity (red dotted line). Note in this specimen how the ventriculo-infundibular (vent.-inf.) fold fuses with the posterior limb of the septomarginal trabeculation, interposing between the leaflets of the tricuspid and aortic valves, and protecting the atrioventricular conduction axis.

muscular outlet septum is an interventricular structure when there are one-to-one ventriculo-arterial connections (Fig. 6). In the setting of double outlet right ventricle, in contrast, the muscular outlet septum is exclusively a right ventricular structure (Figs 2,4,6,7,11,12). It also follows that, should the ventriculo-arterial connection be double outlet left ventricle, the muscular outlet septum would be an exclusively left ventricular structure. Since we first defined these structures, however, we have become aware of the importance of an additional muscular component of the outflow tracts. This is the sleeve of free-standing musculature that lifts the arterial valves away from the base of the ventricle in the setting of complete muscular infundibulums[27]. Sometimes, this sleeve of musculature can be completely absent, along with the muscular outlet septum. In this setting, the arterial valvar leaflets will be in fibrous continuity, albeit that a fibrous raphe can be found between the sub-arterial outlets in the ventricular roof (Fig. 15). These definitions are particularly important for the surgeon, since they are guides to the areas which are at greatest danger during operative procedures (Fig. 16). Thus, the muscular outlet septum is a true septal structure which can be excised with exiting from the cavity of the heart. This structure never harbours conduction tissues. The ventriculo-infundibular fold, irrespective of its right or left ventricular location, is also free from conduction tissues. But, if the surgeon resects this structure, then an opening is made to the inner curvature of the heart. This can place at risk the major coronary arteries, which occupy the atrioventricular grooves bounded by the fold. The septoparietal trabeculations can be resected with impunity, since they harbour neither conduction tissues nor coronary arteries. The septomarginal trabeculation, in contrast, can be a danger area. The bundle of His penetrates in relation to its posterior limb, while the septal perforating artery runs in relation to its anterior limb. It is the anterior limb, nonetheless, which provides the safest area for resection should it prove necessary to enlarge the interventricular communication (Fig. 16). The right bundle branch runs down within the body of the septomarginal

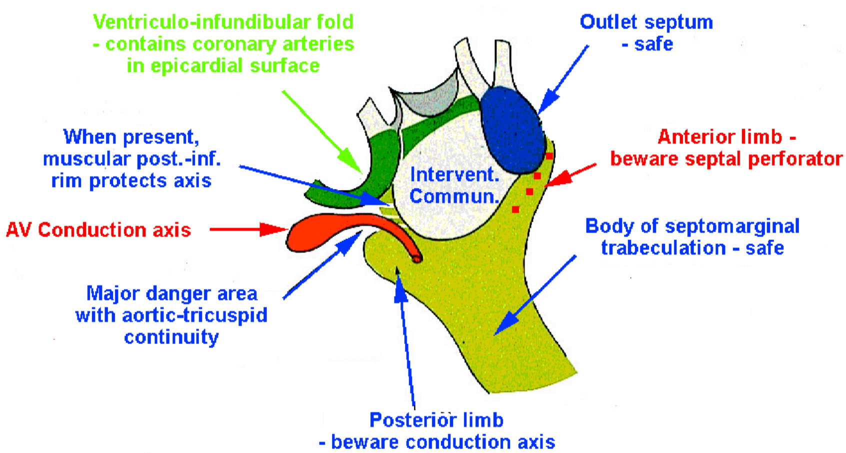

Figure 16.
This cartoon shows the potential dangers to be found in the environs of the interventricular communication when both arterial trunks arise exclusively from the right ventricle.

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trabeculation from the site of the medial papillary muscle, crossing the cavity of the right ventricle in the moderator band. It is known that haemorrhage can track cranially in the sheath surrounding this bundle[28], so it is safest also to avoid resection of the body of the septomarginal trabeculation.

The interventricular communication

As was emphasised by Lev and his colleagues,[24] it is the location of the interventricular communication, particularly its relationship to the sub-arterial outlets, which underscores the clinical presentation of patients with double outlet ventriculo-arterial connection from the right ventricle. In this respect, de la Cruz and her associates[29] highlighted the relationships of the arterial trunks as a useful guide to the location of the septal defect. Thus, when the arterial trunks spiral round one another, in the arrangement known as "normal relations", then most frequently the ventricular septal defect is in sub-aortic position, giving the so-called "Fallot" variant of double outlet. When the arterial trunks arise in parallel fashion, with the aorta in anterior position, then typically the interventricular communication is sub-pulmonary. Indeed, some still continue to use this anterior location of the aorta as their definition for "transposition".[30] For those who continue to espouse this usage, there is no reason why double outlet cannot co-exist with transposition. It to avoid this potential conflict that we prefer to describe the ventriculo-arterial connections as discordant when the arterial trunks arise from morphologically inappropriate ventricles, and account directly for an anterior location of the aorta irrespective of its ventricular origin. There is then a third variation in arterial relationships when both trunks arise from the right ventricle (Fig. 13). This is when the aorta arises anteriorly and to the left relative to the pulmonary trunk. In this situation, the interventricular communication tends to be sub-aortic, and the right coronary artery crosses in front of the sub-pulmonary outflow tract.[31,32] None of these relationships, however, is found with sufficient constancy to provide a reliable guide to the location of the interventricular communication. The arterial relationships should be described, therefore, along with infundibular morphology, but the location and morphology of the interventricular communication must be also be described in their own right.

The thrust of the important study of Lev and his associates[24] was to show that the interventricular communication could occupy sub-aortic, sub-pulmonary, doubly committed or non-committed positions relative to the sub-arterial outlets. This convention has stood remarkably well the passage of time, and remains an important descriptor for hearts with double outlet right ventricle. When seen in diagrammatic format, however, (Fig. 17), the impression is gained that the interventricular communication "moves" around the ventricular septum. In reality, when the defect is directly related to the sub-arterial outlets, it occupies a constant position between the two limbs of the septomarginal trabeculation. It is then the variability in the arrangement and attachment of the right ventricular outlet septum which determines the commitment of the interventricular communication. Furthermore, it is the relationship between the posterior limb of the septomarginal trabeculation and the ventriculo-infundibular fold which determines the important structure of the postero-inferior margin of the communication, this reflecting the vulnerability of the atrioventricular conduction axis. Thus, recognition of the interrelationships of the muscular structures making up the outflow tracts is the key to diagnosis of the location and morphology of the interventricular communication.

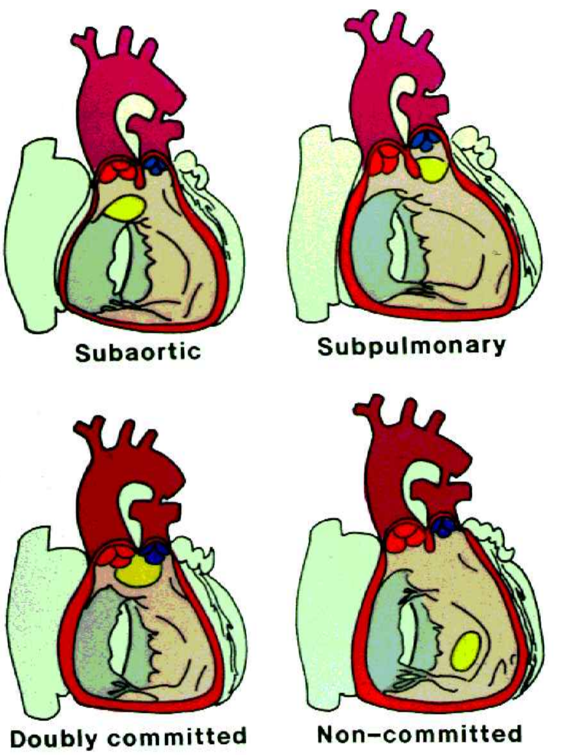

Figure 17.
A cartoon showing the interrelationships between the interventricular communication and the sub-arterial outlets as emphasised in the now classical paper by Lev and his colleagues[24]. See text for further discussion.

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In some hearts, however, the ventricular septum can be intact when both arterial trunks arise from the right ventricle (Fig. 18). This arrangement is rare, and presumably reflects closure of a pre-existing septal defect during fetal life. The existence of such hearts, nonetheless, points to another difficulty in replacing the term "double outlet right ventricle" with "malposition of the great arteries with ventricular septal defect" as suggested by Sidi and Lecompte![10]

When the interventricular communication is in sub-aortic position, and the arterial trunks spiral as they leave the ventricular mass, then the septal attachment of the right ventricular outlet septum is to the anterior limb of the septomarginal trabeculation. The septal defect itself is between the limbs of the septomarginal trabeculation. In most instances with this arrangement, the postero-inferior margin of the defect is fibrous due to continuity between the leaflets of the mitral and tricuspid valves, even in the presence of bilateral infundibulums (Fig. 4). The postero-inferior rim, nonetheless, can be muscular (Fig. 19). The presence of a muscular sub-aortic infundibulum itself lifts the leaflets of the aortic valve away from upper margin of the interventricular communication formed by the ventriculo-infundibular fold. When the infundibulum is particularly long, as in the heart shown in Figure 4, some would nominate the defect as being non-committed. This is an individual judgement. Our own view is that, since no structures interpose in the path which would be used by the surgeon to create a tunnel from the interventricular communication to the sub-aortic infundibulum, the defect should still be considered as sub-aortic. When there is fibrous continuity between the leaflets of the aortic and mitral valves (Fig. 12), then the length of the tunnel will be much shorter, recognising that, on occasions, the interventricular communication may itself be restrictive. The precise location of the septal attachment of the right ventricular outlet septum, and the arrangement of the septoparietal trabeculations, will determine whether the sub-pulmonary outlet shows the morphology of tetralogy of Fallot (Figs 11,12) or is unobstructed (Figs 4,19).

In a second sub-set of hearts with double outlet right ventricle, the key to diagnosis is complete absence of the muscular right ventricular outlet septum (Fig. 15). The septal defect itself remains between the limbs of the septomarginal trabeculation. The roof of the common ventricular outlet component, however, is divided by a fibrous raphe separating the conjoined facing leaflets of the aortic and pulmonary valves. These hearts have the defect in doubly committed location, and in addition the

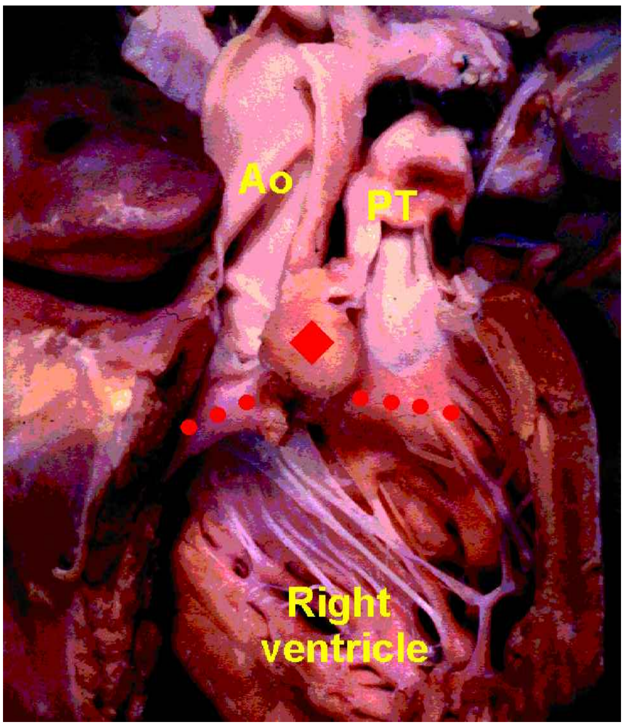

Figure 18.
In this specimen, both arterial trunks arise exclusively from the right ventricle in the setting of an intact ventricular septum. Abbreviations and symbols as before.

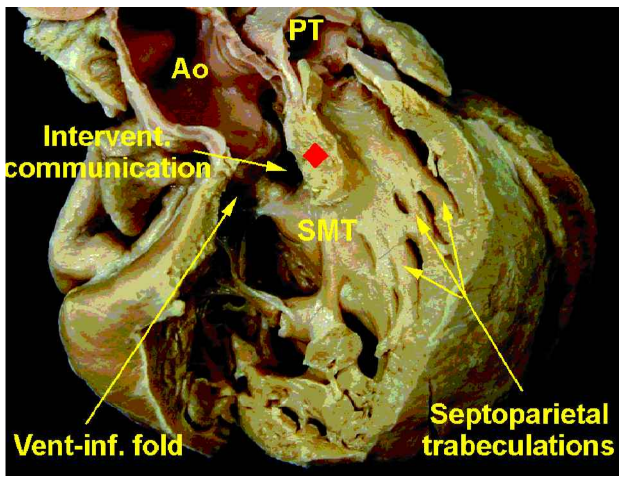

Figure 19.
In this specimen, both arterial trunks arise exclusively from the right ventricle, and the sub-pulmonary outlet is unobstructed, despite the presence of obvious septoparietal trabeculations. The roof of the interventricular communication is formed by fibrous continuity between the leaflets of the aortic and mitral valves. Note, however, that the ventriculo-infundibular fold fuses with the posterior limb of the septomarginal trabeculation (SMT) to give a muscular postero-inferior rim to the defect which protects the atrioventricular conduction axis. The red diamond marks the right ventricular outlet septum.

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defect is directly juxta-arterial. The postero-inferior rim is usually muscular, due to fusion of the posterior limb of the septomarginal trabeculation with the ventriculo-infundibular fold (Figs 15,20), but the defect can be perimembranous when the postero-inferior rim is fibrous, this arrangement placing the conduction axis at greater risk (Fig. 21). The fibrous raphe between the arterial valvar leaflets can itself be deviated towards the aorta (Fig. 20) or the pulmonary trunk (Fig. 21), producing narrowing of one or other outflow tract. The arterial valves themselves can also override the septum, leading to a spectrum of anomalies ending in double outlet from the left ventricle.[23]

In the third sub-set of hearts with the interventricular communication committed to the ventricular outflow tracts, the defect itself continues to be positioned between the limbs of the septomarginal trabeculation (Fig. 5). In these hearts, the key feature is the attachment of the right ventricular muscular outlet septum to the ventriculo-infundibular fold rather than to the anterior limb of the septomarginal trabeculation and the underlying muscular ventricular septum (Figs 5, 22). This location of the right ventricular outlet septum commits the interventricular communication to sub-pulmonary position. Should the ventriculo-infundibular fold also fuse with the septomarginal trabeculation, then the conduction axis will be protected. In contrast, when the defect is perimembranous, with fibrous continuity between the mitral and tricuspid valves (Fig. 23), and usually also the pulmonary valve (Fig. 5), then the conduction bundle is exposed in the region of fibrous continuity as it penetrates from the apex of the triangle of Koch. These hearts are at the double outlet end of the Taussig-Bing malformation.[22,24] Frequently the right ventricular outlet septum is deviated so as to produce sub-aortic infundibular stenosis, and usually there is then severe aortic coarctation, or even interruption of the aortic arch. It is also frequent to find straddling and overriding of the mitral valve, often with a cleft in the straddling leaflet.[33,34]

The final sub-set of hearts divided according to the location of the interventricular communication is made up of those with non-committed defects. We have already discussed the difficulties in determining the definition of commitment in the presence of a long sub-arterial infundibulum. Difficulties can also be produced for the surgeon when an anatomically committed defect is rendered non-committed by presence of tension apparatus of the atrioventricular valves crossing the

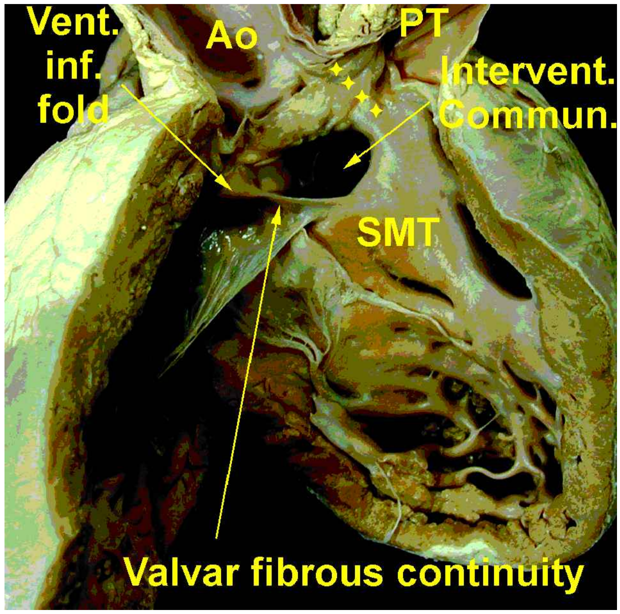

Figure 21.
This specimen, like the one shown in Fig. 20, again has a doubly-committed interventricular communication. In this heart, the fibrous raphe (yellow dots) is deviated in antero-cephalad direction, giving mild sub-pulmonary obstruction. Note that there is fibrous tissue in the postero-inferior rim of the defect, making it perimembranous. The conduction axis is at risk beneath the fibrous flap forming the postero-inferior margin. Abbreviations as before.

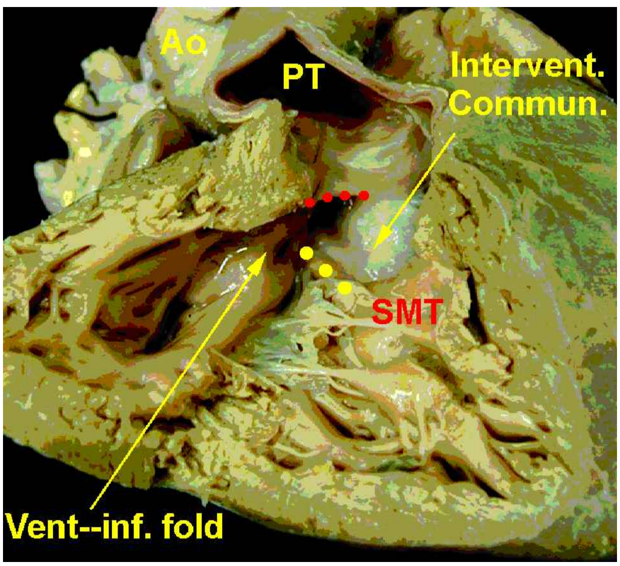

Figure 20.
In this specimen with a doubly-committed interventricular (intervent.) communication (commun.), the fibrous raphe between the arterial valvar leaflets (red dotted line) is deviated posteriorly to produce sub-aortic narrowing. Note how the posterior limb of the septomarginal trabeculation (SMT) fuses (yellow dots) with the ventriculo-infundibular (vent.-inf) fold, protecting the atrioventricular conduction axis.

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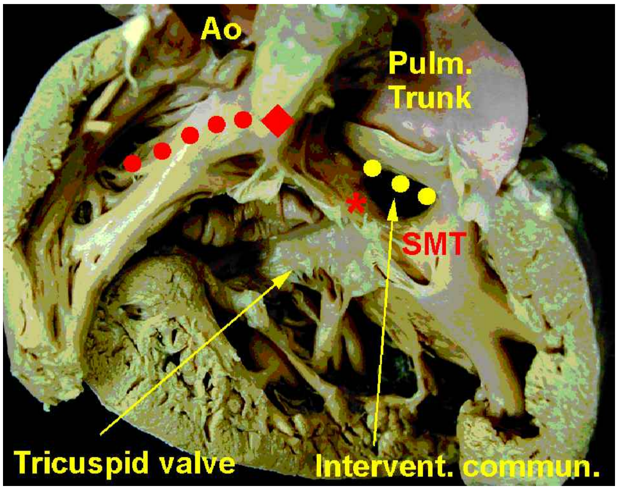

Figure 22.
In this heart with double outlet right ventricle and sub-pulmonary interventricular communication, the so-called "Taussig-Bing" malformation, the posterior limb of the septomarginal trabeculation (asterisk) fuses with the right ventricular outlet septum (red diamond) to produce a muscular postero-inferior rim to the defect which protects the conduction axis. Note that, although the aortic valve has a completely muscular infundibulum (red dotted line), the pulmonary valve is in fibrous continuity with the mitral valve in the roof of the interventricular communication (yellow dots).

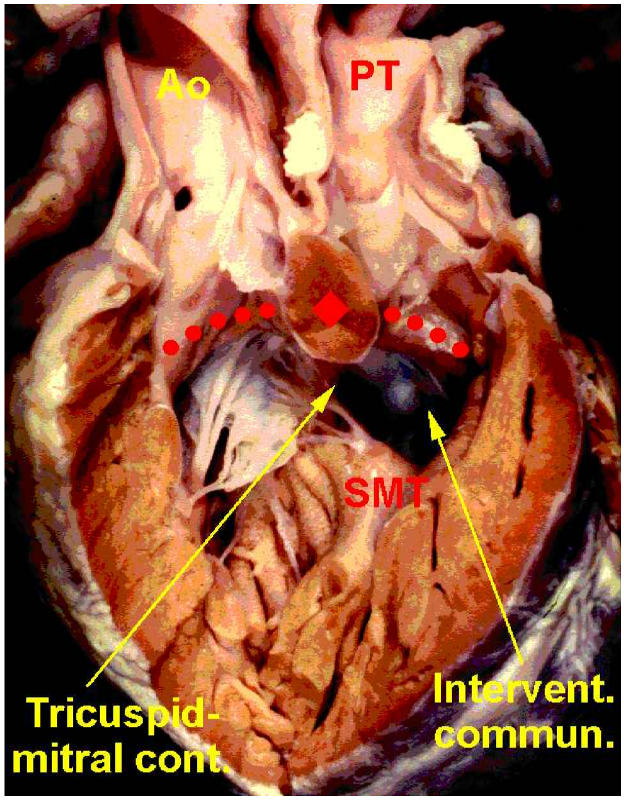

Figure 23.
This heart, as with the specimen shown in Fig. 19, is an example of the "Taussig-Bing" malformation, having double outlet right ventricle with sub-pulmonary interventricular communication. In this specimen, however, there are bilateral infundibulums (red dotted lines), but the defect is perimembranous because of fibrous continuity between the leaflets of the tricuspid and mitral valves. Note that the right ventricular outlet septum (red diamond) is inserted to the mid-point of the ventriculo-infundibular fold.

potential location of an interventricular tunnel.[35] Nowadays, such complications should be diagnosed pre-operatively using cross-sectional echocardiography. There remain further hearts, nonetheless, in which the defect is unequivocally non-committed because it is no longer positioned between the limbs of the septomarginal trabeculation. For example, muscular defects can be found opening to the inlet of the right ventricle (Fig. 24). Although we have not seen such a case, muscular defects must also be anticipated at the apex of the right ventricle. Perimembranous defects can similarly occupy a non-committed position when they open to the inlet of the right ventricle (Fig.25), and defects in this location can be found with common atrioventricular junctions. Problems can then arise in determining when such defects are non-committed or sub-aortic (Fig. 26), and resolution of this debate this will again be a matter for personal judgement.

Additional associated malformations

We have emphasised throughout our review that double outlet connection involves only a small part of the congenitally malformed heart, and that full diagnosis demands complete sequential analysis. We have also focussed attention on the associated malformations, particularly the interventricular communication. It will be appreciated, nonetheless, that malformations can also co-exist in all other parts of the heart, and all must be diagnosed. For example, the case described by Birmingham[15] was associated with left juxtaposition of the atrial appendages, and we now know that this arrangement is frequent in the hearts with double outlet having a left-sided and anterior aorta. Abnormal venous connections will be anticipated whenever there is isomerism of the atrial appendages, with problems of pulmonary venous connection expected with right isomerism, and interruption of the inferior caval vein with left isomerism. And so on. Space does not permit us to catalogue all the associated malformations but, when present, each must be recognised, since any malformation can be of potential clinical significance.

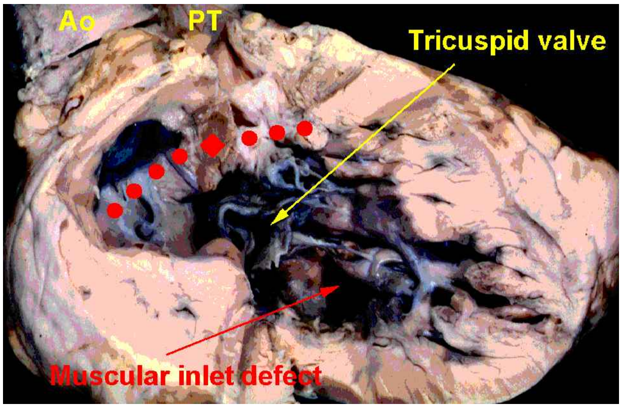

Figure 24.
In this heart, again with bilateral infundibulums (red dotted lines), and with the right ventricular outlet septum (red diamond) inserted to the mid-point of the fold, the interventricular communication is enclosed within the muscular septum, opening to the inlet of the right ventricle. The tricuspid valve is interposed between the defect and both outflow tracts, making it non-committed.

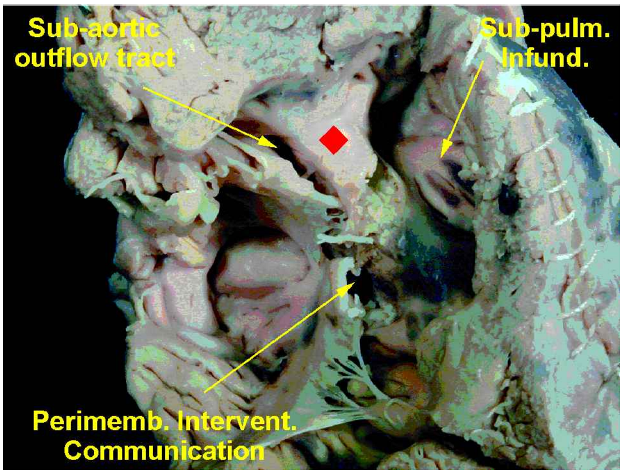

Figure 25.
This heart again has bilateral infundibulums, with the right ventricular muscular outlet septum (red diamond) separating them. The interventricular communication is directly beneath the septal leaflet of the tricuspid valve, and is perimembranous. Its position, and its relationship to the tricuspid valve, make it non-committed.

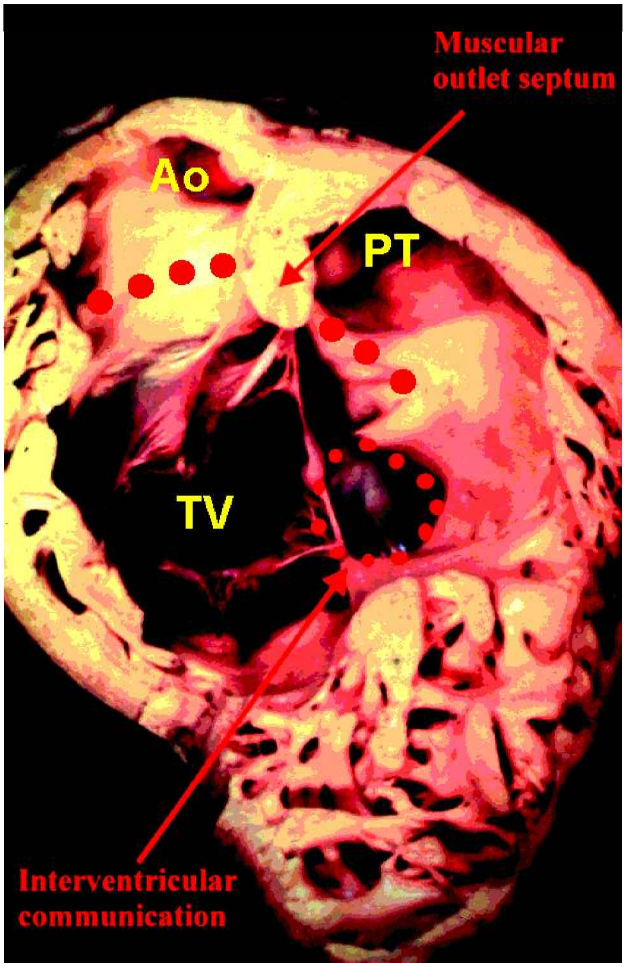

Figure 26.
Like the specimen shown in Fig. 25, this heart has bilateral infundibulums (red dotted lines) and a perimembranous interventricular communication opening to the inlet of the right ventricle. Although well separated from the pulmonary infundibulum, it would be feasible to construct a long tunnel between the defect and the pulmonary valve. Despite this, we would classify this defect as being non-committed, but we would recognise the possibility of considering it to be sub-pulmonary.

Conclusions

In the forty or so years which have passed since Witham[17] coined the term "double outlet right ventricle", we have learnt much concerning this particular ventriculo-arterial connection, albeit that the evolution of knowledge has not always been straightforward. For many years, controversy focussed on the necessity for finding bilateral infundibular structures so as to diagnose this connection. In our opinion, this controversy was resolved by Van Praagh and his colleagues[9] when they introduced their important "morphological method". If we accept this sensible principle, namely that one variable morphologic feature cannot be used to define another feature which is itself variable, then it follows that a specific infundibular arrangement cannot be used to determine a particular ventriculo-arterial connection or alignment. The objections raised by Sidi and Lecompte[10] to the continued use of double outlet are much more substantial, and worthy of reasoned debate. In essence, we infer from their chapter that they believe the term has outlived its value, since markedly different surgical procedures are now needed to repair the various anatomic patterns found in hearts with both arterial trunks supported by the morphologically right ventricle. This is undoubtedly true, and it is also true that the innovative concepts proposed by Lecompte and his colleagues[36,37] have done much to improve the selection of the most appropriate surgical procedure. They state, however, "…. the only way to achieve appropriate surgical decisions is not to classify subgroups of patients anatomically but to consider each patient with malposition and ventricular septal defect as unique." We agree with the second part of this statement, but we are at a loss to understand how the uniqueness can be assessed without paying full attention to the anatomic characteristics. Thus, in our opinion, the arguments marshalled by Sidi and Lecompte[10] in no way detract from the value of double outlet right ventricle as a diagnostic term. On the contrary, recognition of the fact that both arterial trunks are supported predominantly by the outflow tract of the right ventricle enhances the concepts they propose in terms of surgical treatment. This is because, when both arterial trunks arise from the right ventricle, the outlet septum is, of necessity, a right ventricular structure. The structure which they have discussed "at the left edge of the conus"[10] is not an infundibular septum, as they imply. It is appreciation of this fact which focuses attention on the interventricular communication. As long ago as 1793, this defect was recognised as functioning as the outflow tract of the left ventricle when both arterial trunks arose from the right ventricle.[12] This single feature, in its own right, continues to substantiate the need for recognition of double outlet right ventricle as a diagnostic entity, and its distinction from hearts with discordant or concordant ventriculo-arterial connections.

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References

1. Edwards WD. Double-outlet right ventricle and tetralogy of Fallot. Two distinct but not mutually exclusive entities. J Thorac Cardiovasc Surg. 1981;82: 418–422.
2. Wilcox BR, Ho SY, Macartney FJ, Becker AE, Gerlis LM, Anderson RH. Surgical anatomy of double-outlet right ventricle with situs solitus and atrioventricular concordance. J Thorac Cardiovasc Surg 1981;82: 405–417.
3. Van Praagh S, Davidoff MD, Chin A, Shiel FS, Reynolds J, Van Praagh R. Double outlet right ventricle: anatomic types and developmental implications based on a study of 101 autopsied cases. Coeur 1982;13: 389–440.
4. Baron MG. Radiologic notes in cardiology: angiographic differentiation between tetralogy of Fallot and double outlet right ventricle. Circulation 1971;43: 451–455.
5. Van Praagh R. What is the Taussig-Bing malformation? Circulation 1968;38: 445–449.
6. Hinkes P, Rosenquist GC, White RIJ. Roentgenographic reexamination of the internal anatomy of the Taussig-bing heart. Am Heart J 1971;81: 335–339.
7. Kirklin JW, Pacifico AD, Bargeron LM, Soto B. Cardiac repair in anatomically corrected malposition of the great arteries. Circulation 1973;48 153–159.
8. Macartney FJ, Rigby ML, Anderson RH, Stark J, Silverman NH. Double outlet right ventricle. Cross sectional echocardiographic findings, their anatomical explanation, and surgical relevance. Br Heart J 1984;52: 164–177.
9. Van Praagh R, David I, Wright GB, Van Praagh S. Large RV plus small LV is not single RV. Circulation 1980;61: 1057–1058.
10. Sidi D, Lecompte Y. Transposition and Malposition of the great arteries with ventricular septal defects. In: Moller JH, Hoffman JIE (eds). Pediatric Cardiovascular Medicine. New York: Churchill Livingstone. 2000: pp 363–373
11. Anderson RH, Becker AE, Van Mierop LHS. What should we call the "crista"? Br Heart J 1977;39: 856–850.
12. Abernethy J. Surgical and Physiological Essays. London: James Evans. 1793: p163
13. Farre JR. Pathological Research. London: Longman, Hurst, Orme and Brown. 1814: 1–46.
14. von Rokitansky CF. Die Defecte der Scheidewande des Herzens. Vienna: Pathologisch-Anatomisch Wilhelm Braumuller, 1875: 27–36.
15. Birmingham A. Extreme anomaly of the heart and great vessels. J Anat Physiol 1893;27: 139–150.
16. Wilkinson JL. Double outlet ventricle. In: Anderson RH, Macartney FJ, Shinebourne EA, Tynan M (eds). Paediatric Cardiology. Edinburgh: Churchill Livingstone. 1987; pp 889–911
17. Witham AC. Double outlet right ventricle. A partial transposition complex. Am.Heart J. 1957;53: 928–933.
18. Neufeld HN, DuShane JW, Wood EH, Kirklin JW, Edwards JE. Origin of both great vessels from the right ventricle. I. without pulmonary stenosis. Circulation 1961;23: 399–412.
19. Neufeld HN, DuShane JW, Edwards JE. Origin of both great vessels from the right ventricle. II. With pulmonary stenosis. Circulation 1961;23: 603–612.
20. Neufeld HN, Lucas RVJ, Lester RG, Adams PJ, Anderson RC, Edwards JE. Origin of both great vessels from the right ventricle without pulmonary stenosis. Br.Heart J. 1962;24: 393–408.
21. Alva C, Rigby ML, Ho SY, Anderson RH. Overriding and biventricular connection of aortic valves. Cardiol.Young 1998;8: 150–164.
22. Stellin G, Zuberbuhler JR, Anderson RH, Siewers RD. The surgical anatomy of the Taussig-Bing malformation. J.Thorac.Cardiovasc.Surg 1987;93: 560–569.

https://doi.org/10.1017/S1047951101000373 Published online by Cambridge University Press

23. Brandt PWT, Calder AL, Barratt-Boyes BG, Neutze JM. Double outlet left ventricle. Morphology, Cineangiography, diagnosis and surgical treatment. Am J Cardiol 1976;38: 897–909.
24. Lev M, Bharati S, Meng CCL, Liberthson RR, Paul MH, Idriss F. A concept of double-outlet right ventricle. J Thorac Cardiovasc Surg 1972;64: 271–281
25. Goor DA, Lillehei CW. Congenital Malformations of the Heart. 1 Ed. New York: Grune and Stratton, 1975: 1–37.
26. Kirklin JW, Karp RB. The tetralogy of Fallot. From a surgical viewpoint. 1970:
27. Merrick AF, Yacoub MH, Ho SY, Anderson RH. Anatomy of the muscular subpulmonary infundibulum with regard to the Ross procedure. Ann Thorac Surg. 2000;69: 556–561.
28. Latham RA, Anderson RH. Anatomical variations in atrioventricular conduction system with reference to ventricular septal defects. Br Heart J 1972;34: 185–190.
29. de la Cruz MV, Cayre R, Arista-Salado Martinez O, Sadowinski S, Serrano A. The infundibular interrelationships and the ventriculoarterial connection in double outlet right ventricle. Clinical and surgical implications. Int J Cardiol 1992;35: 153–164.
30. Van Mierop LHS. Transposition of the great arteries. Clarification or further confusion? Editorial. Am J Cardiol 1971;28: 735–738.
31. Danielson GK, Ritter DG, Coleman HN, DuShane JW. Successful repair of double outlet right ventricle with transposition of the great arteries (aorta anterior and to the left), pulmonary stenosis, and subaortic ventricular septal defect. J Thorac Cardiovasc Surg. 1972;63: 741–746.
32. Lincoln C, Anderson RH, Shinebourne EA, English TAH, Wilkinson JL. Double outlet right ventricle with l-malposition of the aorta. Br Heart J 1975;37: 453–463.
33. Muster AJ, Bharati S, Aziz KU, Idriss FS, Paul MH, Lev M, Carr I, DeBoer A, Anagnostopoulos C. Taussig Bing anomaly with straddling mitral valve. J Thorac Cardiovasc Surg 1979;77: 832–842.
34. Soto B, Ceballos R, Nath PH, Bini RM, Pacifico AD, Bargeron LMJr. Overriding atrioventricular valves. An angiographic-anatomical correlate. Int J Cardiol 1985: 327–340.
35. Stellin G, Ho SY, Anderson RH, Zuberbuhler JR, Siewers RD. The surgical anatomy of double-outlet right ventricle with concordant atrioventricular connection and noncommitted ventricular septal defect. J Thorac Cardiovasc Surg 1991;102: 849–855.
36. Sakata R, Lecompte Y, Batisse A, Borromee L, Durandy Y. Anatomic repair of anomalies of ventriculoarterial connection associated with ventricular septal defect. I. Criteria of surgical decision. J Thorac Cardiovasc Surg 1988; 95: 90–95
37. Lecompte Y, Batisse A, DiCarlo D. Double-outlet right ventricle: a surgical synthesis. Adv Card Surg 1993; 4: 109–118.

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