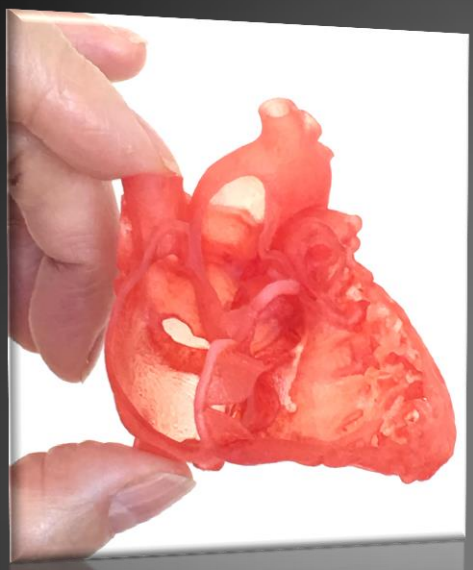

DOUBLE OUTLET RIGHT VENTRICLE IN YOUR HANDS

2nd Edition

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Glen S. van Arsdell

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CONTENTS

List of Abbreviations

Ao, ascending aorta
AV, aortic valve
CS, crista supraventricularis
DORV, double outlet right ventricle
LA, left atrium
LAA, left atrial appendage
LBA, left brachiocephalic artery
LCCA, left common carotid artery
LSA, left subclavian artery
LV, left ventricle
MV, mitral valve
OS, outlet septum
PDA, patent ductus arteriosus
PT, pulmonary arterial trunk
PV, pulmonary valve
RA, right atrium
RAA, right atrial appendage
RBA, right brachiocephalic artery
RCCA, right common carotid artery
RSA, right subclavian artery
RV, right ventricle
TSM, trabecula septomarginalis
TV, tricuspid valve
VIF, ventriculoinfundibular fold
VSD, ventricular septal defect

CHAPTER 1. WHAT'S IN THE NAME?

Historic Background

As far as we know, the earliest description of the condition that we now call "double outlet right ventricle" was in 1793 by Mr. John Abernethy (1764-1831) of London, who was the Professor of Anatomy to the Corporation of Surgeons, the Assistant Surgeon to the St. Bartholomew's Hospital and the Lecturer in Anatomy and Surgery (**Figure 1-1**) (**Table 1-1**) [1]. As a distinguished anatomist, physiologist and surgeon, he began lecturing at his house in Bartholomew Close and speedily attracted a large class. This inspired the governors of St. Bartholomew's to build a lecture theater, and Mr. Abernethy became the founder of the medical school attached to that historic hospital [2]. Mr. Abernethy described the autopsy findings on a male child who died a little more than two years of age with two elegant illustrations (**Figure 1-2**) and text as follows:

> *"From this (right) ventricle, a large vessel proceeded, in the usual course of the pulmonary artery: I laid it open to some extent, but it proved to be the aorta. --- The communication between the ventricle and this artery was large and direct, and it appeared, that it would allow an easy passage of the blood, on the contraction of the ventricle. The pulmonary artery also arose from the right ventricle, but the communicating orifice was small, and the artery was one-third less than its usual size: its sides were uncommonly thin, resembling those of a vein. --- No artery proceeded from the left ventricle, but there was an opening in the upper part of the septum ventriculorum, by which the blood could be projected their blood into the aorta.--- A heart thus constructed, was well calculated for carrying on the foetal circulation, since both ventricles could project their blood into the aorta."*.

Figure 1-1. Mr. John Abernethy's book "Surgical and Physiological Essays" published in 1793 and its title page. A heart with what we now call double outlet right ventricle is described in pages 157-165.

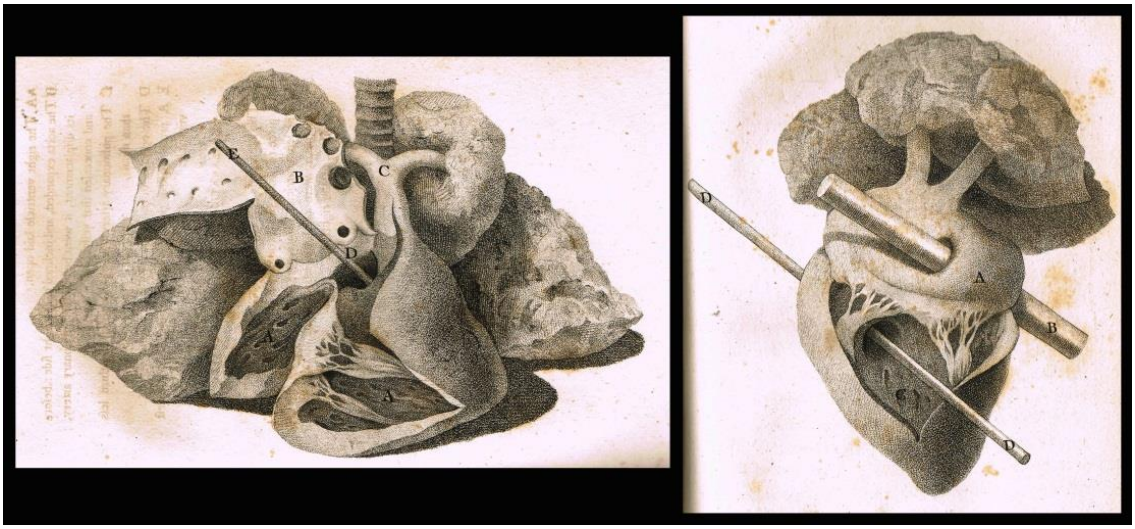

Figure 1-2. Illustrations from the Abernethy's autopsy case. The original legends read:

Left hand plate: AA The right ventricle laid open. B The aorta expanded, and inclined to the right side: before its displacement, it went before the pulmonary artery, and concealed that vessel. C The pulmonary artery, which appeared one-third less than usual. D The opening in the septum ventriculorum. E A probe passed through that opening, into the left ventricle.

Right hand plate: A The left auricle laid open. B A piece of wood put through the foramen ovale. C The left ventricle. Both the cavities of the left side of the heart, were one-third less than the corresponding cavities, of the right side.

In 1898, Vierordt opted to use the term "partial transposition" meaning that only the aorta is transposed above the right ventricle, while the pulmonary arterial trunk is not transposed but maintains its normal origin from the right ventricle [3, 4]. This concept prevailed until the 1950s and the hearts having both great arteries arising from the right ventricle had been regarded as a type of transposition of the great arteries [5-7].

Braun et al used the words "double outlet" in their 1952 report on a postmortem case showing the anatomy of so-called partial transposition of the Fallot type stating [8]:

> *"In the case presented, both the aorta and the stenotic pulmonary artery originate from the right ventricle. It differs from a classic tetralogy of Fallot in that the aorta does not arise from the left ventricle and partially override the ventricular septum, but is transposed and arises entirely from the right ventricle. Thus the right ventricle serves as a "double outlet ventricle"."*

Calhoun Witham introduced the specific term "double outlet right ventricle" in 1957, stating [9]:

> *"The purpose of this report is to describe 4 cases, representing 3 varieties of another rare partial transposition complex characterized by complete aortic transposition with the pulmonary artery in normal position. The description "double outlet right ventricle" has been applied to such anomalies since both great vessels arise completely from this chamber."*

Although Witham's paper cited Braun's case report in the discussion, it did not acknowledge Braun et al's introduction of the words "double outlet ventricle".

In the 1960's, Edwards, Neufeld, et al described a large number of cases with clinicopathological correlation using the long name "origin of both great vessels from the right ventricle" [10-13]. Gradually, however, Witham's "double outlet right ventricle" became the universal term despite ongoing debates and disagreements regarding how it should be defined.

Table 1-1. Evolution and controversy of concept and definition of double outlet right ventricle.

Year	Authors	Remarks
1793	Abernethy J [1]	First description of a heart with origin of both great arteries from the right ventricle
1898	Vierordt H [4]	Partial transposition meaning that only the aorta is transposed above the right ventricle
1923	Spitzer A [5]	Type II (simple transposition) of four types of transposition
1949	Taussig HB and Bing RJ [6]	Complete transposition of the aorta and levoposition of the pulmonary artery
1950	Lev M and Volk BM [7]	Reported a case that they called "Taussig-Bing heart"
1952	Braun K, et al [8]	Both the aorta and stenotic pulmonary artery originate from the right ventricle which serves as a double outlet ventricle.
1957	Witham AC [9]	Earliest use of the specific term "double outlet right ventricle". Complete aortic transposition with the pulmonary artery in the normal position, resulting in both great vessels arising completely from the right ventricle.
1957	Kirklin JW [14]	First surgical repair of double outlet right ventricle with the diagnosis made at operation
1967	Sakakibara S, et al [15]	First report of double outlet left ventricle
1970 1970 1971	Hallerman FJ, et al [16] Deutsch V et al [17] Baron MG [18]	Both aorta and pulmonary artery arise from the right ventricle. An abnormal mitral-aortic relationship is the *sine qua non* of a double-outlet right ventricle.
1972	Lev M, et al [19]	Both arterial trunks emerge almost completely or completely from the right ventricle and there may or may not be mitral aortic or mitral-pulmonic continuity. Suggested four types of DORV according to the relationship between the VSD and the great arteries
1981	Wilcox BR, et al [20]	One form of ventriculoarterial connection in which more than half of both great arteries are connected to the right ventricle.

Definition of Double Outlet Right Ventricle

Double outlet right ventricle (DORV) encompasses a wide spectrum of anatomic malformations that are characterized by origin of both arterial trunks from the right ventricle. Because of the extreme heterogeneity of the hearts classified as DORV, there have been debates on how to define "double outlet right ventricle" (**Table 1-1**) [3, 21, 22]. When it was introduced in the literature in the 1950's, DORV was defined as a condition where both great vessels arise completely from the right ventricle. The major controversy occurred when DORV was considered as a specific pathological entity characterized by the origin of both arterial trunks from the right ventricle with the bilateral muscular infundibulum supporting both arterial valves. When this concept prevailed, this finding was regarded as the *sine qua non* of a DORV [16-18, 23]. Although this concept appears to have a sound anatomic and embryologic basis, it leaves the hearts with both great arteries arising from the right ventricle but showing an aorto-mitral or pulmonary-mitral continuity no place to be categorized [19]. A pathological series showed that bilateral infundibulum was present in only one quarter to one third of the hearts with both great arteries arising exclusively from the right ventricle [24, 25]. On the other hand, bilateral infundibulum was observed in hearts that do not have both great arteries arising from the right ventricle. They include some complex forms of complete or congenitally corrected TGA and so-called anatomically corrected malposition of the great arteries [26-31]. Furthermore, when considering the intracardiac anatomy, the hearts having both great arteries arising from the right ventricle are not a single spectrum of pathology. There are multiple distinct clusters of cases showing very different intracardiac anatomy and each cluster shows a spectrum of abnormality. The variability of the infundibular and intracardiac morphology is responsible for distinctively different clinical manifestations and requires different surgical management options. Therefore, the term "DORV" cannot be regarded as a specific pathological entity. It is simply a specific type of ventriculoarterial connection, which is one of the three facets of the cardiac construction, namely, the morphology, the connection and the relationship [32].

In our discussion, we define the term "DORV" as a form of ventriculoarterial connection in which both the aorta and pulmonary arterial trunk arise entirely or predominantly from the morphologically right ventricle (**Figure 1-3**) [3, 19, 20]. When an arterial valve or valves override(s) the ventricular septum through a ventricular septal defect (VSD), a '50% rule' is applied [20]. By this rule, an overriding arterial trunk is considered as arising from the right ventricle when more than half of the circumference of its valve belongs to the right ventricle (**Figure 1-4**). When the '50% rule' is applied, the extent of override of an arterial valve is determined by observing the short axis of the ventricular mass seen from the apex toward the base [25]. The extent of override seen in an oblique long axis cut of the ventricles can be misleading as the ventricular septum often shows a sigmoid configuration with the subarterial part of the ventricular septum projecting forward relative to the apical muscular part.

As the term "DORV" describes a form of ventriculoarterial connection, it occurs with any combination of visceroatrial situs and atrioventricular connection.

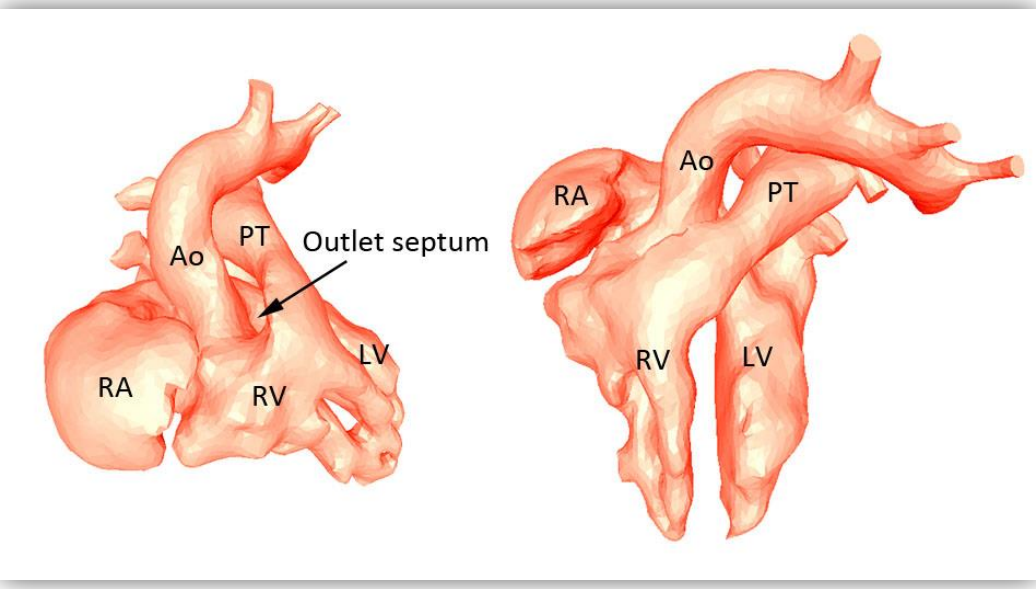

Figure 1-3. Frontal and long-axial views of volume rendered MR angiograms show both pulmonary arterial trunk and aorta arising from the right ventricle.

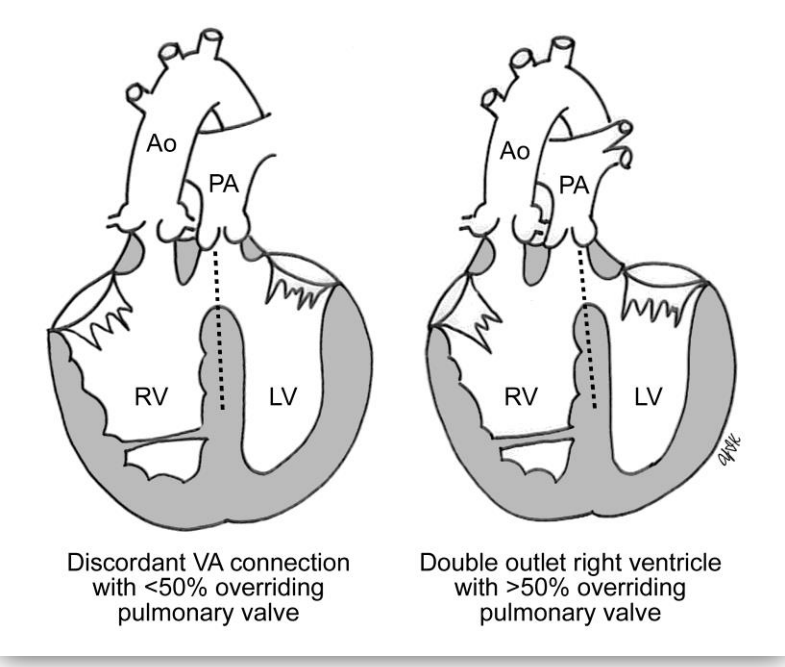

Figure 1-4. The '50% rule' in assigning the commitment of the arterial trunk to a ventricle.

Double outlet right ventricle and tetralogy of Fallot

Tetralogy of Fallot is a specific malformation characterized by anterior, leftward and superior displacement of the outlet or infundibular septum that causes a VSD of an anterior malalignment type, narrowing of the subpulmonary outflow tract, overriding of the aortic valve and right ventricular hypertrophy. Not infrequently, more than 50% of the aortic valve overrides the ventricular septum and therefore belongs to the right ventricle. This subset of tetralogy cases unequivocally has DORV (**Figure 1-5**). Tetralogy of Fallot and DORV represent two different aspects of the abnormality. Among three facets of construction of the heart, i.e., the morphology, the connection and the relationship, tetralogy of Fallot represents the morphology of the right ventricular outflow tract and ventricular septum, whereas DORV describes the connection between the ventricles and arterial trunks. Therefore, the two terms are not mutually exclusive [33]. In an attempt to differentiate DORV and tetralogy of Fallot, Kirklin et al classified the cases with cardiac morphology of tetralogy as DORV only when the aorta arises more than 90% from the right ventricle [34]. We think that this definition is arbitrary and not logical.

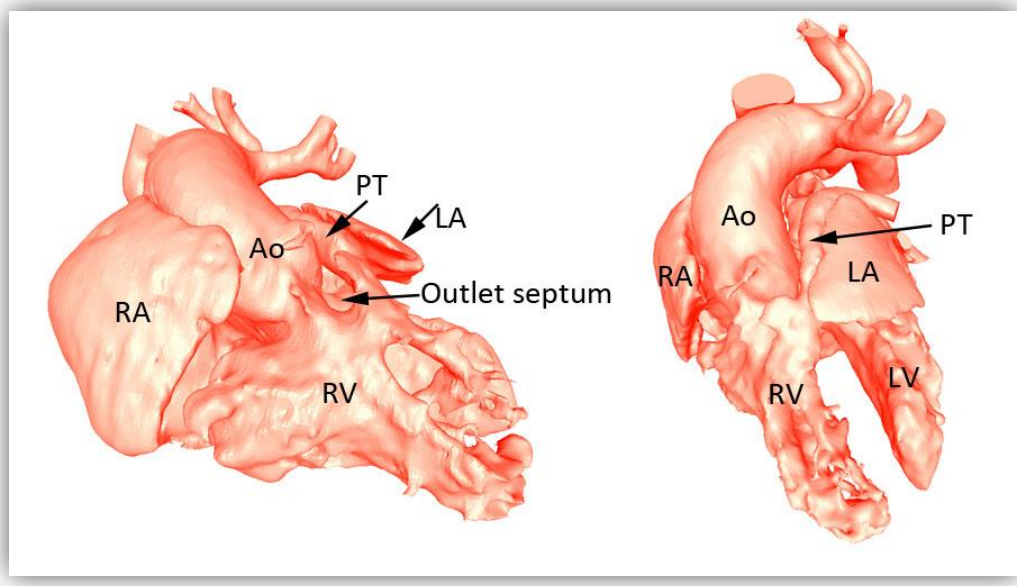

Figure 1-5. Tetralogy of Fallot with DORV seen in volume rendered contrast-enhanced MR angiograms. Long-axial oblique view (right panel) shows that the aorta arises exclusively from the right ventricle. The subpulmonary right ventricular outflow tract shown in frontal view (left panel) demonstrates typical morphology of tetralogy of Fallot with anterior, leftward and superior deviation of the outlet septum.

Double outlet right ventricle and transposition

Transposition of the great arteries (TGA) is another form of ventriculoarterial connection in which the pulmonary arterial trunk arises from the morphologically left ventricle and the aorta arises from the morphologically right ventricle, i.e., discordant ventriculoarterial connection. There is a spectrum between the unequivocal forms of TGA and DORV, showing varying degrees of overriding of the pulmonary valve or both pulmonary and aortic valves across the VSD (**Figure 1-6**). As both TGA and DORV represent the specific forms of ventriculoarterial connection, they are mutually exclusive terms based on the '50% rule' (**Figure 1-4**).

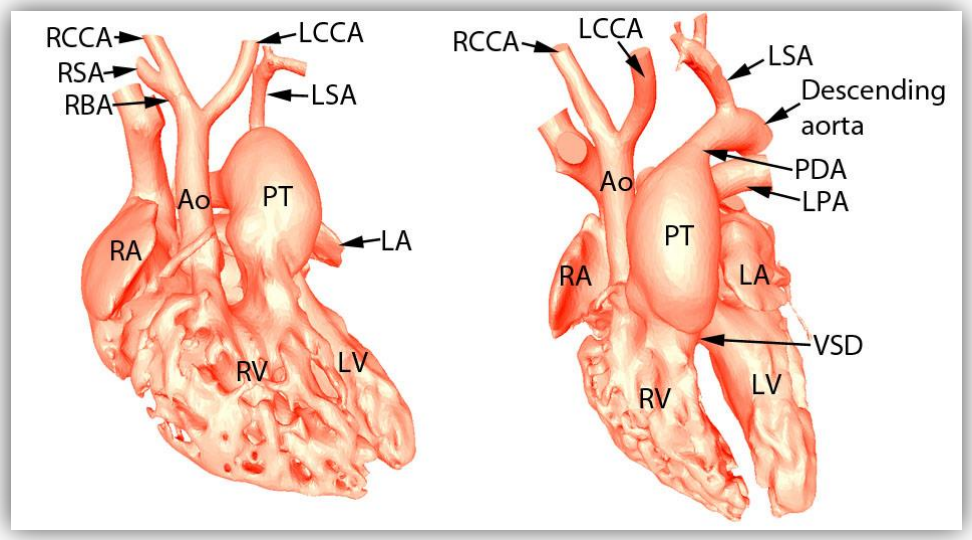

Figure 1-6. DORV with a subpulmonary ventricular septal defect, so-called Taussig-Bing malformation, seen in volume rendered contrast-enhanced CT angiograms. Long-axial oblique view (right panel) shows that pulmonary valve overrides the ventricular septum with approximately 80% of the pulmonary arterial trunk arising from the right ventricle. The subaortic outflow tract and aorta are small and the aortic arch is interrupted after the origin of the left common carotid artery (LCCA). LSA, left subclavian artery; RBA, right brachiocephalic artery; RCCA, right common carotid artery; RSA, right subclavian artery.

Taussig-Bing Malformation

In 1949, Taussig and Bing reported a case with the aorta arising entirely from the right ventricle and the pulmonary orifice overlying the ventricular septum through a VSD [6]. They described that a muscular ridge extended from the upper margin of the ventricular septum close to the defect: forward to the outer wall of the right ventricle. They went on to describe that this ridge separated the aorta from the pulmonary artery. It is apparent that what they named as a muscular ridge is an identical structure to

what we now call outlet or infundibular septum. When Richard van Praagh re-examined the heart reported by Taussig and Bing, he found that the heart was characterized by a bilateral conus, hence pulmonary–mitral discontinuity, and viewed it as one form of DORV since the pulmonary valve did not override the left ventricular cavity [35]. Hinkes et al subsequently revisited the same heart again by coating the endocardium with barium sulfate and marking the valve rings with wires, and taking radiographs in various projections [36]. They showed that the pulmonary valve did override the ventricular septum and also connected directly to the left ventricle as it was originally described by Taussig and Bing. They also described that the semilunar valve rings appear side-by-side. Based on the descriptions in these historic papers, some reserve the term "Taussig-Bing malformation" for the DORV cases that have a subpulmonary VSD, overriding pulmonary valve, bilateral infundibulum and side-by-side relationship [35, 37]. More commonly, the term is used for all hearts with the aorta arising from the right ventricle and the pulmonary arterial trunk overriding the underlying VSD to have a biventricular origin [20, 38-41]. With this extended use of the term "Taussig-Bing heart", Lev et al classified the spectrum of the hearts into right-sided, intermediate and left-sided forms according to the degree of commitment of the overriding pulmonary valve to the right and left ventricles (**Figure 1-7**) [38]. If the "50% rule" [20] is applied for the definition of DORV with an overriding arterial valve, the Lev et al's left-sided and intermediate types of Taussig-Bing heart are not DORV but transposition. Such variations in usage of the term "Taussig-Bing malformation" have resulted in some inconsistency in statistics regarding its overall prevalence, the incidences of associated malformations and the surgical results. Although it is not ideal for the term to be defined in such different ways, the variability in the use of the term "Taussig-Bing malformation" is unavoidable and will continue since it is named after the authors of a single case report [20].

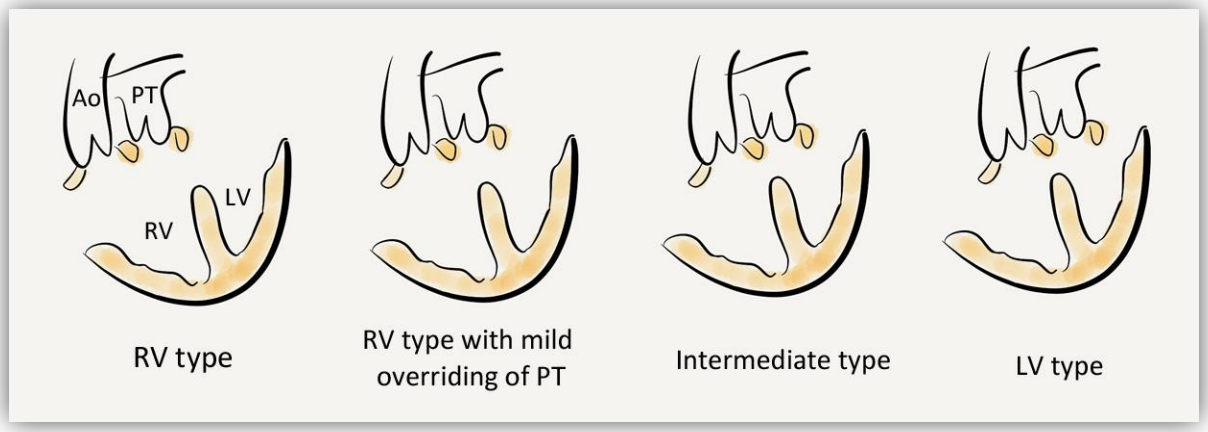

Figure 1-7. Classification of Taussig-Bing malformation by Lev et al [37, 38].

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CHAPTER 2. CLASSIFICATION AND MORPHOLOGCIAL OVERVIEW

Double outlet right ventricle (DORV) may occur in combination with any type of situs and any form of atrioventriculoarterial connection as shown in **Table 2-1**. In this chapter, we will focus our discussion on cases occurring in situs solitus with concordant atrioventricular connection.

Table 2-1. Possible segmental combinations of situs and atrioventricular connections in DORV.

Situs solitus
- Concordant atrioventricular connection
- Discordant atrioventricular connection
- Univentricular atrioventricular connection
Situs inversus
- Concordant atrioventricular connection
- Discordant atrioventricular connection
- Univentricular atrioventricular connection
Right isomerism
- Biventricular atrioventricular connection
- Univentricular atrioventricular connection
Left isomerism
- Biventricular atrioventricular connection
- Univentricular atrioventricular connection
Undetermined situs
- Biventricular atrioventricular connection
- Univentricular atrioventricular connection

Classic Morphological Classification of DORV with Situs Solitus and Concordant Atrioventricular Connection

The hemodyamic physiology and the clinical manifestations of DORV are dependent on two major factors: the relation of the VSD to the arterial valves and the presence or absence of aortic or pulmonary outflow tract obstruction, although other factors such as the size of the VSD and abnormalities of the atrioventricular valves are also important. Traditionally, the relation of the VSD to the arterial valves is classified into subaortic, subpulmonary, doubly committed and non-committed or remote varieties after the seminal publications by Lev, et al in 1972 (**Figure 2-1**) [1]. These terms do not imply that the VSD move around within the ventricular septum [2]. To the contrary, the relationship between the defect and the arterial valves is determined by [3-5]:

1) the location of the defect in the ventricular septum,
2) the orientation of the outlet (infundibular) septum in relation to the defect margin,
3) the spatial relationship between the arterial valves, and
4) the extent of the muscular infundibulum

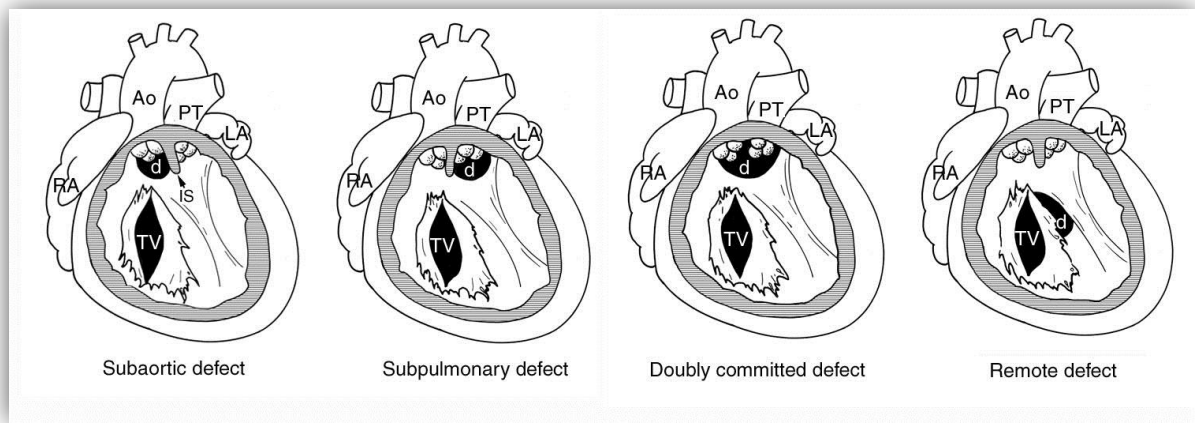

Figure 2-1. Types of VSD according to the relationship between the defect and the arterial valves (after Lev, et al [1]). Only the examples seen in hearts with situs solitus, concordant atrioventricular connection and right-sided aorta are illustrated.

In categorizing the VSD into such varieties, one should be aware that there are grey zones among the classes (**Figure 2-2**). For instance, the defects that one may call a double committed VSD may have more commitment to either arterial valve, so one may be inclined to classify them into either the subaortic or subpulmonary variety. On the other hand, the remoteness of the VSD from the arterial valves is an arbitrary definition. Therefore, there are grey zones between the non-committed variety and other varieties.

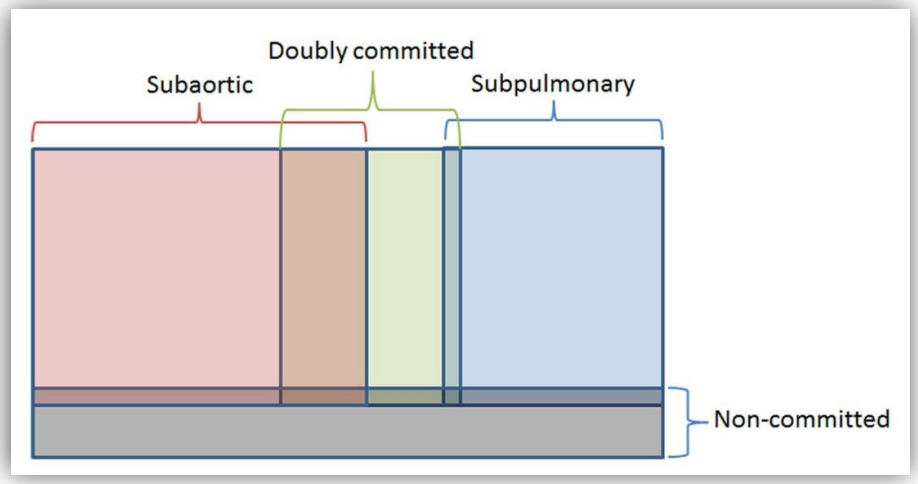

Figure 2-2. Overlapping nature of the types of ventricular septal defect with grey zones among the classes.

Relative incidences of these four types of DORV from the recent large surgical series [6-10] are compiled in **Table 2-2**. DORV with a subaortic VSD accounts for almost 50% of the cases, while DORV with a subpulmonary VSD accounts for approximately 25% of the cases. In contrast to most pathological series [1, 11, 12] where DORV with a non-committed VSD is least common, the surgical series invariably showed a higher incidence of the non-committed variety than the doubly committed variety. In surgical series, DORV with a non-committed VSD and DORV with a doubly committed VSD account for approximately 20% and 5%, respectively. Multiple VSDs are seen in approximately 6-12% of the surgical series.

Table 2-2**.** Site of VSD in relation to arterial valves in DORV with situs solitus and concordant atrioventricular connection in contemporary surgical series [6-10].

Authors	Aoki, et al [6]	Kleinert, et al [7]	Belli, et al [8]	Brown, et al [9]	Bradley, et al [10]	Compiled
Institution	Boston	Melbourne	Le Plessis-Robinson	Indiana	Toronto	
Year of publication	1994	1997	1996	2001	2007	
Years included	1981-1991	1978-1993	1980-1995	1980-2000	1980-2000	
Total cases	73	193	180	124	335	905
Subaortic	31(42%)	90 (47%)	106 (59%)	57 (46%)	156 (47%)	440 (49%)
Subpulmonary	27 (37%)	49 (25%)	37 (21%)	39 (31%)	76 (23%)	228 (25%)
Doubly committed	5 (7%)	5 (3%)	17 (9%)	6 (5%)	15 (4%)	48 (5%)
Non-committed	10 (14%)	49 (25%)	20 (11%)	22 (18%)	88 (26%)	189 (21%)

The location of the defect in the ventricular septum in DORV is described using a few important anatomical landmarks that include trabecula septomarginalis, ventriculoinfundibular fold, outlet septum (infundibular or conal septum) and membranous septum (**Figure 2-3**) [13-15]. The trabecula septomarginalis is also called the septal band of the crista supraventiricularis [13-17]. The trabecula septomarginalis is a 'Y'-shaped muscular elevation on the right ventricular aspect of the septum. The body of the 'Y' extends from the superior or inner curvature of the right ventricle to the apex of the right ventricle. The anterior limb of the 'Y' extends forward to support the left-sided arterial valve. The posterior limb extends backward to fuse to the posterior part of the ventriculoinfundibular fold of the right ventricle above the commissure between the septal and anterior leaflets of the tricuspid valve.

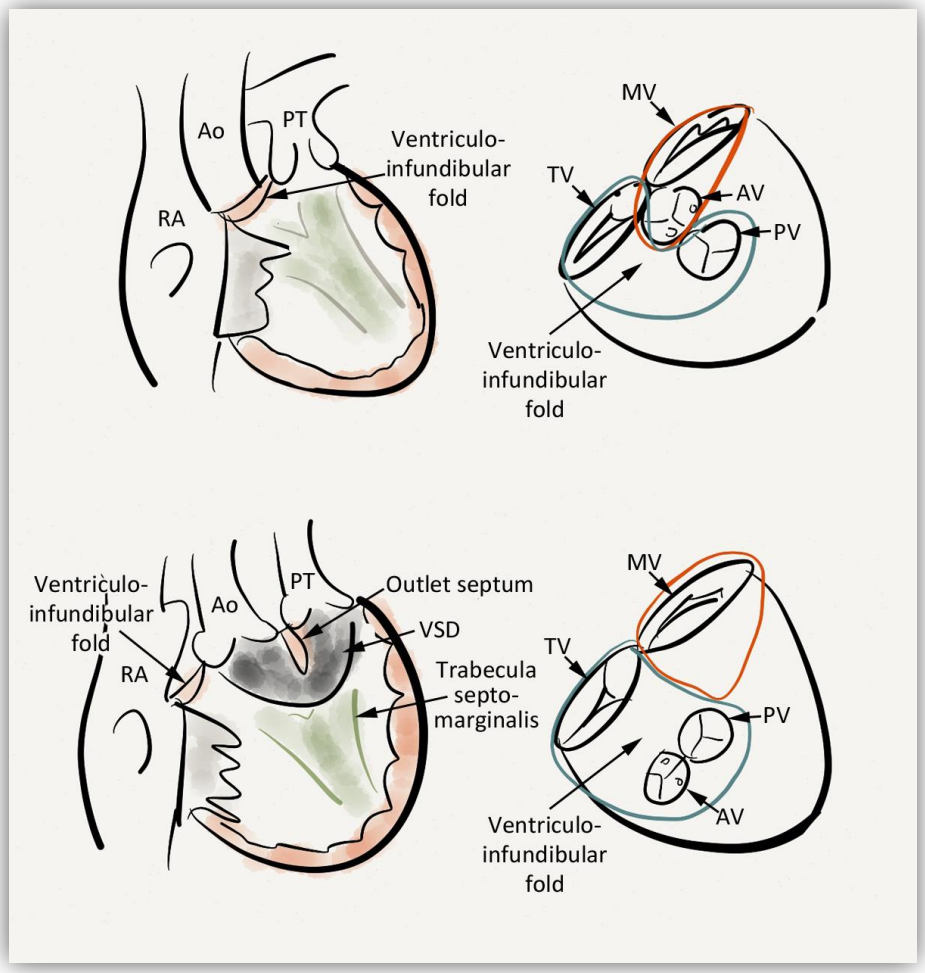

Figure 2-3. Cartoons illustrating the anatomical terms used in description of VSD and ventricular outflow tract abnormalities. The upper panels show the anatomy in a normal heart and the lower panels show the anatomy in an example of DORV. In normal heart, there is a muscular crest, called crista supraventricularis, between the pulmonary and tricuspid valves. The parietal part of this crest is the right ventriculoinfundibular fold. The aortic valve is in fibrous continuity with the mitral and tricuspid valves and, therefore, there is no left ventriculoinfundibular fold. The trabecula septomarginalis is a Y-shaped muscular elevation on the right ventricular aspect of the ventricular septum. A small triangular part between the anterior and posterior limbs of the 'Y' is the septal part separating the right and left ventricular outflow tracts. It is called the outlet or infundibular septum. In DORV, the outlet septum is exclusively a right ventricular structure. In so-called classic form of DORV with bilateral infundibulum, both aortic and pulmonary valves are separated from the mitral and tricuspid valves by the intervening muscular fold. The fold between the semilunar valves and the tricuspid valve is called the right ventriculoinfundibular fold and the fold between the semilunar valves and the mitral valve is called the left ventriculoinfundibular fold.

The trabecula septomarginalis typically gives rise to the moderator band from its apical aspect. The ventriculoinfundibular fold is the parietal muscular structure interposing between the atrioventricular and arterial valves [13-15]. The outlet septum is the septum that separates the aortic and pulmonary outflow tracts below the semilunar valves. In the normal heart, the outlet septum occupies a small triangular area above the anterior and posterior limbs of the trabecula septomarginalis. In hearts with DORV, the outlet septum, although it can be vestigial, is exclusively a right ventricular structure. The membranous septum abuts on the commissure between the septal and anterior leaflets of the tricuspid valve and extends toward the inlet part of the right ventricle along the septal leaflet.

Most commonly, the VSD in DORV primarily involves the outlet component of the septum that is cradled between the anterior and posterior limbs of the trabecula septomarginalis. Less frequently, the defect involves the inlet part of the septum across or below and behind the posterior limb and body of the trabecula septomarginalis or the trabecular part of the septum. The interventricular communication can be through an atrioventricular septal defect. Occasionally, DORV is associated with multiple VSD's. Very rarely, DORV occurs with an intact ventricular septum [14, 18-27].

VSD involving the Outlet Part of the Ventricular Septum

Typically, the VSD involves the outlet septum and extends to the adjacent limbs of the trabecula septomarginalis. Therefore, the VSD appears cradled between the limbs of the attenuated trabecula septomarginalis (**Figure 2-4**) [3, 4, 14, 15]. When the posterior limb is not completely attenuated and fused to the ventriculoinfundibular fold, it separates the VSD from the membranous part of the septum (**Figure 2-4, left panel**). When the VSD involves the posterior limb, it reaches the membranous part of the ventricular septum around the anteroseptal commissure of the tricuspid valve (**Figure 2-4, right panel**).

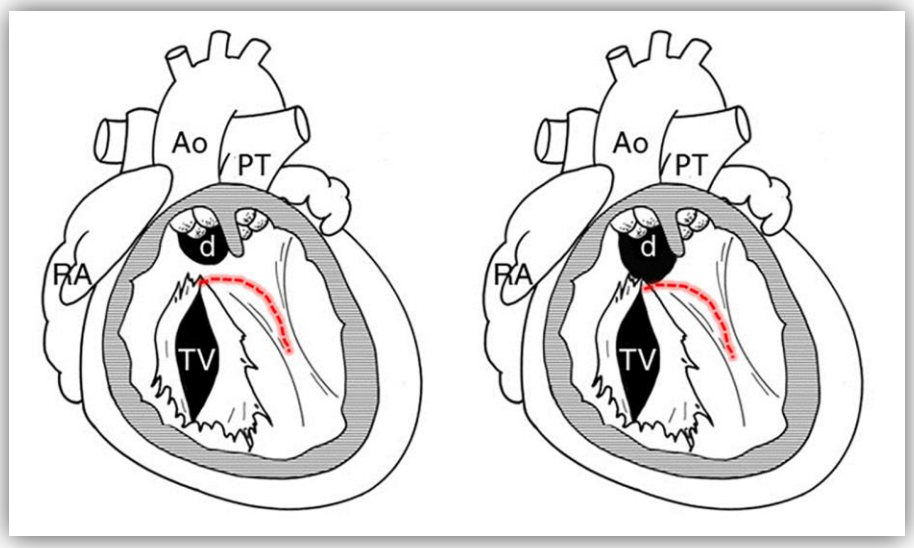

Figure 2-4. Legend in the next page.

Figure 2-4. Cartoons showing DORV with the VSD (d) cradled between the limbs of the trabecula septomarginalis. In left panel, the posterior limb of the trabecula fuses to the ventriculoinfundibular fold, separating the defect from the membranous septum. In right panel, the defect extends posteriorly and inferiorly to involve the posterior limb of the trabecula so that the latter structure does not connect to the ventriculoinfundibular fold. As the defect is in direct contact with the membranous septum, it is a perimembranous defect. When the VSD is a perimembranous type, the atrioventricular conduction axis (red dotted line) courses along the posteroinferior margin of the VSD (right panel). When the VSD does not involve the membranous septum, the conduction axis courses along the posteroinferior aspect of the intact membranous septum and is therefore away from the VSD margin (left panel).

As discussed previously, the commitment of the VSD to an arterial valve or arterial valves depends not only on the locations of the VSD and orientation of the outlet septum but also on the relationship of the arterial valves. The following discussion will focus on the common forms of DORV in the setting of situs solitus and atrioventricular concordant connection with the aorta on the right side of the pulmonary arterial trunk. In most cases of DORV that have a VSD cradled between the two limbs of trabecula septomarginalis, the outlet septum separates the pulmonary and aortic outflow tracts. The orientation of the outlet septum relative to the margins of the VSD determines the commitment of the defect to an arterial valve or both valves [3, 5, 28-32]. Four variations can be seen in this regard (**Figure 2-5**):

- ♥ The outlet septum inserts to the left margin of the VSD, committing the VSD to the right-side arterial valve (**Figure 2-6**).
- ♥ The outlet septum inserts to the right margin of the VSD, committing the VSD to the left-side arterial valve (**Figure 2-7**).
- ♥ The outlet septum is parallel to the plane of the VSD, committing the VSD to the posterior arterial valve (**Figure 2-8**).
- ♥ The outlet septum is absent or only vestigial, committing the VSD to either or both valves (**Figure**s 2-9 and 2-10).

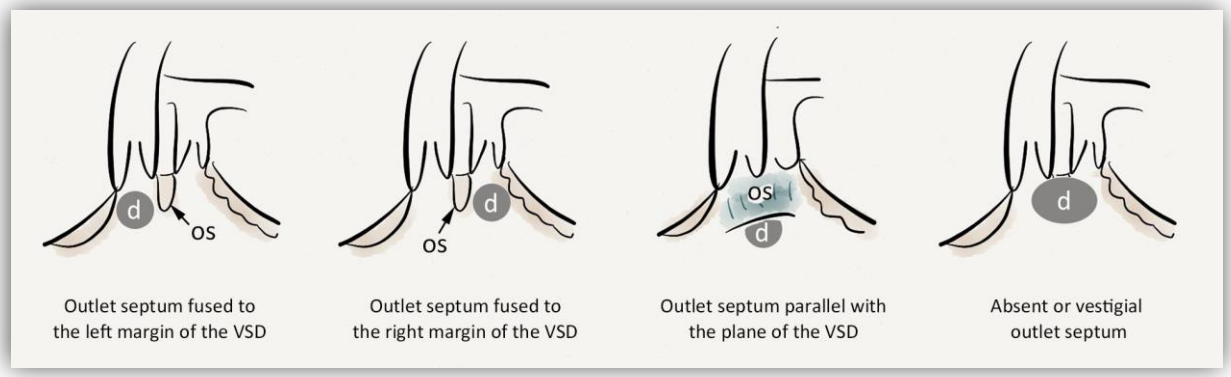

Figure 2-5. Cartoons showing 4 variations of the orientation of the outlet septum (OS) relative to the VSD (d) margin in the VSD involving the outlet part of the ventricular septum.

Outlet septum inserting to the left margin of the VSD: In this variety, the outlet septum is more or less perpendicularly oriented to the ventricular septum harboring the VSD. The outlet septum appears deviated leftward and forward from the area where it should be in the normal heart (**Figure 2-6**). The outlet septum deviates like a trap door with hinges. The door of the outlet septum opens into the right ventricle around its hinges on the left margin of the VSD, which is the attenuated anterior limb of the trabecula septomarginalis. The arterial trunk behind the outlet septum is rotated and displaced into the right ventricle along with the outlet septum. This configuration is the classic feature of DORV with a subaortic VSD in the setting of situs solitus and concordant atrioventricular connection with a right-sided aorta (**Figure** 2-6). With the pulmonary valve being isolated from the VSD by the outlet septum, the pulmonary outflow tract may be widely patent but is prone to be narrowed or completely occluded according to the severity of deviation of the outlet septum. The arterial trunks are usually normally related with the aortic valve located rightward and slightly posterior to the pulmonary arterial trunk.

Outlet septum inserting to the right margin of the VSD: In this variety, the outlet septum is also more or less perpendicularly oriented to the ventricular septum harboring the VSD. The outlet septum appears deviated rightward and forward from the area where it should be in the heart with complete transposition of the great arteries (**Figure** 2-7). The outlet septum deviates like a trap door with hinges. The door of the outlet septum opens into the right ventricle around its hinges on the right margin of the VSD, which is the attenuated posterior limb of the trabecula septomarginalis or the posterior part of the ventriculoinfundibular fold. The arterial trunk behind the outlet septum is rotated and displaced into the right ventricle along with the outlet septum. Such deviation of the outlet septum may cause significant encroachment on the dimension of the outflow tract that leads to the right-sided arterial valve. This configuration is the classic feature of DORV with a subpulmonary VSD in the setting of situs solitus and concordant atrioventricular connection with a right-sided aorta (**Figure 2-7**). The arterial trunks usually show a side-by-side relationship with the aorta on the right.

Outlet septum parallel to the plane of the VSD: In this variety, the outlet septum appears moved forward from the area where it should be in the heart with complete transposition of the great arteries (**Figure 2-8**). The arterial trunks and the outlet septum are moved forward en bloc so that the outlet septum is parallel to the plane of the VSD and inserts to the more anterior part of the ventriculoinfundibular fold. Only the arterial trunk behind the outlet septum is committed to the VSD. The forward displacement of the outlet septum may cause significant encroachment on the dimension of the outflow tract that leads to the anterior arterial valve. This configuration is also classically seen in DORV with a subpulmonary VSD in the setting of situs solitus and concordant atrioventricular connection (**Figure 2-8**). The arterial trunks usually show an anteroposterior relationship with the aorta anteriorly and slightly to the right relative to the pulmonary arterial trunk.

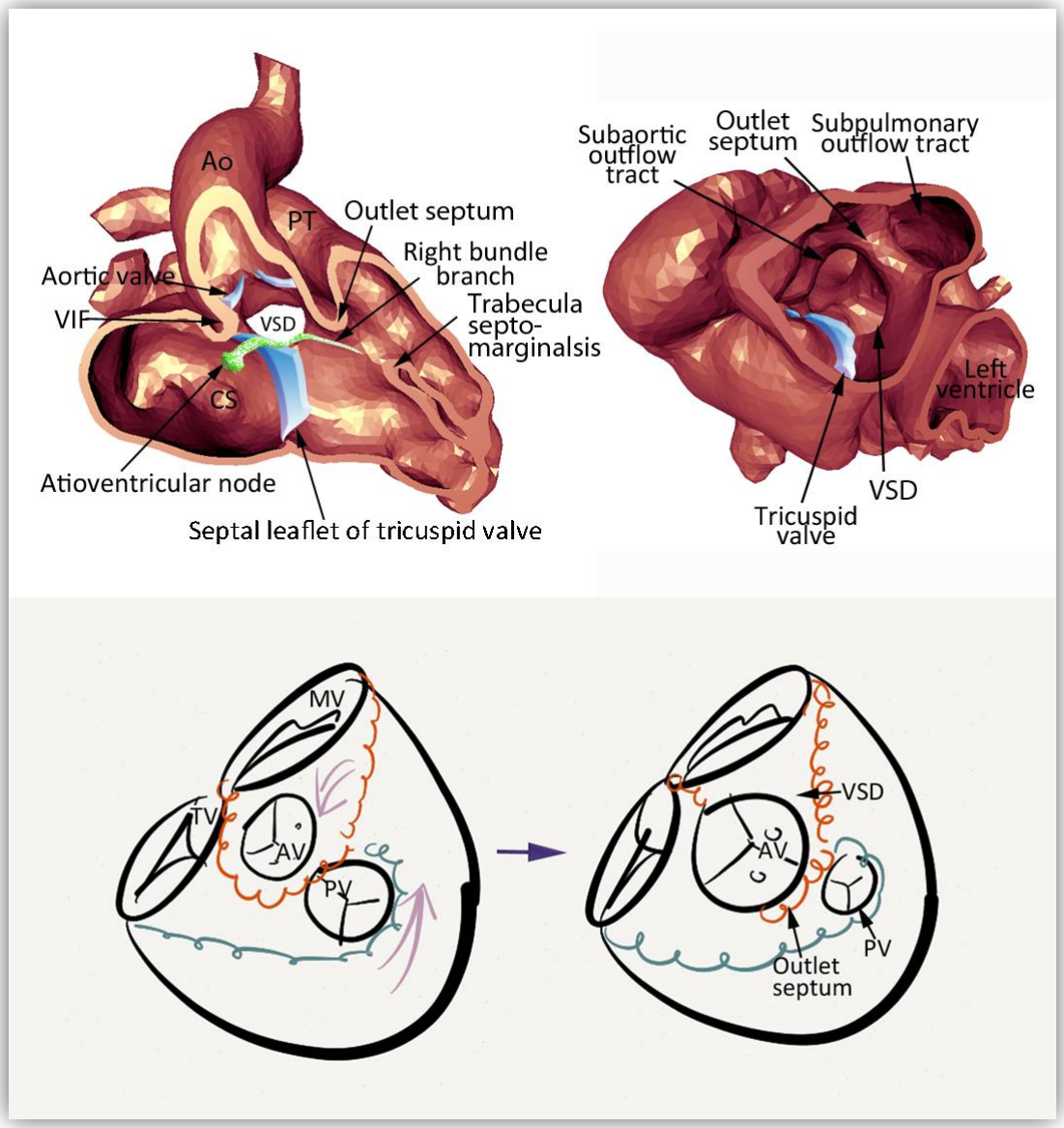

Figure 2-6. DORV with a subaortic VSD in the setting of situs solitus, atrioventricular concordant connection and right-sided aorta. The outlet septum is inserted to the left margin of the VSD, isolating the pulmonary outflow tract from the VSD. Cartoons in the lower panel show the direction of rotation of the arterial trunks and the outlet septum in DORV with a subaortic VSD. The aortic valve of the normal heart appears pushed forward and rotated counter-clockwise seeing from above as indicated by the purple arrows in the left-hand cartoon so that the aortic valve and outlet septum are rotated and displaced into the right ventricle. The hinge plane of the deviation is on the left margin of the VSD.

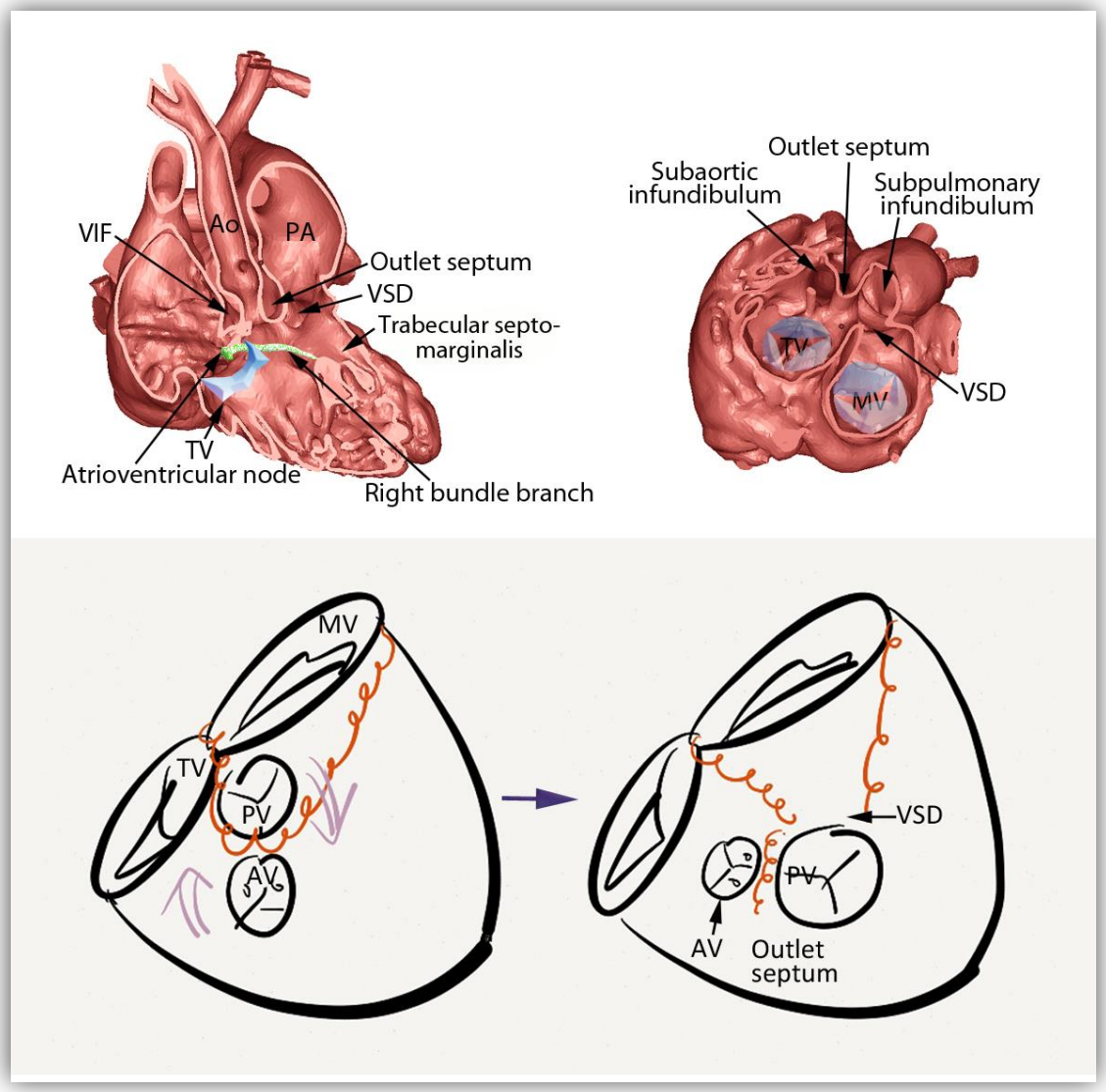

Figure 2-7. DORV with a subpulmonary VSD in the setting of situs solitus, atrioventricular concordant connection and right-sided aorta. The outlet septum is inserted to the right margin of the VSD, isolating the subaortic outflow tract from the VSD. Cartoons in the lower panel show the direction of rotation of the arterial trunks and the outlet septum in DORV with a subpulmonary VSD. The pulmonary valve of the heart with complete transposition of the great arteries appears pushed forward and rotated clockwise seeing from above as indicated by the purple arrows in the left-hand cartoon so that the pulmonary valve and outlet septum are rotated and displaced into the right ventricle. The hinge plane of the deviation is on the right margin of the VSD.

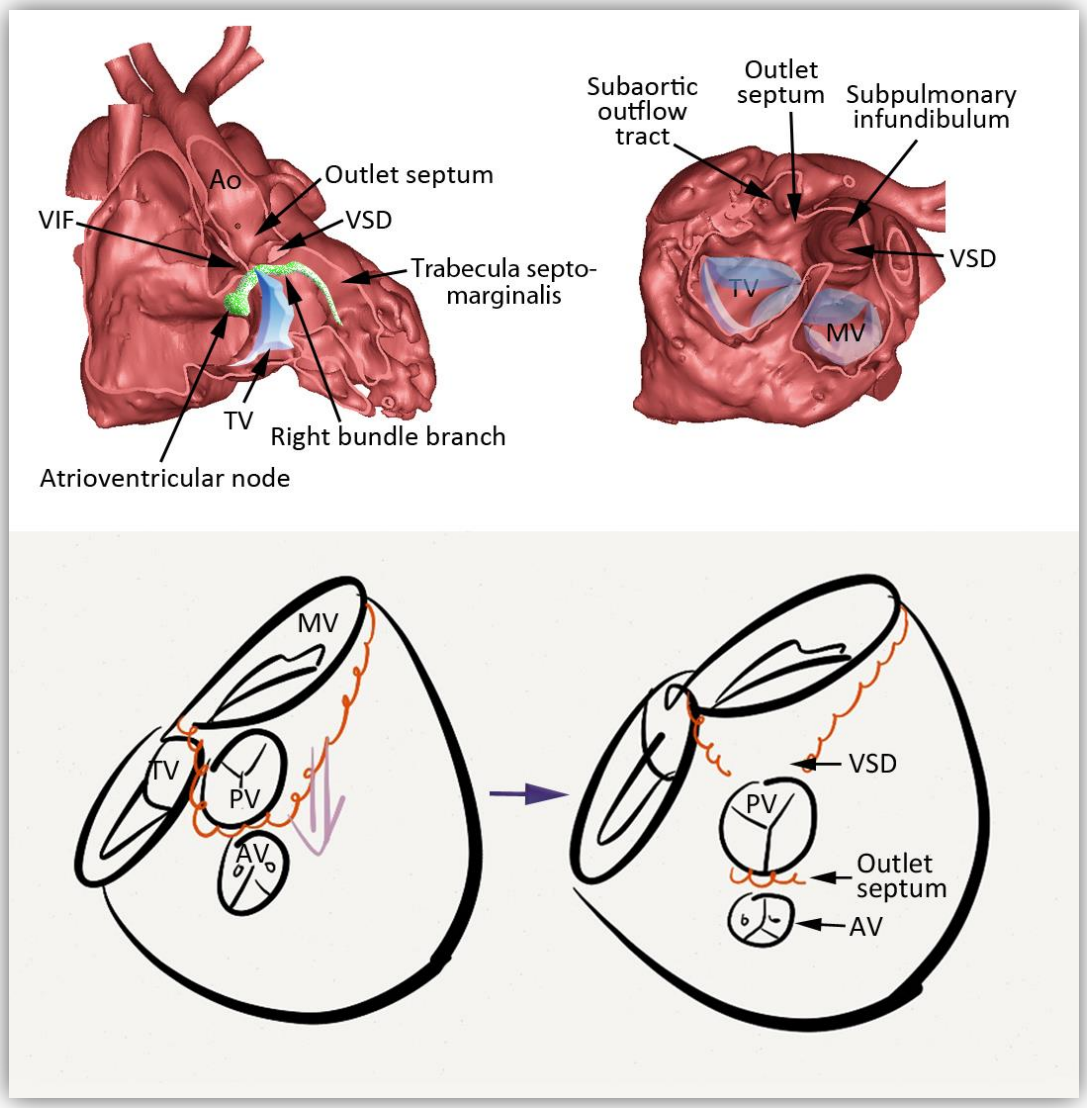

Figure 2-8. DORV with a subpulmonary VSD in the setting of situs solitus, atrioventricular concordant connection, and anterior and right-sided aorta. The outlet septum is parallel to the plane of the VSD with the posterior arterial trunk solely committed to the VSD. Cartoons in the lower panel show the direction of displacement of the arterial trunks and the outlet septum in this variety of DORV. The arterial trunks and the outlet septum of the heart with complete transposition of the great arteries appears pushed forward as indicated by the purple arrow in the left-hand cartoon so that the pulmonary valve and outlet septum are moved into the right ventricle. Note that the outlet septum does not insert to the margin of the VSD but is parallel with the ventricular septum harboring the VSD. VIF, ventriculoinfundibular fold.

Absent or vestigial outlet septum: Uncommonly, the outlet septum is absent or only vestigial. This variety is typically described in DORV with a VSD committed to both arterial valves, i.e., doubly committed (**Figure 2-9**) [3, 4, 14, 32]. The raphe or vestigial outlet septum between the arterial valves inserts to the superior margin of the VSD. However, it should be noticed that an absent or vestigial outlet septum does not necessarily mean that the VSD is doubly committed [5, 15]. The VSD can also be in either the subaortic or subpulmonary location with the raphe or vestigial outlet septum inserted to either side, instead of the superior margin, of the VSD (**Figure 2-10**) [5, 15]. On the contrary, the VSD can be doubly committed in the presence of the sizable outlet septum [15]. The raphe or hypoplastic outlet septum may be deviated toward the aortic or pulmonary valve, producing one arterial valve smaller than the other with or without pulmonary or aortic valve stenosis.

In summary, the VSD involving the outlet part of the ventricular septum seen from the right ventricular aspect is committed to the right-sided arterial valve (usually the aortic valve) when the outlet septum or its vestige inserts to the left margin of the VSD (**Figure 2-6** and left panel of **Figure 2-10**). On the other hand, the VSD involving the outlet part of the septum is committed to the left-sided arterial valve (usually the pulmonary valve) when the outlet septum or its vestige inserts to the right margin of the VSD (**Figure 2-7** and right panel of **Figure 2-10**). The VSD involving the outlet part of the septum is committed to the posteriorly located arterial valve (usually the pulmonary valve) when the outlet septum is parallel to the rest of the septum within the right ventricle (**Figure 2-8**). The situations with the VSD committed to the left-sided or posteriorly located pulmonary valve are the features of so-called Taussig-Bing malformation [33-35]. The displacement or deviation of the outlet septum often encroaches on the dimension of the outflow tract toward which the outlet septum is displaced or deviated. Narrowing of the subpulmonary outflow tract is common in DORV with a subaortic defect, whereas the subaortic outflow tract is often small or overtly narrow in DORV with a subpulmonary defect [6, 7, 36-38].

Although the general principles described above apply in most cases, it is important to note that the commitment of an arterial valve or valves to the margin of the VSD is at a variable distance according to the extent of the muscular infundibulum (**Figure 2-11**) [4, 5]. When the muscular infundibulum is not well formed, the committed arterial valve is in direct contact with or at close proximity to the defect margin. When the arterial valve is supported by an excessively long infundibulum, there is a considerable distance between the arterial valve and the VSD margin. As a result, this type of defect is classified as a non-committed VSD by some (**Figure 2-2**) [4, 39, 40].

When the aortic valve is on the left side (levo- or L-malposed) in the setting of situs solitus and concordant atrioventricular connection, the VSD is usually cradled between the limbs of the trabecula septomarginalis and is committed to either arterial valve [2]. More commonly, the VSD lies more anteriorly and superiorly than it does when the aorta is on the right side (dextro- or D-malposed) and is committed to the aorta.

The presence of continuity or fusion between the posterior limb of the trabecula septomarginalis and the ventriculoinfundibular fold has an important significance on the location of the atrioventricular conduction axis relative to the margin of the VSD (**Figure 2-4**) [3, 4]. When they are not continuous or fused, the defect involves the membranous septum and the conduction axis courses on the

posteroinferior margin of the defect, and therefore is vulnerable to injury at the time of surgical closure of the VSD. When they are continuous or fused, the defect does not involve the membranous septum and the muscle bar separates the conduction axis from the defect margin and is less vulnerable to injury at surgery.

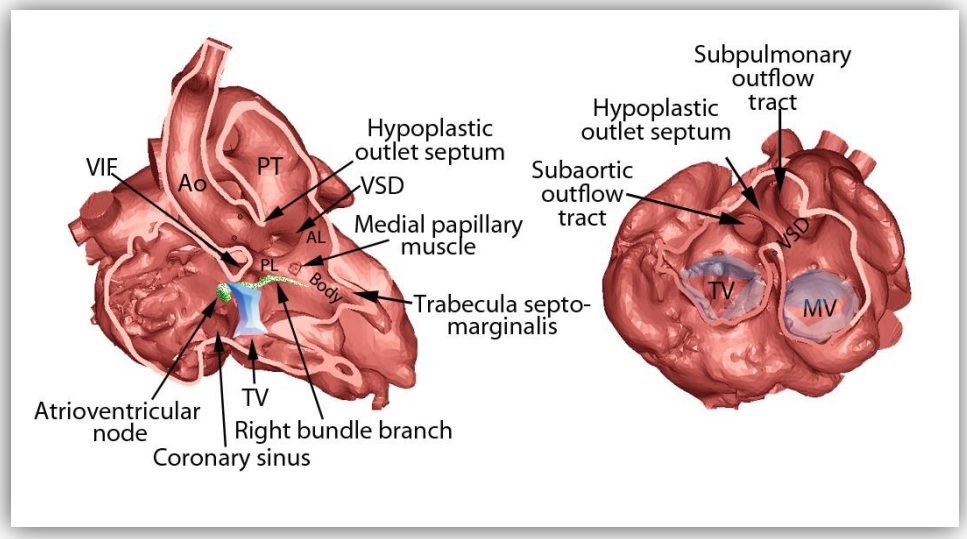

Figure 2-9. DORV with a doubly-committed VSD. A large VSD in the outlet part of the right ventricle is below both arterial valves. The hypoplastic outlet septum is seen in the roof of the VSD.

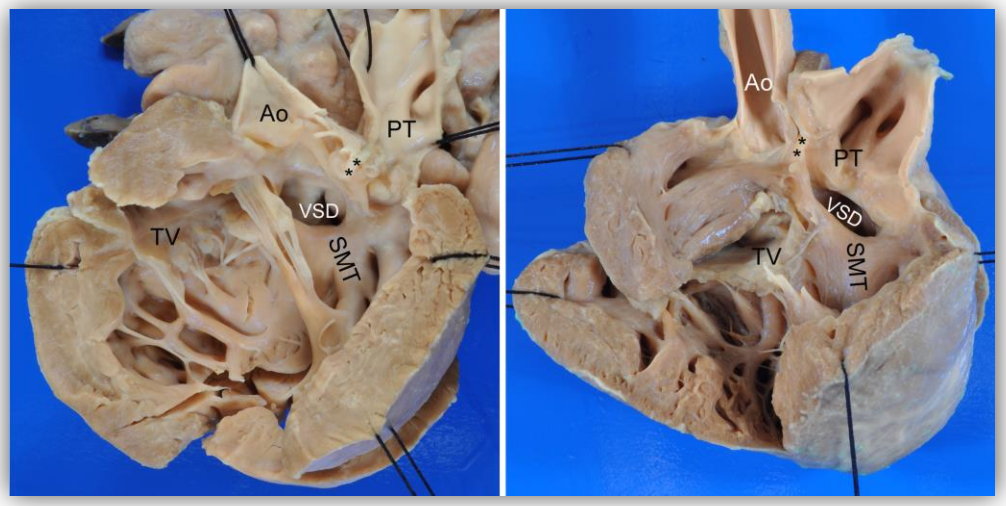

Figure 2-10. Two different DORV cases with a VSD cradled between the limbs of the trabecula septomarginalis and an absent outlet septum. In both **Figure**s, the semilunar valves are in direct contact (asterisks) because of the absence of the outlet septum. The left panel shows the defect solely committed to the right-sided aortic valve. The right panel shows the defect solely committed to the left-sided pulmonary valve.

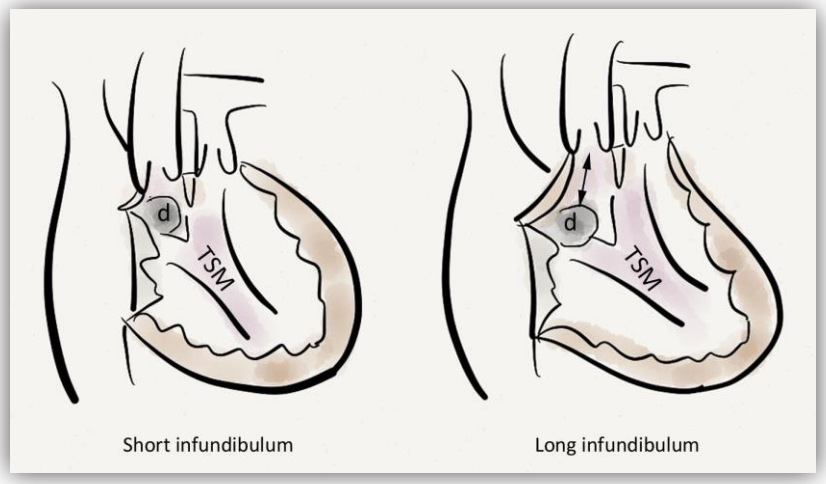

Figure 2-11. Extent of infundibulum versus distance between semilunar valve and VSD margin. Two cartoons show the location of the VSD similar. Left hand panel shows the VSD closely committed to the aortic valve. Right hand panel shows the VSD at a distance (double-headed arrow) from the aortic valve.

VSD involving the Inlet or Trabecular Part of the Ventricular Septum

Infrequently, the VSD involves the inlet component of the ventricular septum behind and below the posterior limb of the trabecula septomarginalis or involves the posterior limb and the body of the trabecula (**Figure 2-1**, right panel and **Figure 2-12**) [39-42]. Such defects are usually perimembranous. Rarely, a muscular defect in the inlet or trabecular part of the ventricular septum can be seen [39]. The defects involving the inlet and/or trabecular part are regarded as non-committed [1] or remote [11]. As it is an arbitrary distinction between the committed and non-committed VSDs in borderline cases, it was suggested that the VSD should be defined as non-committed when the distance between the VSD and both aortic and pulmonary valve is greater than the age-matched diameter of the aortic valve [43-45]. Although this definition appears clear, it should be emphasized, as discussed earlier, that the arterial valve and the upper margin of the otherwise "committed" VSD can be separated by a considerable distance if there is an excessively long infundibulum (**Figure 2-11**) [4, 5, 39, 40, 43]. In addition, it should be taken into account that the VSDs that are committed to an arterial valve by anatomical definition may be rendered surgically non-committed because of the factors that do not allow baffling the VSD to an arterial valve [39].

Although such defects are not directly committed to either arterial valve, there are remote defects that are closer to either arterial valve, and therefore, the blood flow shunting from the left ventricle can surgically be directed toward one arterial valve [39, 40]. Recently there has been increasing interest in creating a long intraventricular baffle that connects the left ventricle to either arterial valve. Therefore, the remote defects should be subclassified into those that are able to be surgically baffled to either aortic or pulmonary valve and those that cannot be baffled to either arterial valve.

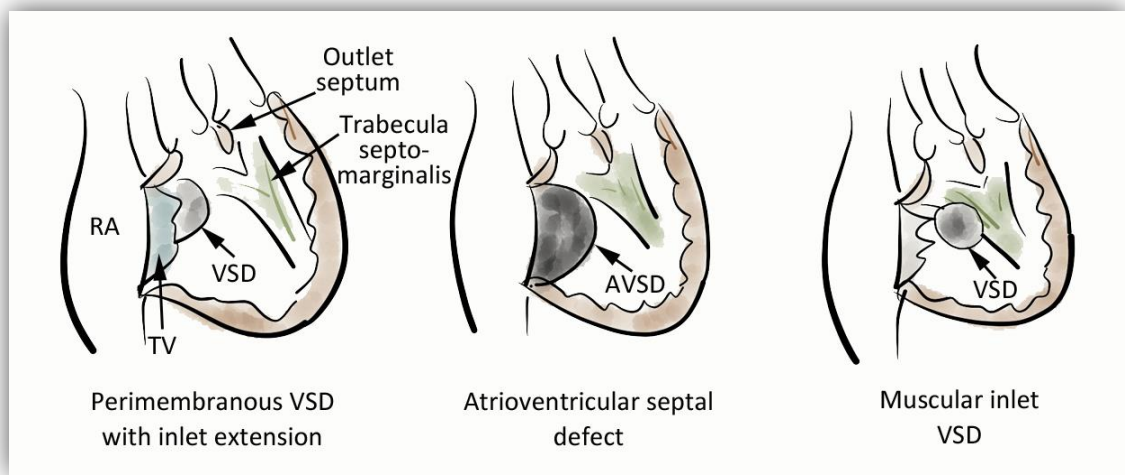

Figure 2-12. DORV with a remote or non-committed VSD. The outlet septum is a free standing structure in the right ventricular outlet. It is not directly related to the VSD margin.

DORV can also be associated with an atrioventricular septal defect in the presence of either a common or partitioned atrioventricular orifice (**Figure 2-12**, middle panel). The combination of DORV and atrioventricular septal defect is commonly seen in the setting of atrial isomerism, particularly the right isomerism [5, 46]. When the defect is not in close proximity to either arterial valve, this uncommon variant is also classified as DORV with a non-committed or remote VSD (**Figure 2-13**, left panel). However, the atrioventricular septal defect often extends toward the outlet so as that the upper margin of the defect is in close proximity to an arterial valve (**Figure 2-13**, right panel). Such a confluent inlet and outlet defect is typically seen in cases of tetralogy of Fallot with an atrioventricular septal defect.

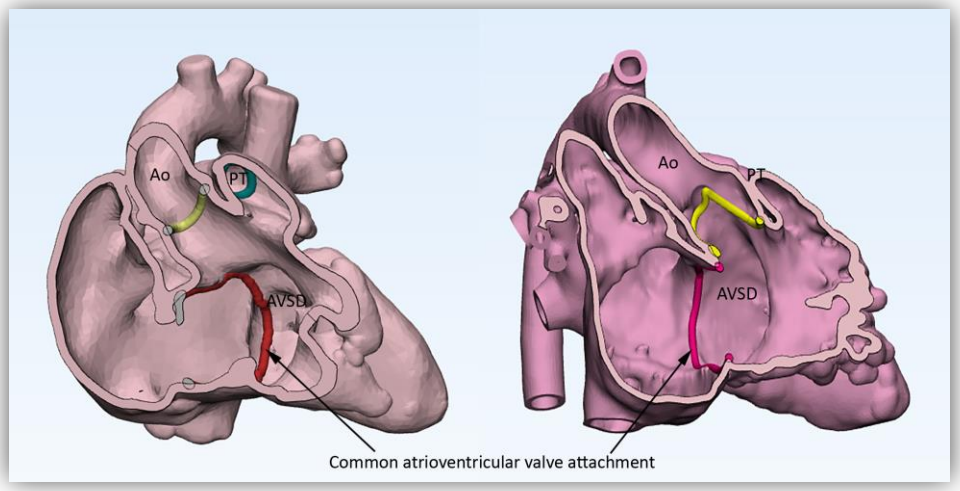

Figure 2-13. Legend in next page

Figure 2-13. DORV with an atrioventricular septal defect (AVSD). Left panel is a case of DORV occurring in heterotaxy with right isomerism. Right panel is a case of DORV occurring in situs solitus. Both cases show a large AVSD and subpulmonary stenosis. In the left panel case, the AVSD does not show significant extension toward the outlet . The aortic valve is located far from the upper margin of the AVSD with a long subaortic infundibulum. The right panel case shows that the AVSD extends toward the outlet to reach the aortic valve annulus.

DORV with restrictive VSD or without VSD

As the VSD is the only outlet of the left ventricle in DORV, its small size implies left ventricular outflow tract obstruction [18, 47-52]. The VSD is considered restrictive when the diameter of the defect is smaller than the age-matched diameter of the aortic valve [53-55]. A restrictive VSD is seen in approximately 10% of DORV cases, most commonly occurring in the variety with a non-committed VSD [44, 56]. The margin of the restrictive VSD is commonly muscular but proliferation of fibrous tissue forming a ridge or diaphragm can further compromise the size of the defect [49-51]. The fibrous tissue may arise from the atrioventricular valve [50] or, less frequently, from the aortic valve [58]. The right ventricular muscle bundles crossing the defect may also restrict the effective size of the VSD. When present, surgical treatment requires enlargement of the VSD and resection of the muscle bundles to establish the unobstructed left ventricular outlet.

Very rarely DORV occurs with an intact ventricular septum [11, 13, 18-27, 56]. This variety is usually associated with severe hypoplasia of the mitral valve and left ventricle as the left ventricle does not have any outlet. The only outlet of the left side of the heart is a patent foramen ovale or an atrial septal defect. Most of the reported cases show findings suggesting spontaneous closure of the restrictive VSD postnatally or during fetal life [18, 21, 57].

Variations in Infundibular Morphology, Great Arterial Relationship, Relationship of the VSD to the Atrioventricular Conduction Axis and Coronary Artery Anatomy

Infundibular Morphology

The ventriculoinfundibular fold, when present, separates the semilunar and atrioventricular valves and therefore completes the muscular infundibulum that supports the semilunar valve. When the ventriculoinfundibular fold separates both semilunar valves from the tricuspid valve, the right ventricular outflow tract is divided into the subaortic and subpulmonary infundibulum. The presence of bilateral infundibulum, which was once considered an essential feature of DORV, is seen in only one quarter or third [14, 58]. (**Figure 2-14**). A bilateral infundibulum is seen in most cases with a non-committed VSD [44, 56], although it is also seen in cases with a subpulmonary or subaortic VSD. Most hearts with unilateral subpulmonary infundibulum have either subaortic or doubly committed VSD, while most with unilateral subaortic infundibulum have subpulmonary VSD.

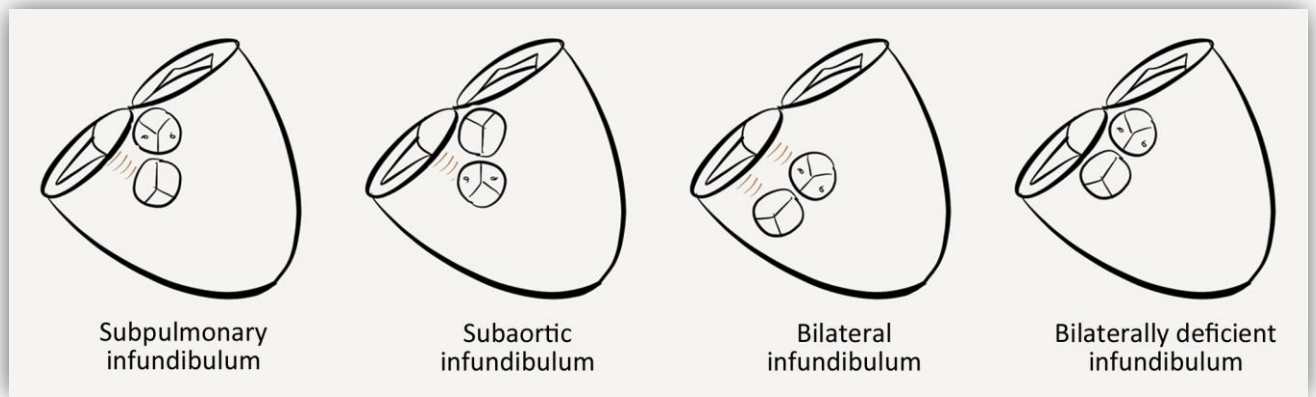

Figure 2-14. Variations of infundibular morphology in DORV.

Great Arterial Relationship

Although the majority of the cases with DORV in the setting of situs solitus and atrioventricular concordant connection have the aorta located on the right side of the pulmonary arterial trunk, there is ample variation of the great arterial relationship (Figure 2-15). With the aorta on the right, the great arterial relationship varies according to the relative position of the semilunar valves. When the right sided aorta has its valve positioned posteriorly relative to the pulmonary valve, the great arteries tend to keep their spiral relationship as in normal hearts (normal relationship). When the aortic valve is positioned more anteriorly and aligned 'side-by-side' with the pulmonary valve in a coronal plane, the great arteries take a parallel course (D-malposition). A further anterior location of the aortic valve results in the great arterial relationship similar to that seen in typical complete TGA; i.e., a parallel course with the aorta rightward and anterior to the pulmonary arterial trunk (D-malposition). Infrequently, the aorta can be directly anterior to the pulmonary arterial trunk (A-malposition). Rarely, the aorta is leftward and anterior to the pulmonary arterial trunk (L-malposition). DORV with L-maplosed great arteries in the setting of situs solitus and concordant atrioventricular connection typically has a subaortic VSD but the defect can infrequently be subpulmonary, doubly committed or non-committed. In general, the more extensive the infundibulum is, the more anterior the semilunar valve is (**Figure 2-16**).

Although a great arterial relationship is not precisely predictive of a VSD location, there is a certain degree of association between the two variables. The normally related great arteries are most commonly seen with a subaortic VSD. A D-malposed side-by-side relationship is usually associated with a subpulmonary VSD, while an L-malposition is usually associated with a subaortic VSD [59].

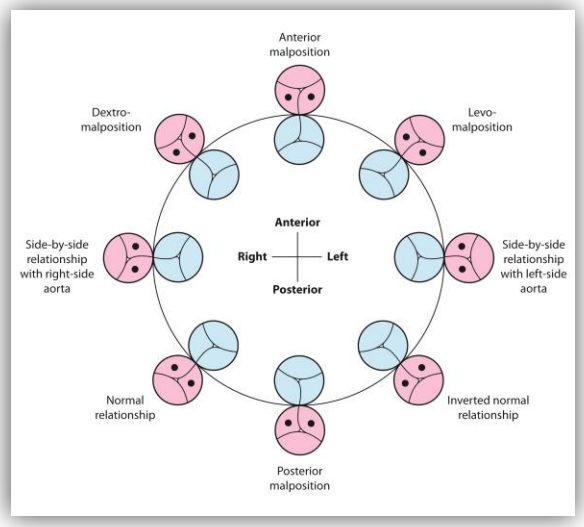

Figure 2-15. Great arterial relationship in situs solitus and atrioventricular concordant connection.

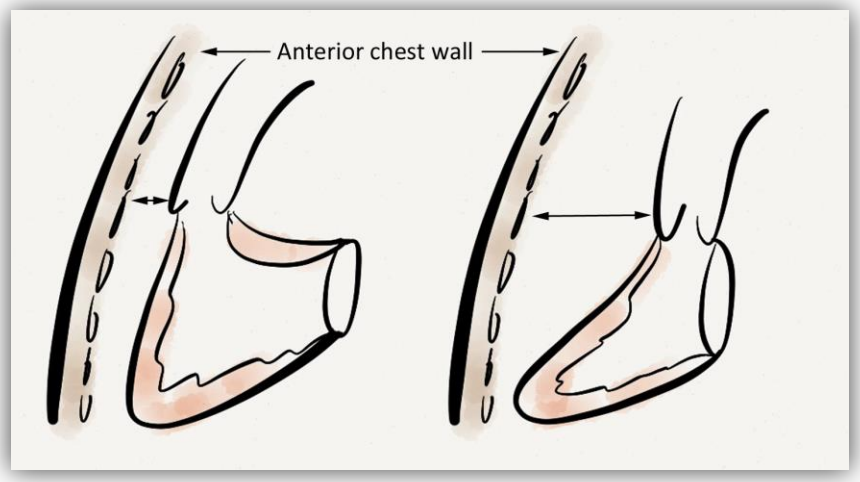

Figure 2-16. Relationship between the extent of the infundibulum and the location of the semilunar valve. The muscular infundibulum pushes the semilunar valve forward. The larger the muscular infundibulum, the closer the semilunar valve to the anterior chest wall (double- headed arrows) and the more superior the semilunar valve.

Relationship of the VSD to the Atrioventricular Conduction Axis

An important feature in the surgical management of DORV is the relationship of the atrioventricular conduction axis to the margin of the VSD [3, 4]. In cases with situs solitus and atrioventricular concordant connection, the atrioventricular node lies in the normal position in the atrioventricular septum behind its

membranous part. When the VSD has a completely muscular rim, the conduction axis keeps its normal course along the posteroinferior margin of the intact membranous septum, and, therefore, it is safe to stitch a patch on the VSD margin. More commonly, the VSD involves the membranous septum, making it a perimembranous type (**Table 2-4**). When the VSD is perimembranous, the atrioventricular conduction axis lies on the posteroinferior margin of the VSD and can be damaged by stitching a patch directly on this region.

Table 2-4. Incidence of perimembranous VSD according to the VSD types in pathology and surgical series.

	Wilcox et al (Pathology series) [12]	Kirklin et al (Surgical Series) [60]
Subaortic VSD	27 of 34 (79%)	46 of 57 (80%)
Subpulmonary VSD	9 of 16 (56%)	12 of 34 (35%)
Doubly committed VSD	4 of 7 (57%)	3 of 10 (30%)
Non-committed VSD	5 of 8 (63%)	15 of 26 (58%)
Total	45 of 65 (69%)	76 of 127 (60%)

Coronary Artery Anatomy

There is wide variation in the origins and epicardial courses of the coronary arteries in DORV. Understanding of the coronary arterial origins and courses is particularly important when an arterial switch operation needs to be performed in patients with a VSD in subpulmonary location or with a remote VSD that is bafflable only to the pulmonary trunk [61]. A major coronary arterial branch crossing the pulmonary outflow tract may preclude transannular augmentation of the stenotic or hypoplastic pulmonary valve.

Despite wide variations in great arterial relationship and coronary arterial anatomy, there is a certain degree of association between these two variables (**Figure 2-17**). When the great arteries are normally related, the coronary arteries tend to have the normal origins and epicardial courses. With the aortic valve more anteriorly located to have a side-by-side relationship, the left coronary artery originating from the Sinus 2 or posterior facing sinus takes a long course behind the subpulmonary outflow tract or pulmonary valve to bifurcate into the left anterior descending and circumflex coronary arteries. When the aorta is in a right anterior or directly anterior location to the pulmonary arterial trunk, the left coronary artery commonly arises from the Sinus 1 or left anterior facing sinus to course along the left and anterior aspect of the subpulmonary outflow tract and bifurcates into the left anterior descending and circumflex coronary arteries. Not infrequently, the left anterior descending coronary artery arises from the right coronary artery and courses anteriorly on the subaortic outflow tract. Infrequently, a single coronary artery arises from the Sinus 1 or 2. When the aorta is L-malposed, the right coronary artery usually courses anteriorly on the right-sided subpulmonary outflow tract to reach the right atrioventricular groove. An intramural course of the right or left coronary artery, although uncommon, can be a risk factor when an arterial switch procedure is attempted.

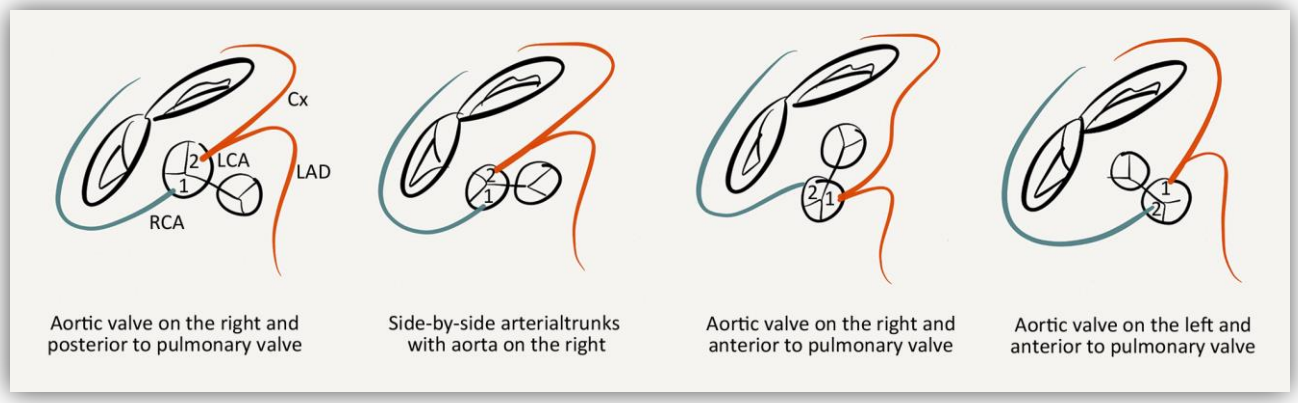

Figure 2-17. Coronary arterial origins and courses in DORV. Only typical examples are illustrated. The right (RCA) and left (LCA) coronary arteries tend to arise from the closest sinuses. The right coronary artery arises usually from the right-sided facing sinus, while the left coronary artery arises usually from the left-hand facing sinus, regardless of whether the aorta is anterior or posterior. When the arterial trunks show a side-by-side relationship with the aorta on the right, the left coronary artery tends to arise from the posterior facing sinus and the right coronary artery from the anterior facing sinus. Numbers 1 and 2 on the aortic valve indicates Sinus 1 and Sinus 2, respectively. Cx, circumflex branch; LAD, left anterior descending branch.

Associated Abnormalities of Clinical and Surgical Importance

Juxtaposition of the atrial appendages

Juxtaposition is a condition where the whole or part of the appendage of one atrium is displaced to the other side and positioned above the appendage of the other atrium. Left juxtaposition of the right atrial appendage is much more common than right juxtaposition [62-64]. When it occurs, this rare condition accompanies tricuspid atresia, double outlet right ventricle, complete transposition of the great arteries, or so-called criss-cross or twisted heart. DORV is typically associated with left juxtaposition (**Figure 2-18**). Juxtaposition of the atrial appendages causes a horizontal orientation of the atrial septum and can be associated with hypoplasia of one or both atria. Awareness of this rare condition is important as the unusual atrial anatomy may prolong the echocardiographic examination, or may complicate the surgical or interventional procedure when the atrial septal defect needs to be closed or when intra-atrial rerouting of the pulmonary or systemic venous pathways is required.

Abnormalities of the Atrioventricular Valves

Although infrequent, an abnormality of an atrioventricular valve is an important additional pathology that may complicate the surgical repair or preclude a biventricular repair. The atrioventricular valve abnormalities include stenosis, atresia, abnormal attachment of the chordae tendinae of the mitral and tricuspid valves to the margin of the VSD or subaortic or subpulmonary outflow tract, and straddling and/or overriding of one or both valves.

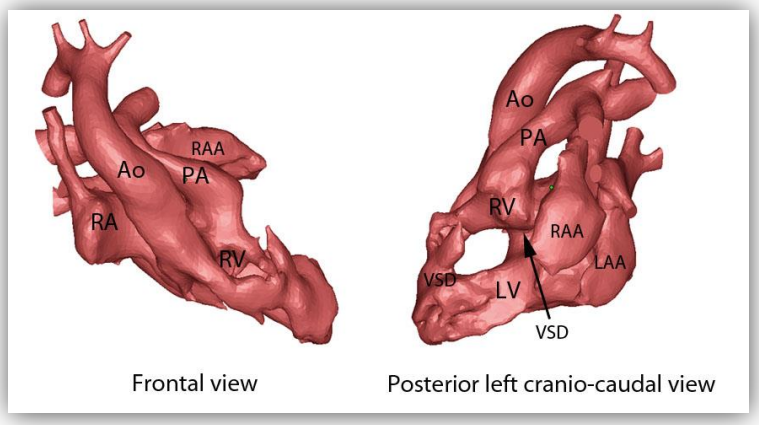

Figure 2-18. Left juxtaposition of the atrial appendages in a heart with superoinferior ventricular relationship and DORV.

Abnormal chordal attachment of the tricuspid or mitral valve is not infrequently seen in DORV [28, 65]. The chordae may insert to the outlet septum, the subaortic or subpulmonary outflow tract or the margin of the VSD. The abnormal chordal distribution and attachment may interfere with optimum construction of the intraventricular pathway to the aorta, although various approaches have been employed to avoid such complication. Rarely, the curtain-like chordae of the tricuspid valve may be seen.

Straddling or overriding of the tricuspid valve, mitral valve, or both valves is not frequent. Straddling implies the biventricular disposition of the tension apparatus of the atrioventricular valve across the ventricular septum, while overriding implies the biventricular spatial commitment of the valvular annulus across the ventricular septum [66-68]. Usually straddling is associated with a degree of overriding. However, straddling may occur without overriding, and overriding may occur without straddling. It has been shown that the tricuspid valve usually straddles through a VSD involving the inlet part of the septum that extends to the diaphragmatic surface of the ventricles, while the mitral valve typically straddles through a VSD involving the anterior part of the septum (**Figure 2-19**) [69-70]. Straddling atrioventricular valve occurring in association with DORV usually involves the mitral valve in cases with a subpulmonary VSD (**Figure 2-20**) [11, 67, 71-77]. It was postulated that the anterior part of the ventricular septum is displaced leftward and backward so that the anterior part of the mitral valve overrides the septum and its supporting structures remain in the right side of the septum [73]. The degree of straddling and overriding of the mitral valve varies according to the degree of inclination of the anterior part of the septum toward the left ventricle. With a greater degree of straddling and overriding of the mitral valve, the left ventricle becomes hypoplastic. The papillary muscle of the straddling mitral valve may arise from the crest of the VSD, the right ventricular aspect of the septum, the apex of the right ventricle, or the free wall of the right ventricle. As the straddling mitral valve frequently has its abnormal chordal attachment to the subpulmonary outflow tract, it causes some degree of subpulmonary stenosis [75]. In the case with a larger part of the pulmonary artery arising from the left ventricle, the straddling mitral valve more severely impedes the pulmonary blood flow as the mitral leaflet remains anchored in the pulmonary outflow tract during systole [75]. Straddling mitral valve can be seen with a subaortic VSD when the great arteries are L-malposed [78].

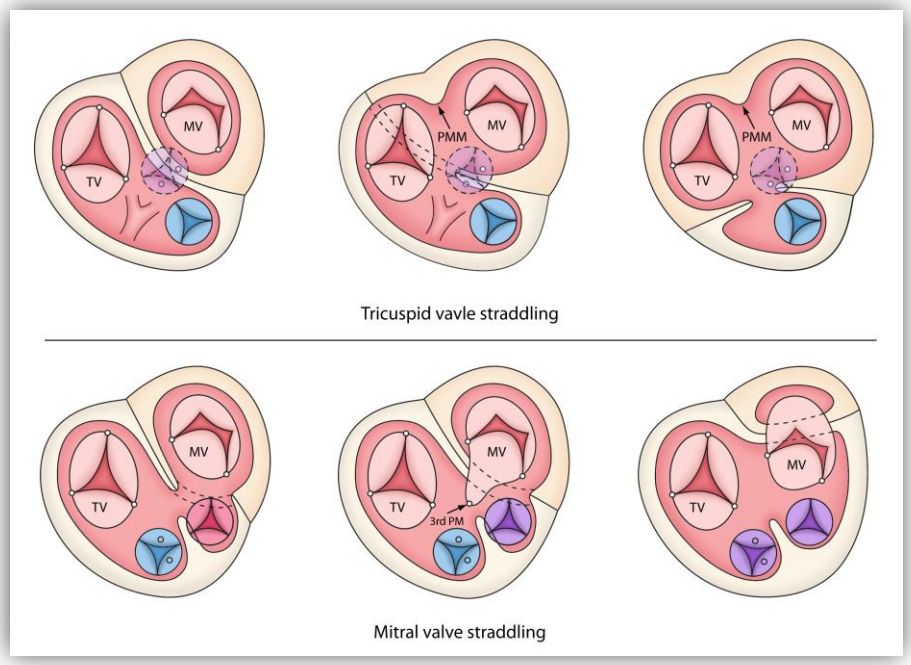

Figure 2-19. Straddling tricuspid versus straddling mitral valve. The tricuspid valve straddles through the defect involving the inlet part of the ventricular septum and extending to the diaphragmatic wall of the ventricles, while the mitral valve straddles through the defect involving the anterior part of the ventricular septum. With extreme straddling and overriding, the atrioventricular connection becomes univentricular with double inlet left or right ventricle.

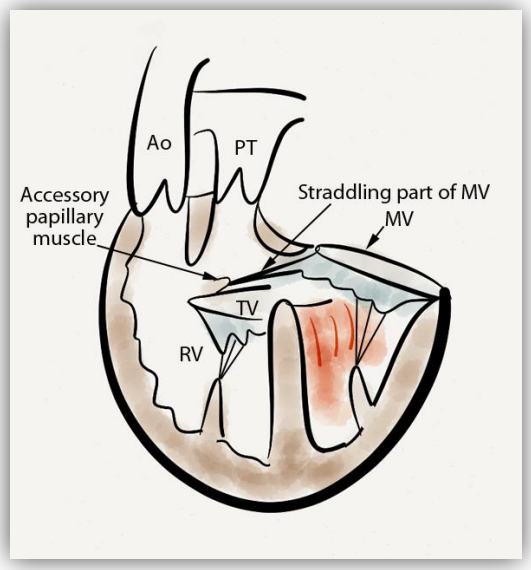

Figure 2-20. Cartoon showing straddling mitral valve in DORV with subpulmonary VSD. An accessory papillary muscle supporting the straddling part of the mitral valve is inserted to the subpulmonary outflow tract.

Ventricular outflow tract obstruction

Ventricular outflow tract is common in DORV with the pulmonary outflow tract obstruction more common than the aortic outflow tract obstruction (**Table 2-5**) [6, 7, 36]. Approximately two thirds of the DORV cases with a subaortic or doubly committed VSD and one third of the cases with a non-committed VSD are associated with pulmonary outflow tract obstruction. In DORV with a subpulmonary VSD, the pulmonary outflow tract obstruction is not common. In contrast, the aortic outflow tract dimension is usually smaller than the pulmonary outflow tract dimension, although there might not be overt stenosis in the beginning. With hypertrophy of the right ventricle and the outlet septum, especially sometime after pulmonary artery banding, the small subaortic outflow may develop significant stenosis [37, 38]. Frequent association of an obstructive lesion of the aortic arch in a form of tubular hypoplasia, coarctation or interruption in DORV with a subpulmonary VSD may be explained by the reduced flow into the small aortic outflow in fetal life [6, 7, 28, 36, 79, 80]. As discussed earlier, subpulmonary stenosis may also be due to the displaced leaflet and tension apparatus of the straddling mitral valve [39, 75].

Table 2-5. Compiled incidence of pulmonary and aortic outflow tract obstruction in DORV (6, 7, 36).

	Pulmonary stenosis	Subaortic stenosis	Obstructive lesion of the aortic arch
Subaortic VSD (181)	119 (66%)	5 (3%)	8 (4%)
Subpulmonary VSD (115)	16 (14%)	7 (6%)	35 (30%)
Doubly committed (22)	13 (59%)	3 (14%)	2 (9%)
Non-committed (68)	22 (32%)	4 (6%)	7 (10%)

Other associated abnormalities

DORV can be associated with all kinds of pathology including abnormal pulmonary or systemic venous connection, atrial septal defect or patent foramen ovale, unroofed coronary sinus and patent ductus arteriosus.

DORV with Situs Solitus and Discordant Atrioventricular Connection

This combination can be seen in either situs solitus or inversus and most cases share the features of congenitally corrected TGA [81]. When seen in situs solitus, there is a higher incidence of dextrocardia and L-malposition of the great arteries. The VSD is more frequently a subpulmonary than subaortic defect.

Summary

Double outlet right ventricle is a form of ventriculoarterial connection in which both arterial trunks arise entirely or predominantly from the right ventricle. Double outlet right ventricle encompasses a wide spectrum of malformations that require systematic assessment of the morphological modifiers at each anatomical level as summarized in **Table 2-6** [5]. Most importantly, feasibility of biventricular repair should be determined based on detailed assessment of the listed modifiers.

Table 2-6. List of essential modifiers of surgical anatomy of double outlet right ventricle [5].

Features	Primary	Secondary
Relationship of the atrioventricular conduction axis to the VSD margin	□ Perimembranous VSD □ Non-perimembranous VSD □ Atrioventricular septal defect	
Location of the VSD seen from the right ventricle	□ Predominantly outlet □ Predominantly inlet □ Confluent Inlet and outlet □ Predominantly apical trabecular □ Confluent involving all three parts	Relationship of the VSD to the tricuspid valve annulus: □ Along < upper 1/3 □ Along upper 1/3 to 2/3 □ Along > upper 2/3
Size and multiplicity of the VSD	□ Unrestrictive □ Restrictive □ No identifiable VSD	□ Single □ Multiple
Orientation of the outlet septum relative to the VSD margin	□ To the left margin of the VSD □ To the right margin of the VSD □ Parallel with the plane of the VSD □ Not related to the VSD margin □ Deficient or vestigial	
Muscular infundibulum	□ Subaortic □ Subpulmonary □ Bilateral □ Bilaterally deficient	Extent of the muscular infundibulum □ Long □ Short
Great arterial relationship	□ Normally related □ Mirror-image of normal □ Dextro-malposed □ Levo-malposed □ Side-by-side with aorta on the right □ Side-by-side with aorta on the left	
Outflow tract stenosis	□ Subaortic stenosis □ Aortic valvar stenosis □ Subpulmonary stenosis □ Pulmonary valvar stenosis □ Pulmonary valvar atresia	Aortic arch □ Unobstructed □ Tubular hypoplasia □ Coarctation □ Interruption
Type of DORV	VSD location per Lev, et al's classification: □ Subaortic □ Subpulmonary □ Doubly committed □ Non-committed or Remote □ Aligned with the subaortic outflow □ Aligned with the subpulmonary outflow □ Aligned with neither outflow	STS-EACTS-AEPC class*: □ VSD type □ Tetralogy type □ TGA type □ Non-committed VSD type □ AVSD
Atriovenricular valve abnormalities	□ Stenosis of the tricuspid or mitral valve □ Straddling and/or overriding of the tricuspid or mitral valve □ Insertion of the atrioventricular valve tension apparatus to the margin of the VSD or outlet septum	
Ventricular volumes	Right ventricular volume □ Enough space for intraventricular baffling □ Too little space for intraventricular baffling	Left ventricular volume □ Normal □ Borderline hypoplasia □ Too small
Other findings and associated abnormalities	□ Anomalous systemic venous connection □ Anomalous pulmonary venous connection □ Juxtaposition of the atrial appendages □ Coronary arterial origins and distribution	

*STC, Society of Thoracic Surgeons; EACTS, European Association of Cardiothoracic Surgery; AEPC, Association of European Pediatric Cardiology

AVSD, atrioventricular septal defect; DORV, double outlet right ventricle; TGA, transposition of the great arteries; VSD, ventricular septal defect

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CHAPTER 3. SURGICAL OPTIONS

The surgical approach to double outlet right ventricle (DORV) varies according to the given anatomic features [1-5]. The ideal surgical repair is a biventricular repair by connecting the morphologically left ventricle to the aorta and the morphologically right ventricle to the pulmonary artery. There are occasions when the left ventricle can only be connected to the pulmonary artery and the repair is completed by an arterial or atrial switch operation. A biventricular repair is feasible in the majority of patients with two well-developed ventricles, while severe hypoplasia of a ventricle, significant mitral or tricuspid valve pathology, a remote location of the ventricular septal defect (VSD), multiple VSDs and complex form of pulmonary atresia may necessitate a univentricular repair or other type of palliative procedure.

Although well-known pathological classification of DORV based on the relationship between the VSD and the great arterial trunks is useful, the commitment of the VSD to an arterial valve or valves does not always predict the surgical approach [1-3]. Recent classification named by the STS-EACTS [International Nomenclature named after the Society of Thoracic Surgeons (STC) and the European Association of Cardiothoracic Surgery (EACTS)] and adopted by the Association of European Pediatric Cardiology (AEPC) defines four types of DORV based on the clinical presentation and treatment [1, 6-9]: 1) VSD-type, 2) tetralogy-type, 3) transposition of the great arteries (TGA)-type, and 4) non-committed VSD-type (**Table** 3-1).

Table 3-1. STS-EACTS-AEPC classification of VSDs in DORV.

- VSD-type (25%) - Tetralogy-type (35%) - TGA-type (20%) - Non-committed VSD-type (20%)

* STC, Society of Thoracic Surgeons; EACTS, European Association of Cardiothoracic Surgery; AEPC, Association of European Pediatric Cardiology

1. DORV, VSD-type

This type is characterized by a subaortic or doubly committed VSD without pulmonary stenosis showing the clinical signs of overcirculation from an unrestrictive VSD. Usually, it requires a one-stage complete repair within the first 6 months of life. Rarely, in patients with refractory congestive heart failure, initial palliation with a pulmonary artery band to protect the pulmonary vascular bed is required. The repair consists of intraventricular routing of the VSD to the aortic valve using a tunnel-like patch. When the VSD is restrictive, enlargement of the VSD can be required to avoid left ventricular outflow tract obstruction. The VSD is considered restrictive when its diameter is smaller than the diameter of the aortic valve [10-12]. The restrictive VSD is enlarged by making an incision or by resecting a wedge of the septum in an anterior and superior direction as the atrioventricular conduction axis courses along or is a few millimeters away from the postero-inferior margin of the VSDt. Occasionally, the outlet septum should be resected to construct a

straight tunnel in order to avoid pulmonary outflow tract obstruction. Repair can usually be achieved through a right atrium but occasionally needs a right ventriculotomy.

2. DORV, Tetralogy-type

This type is characterized by a subaortic or doubly committed VSD with pulmonary outflow tract obstruction showing various degree of cyanosis. It requires a complete repair during the first year of life. The VSD can also be a complete atrioventricular septal defect extending toward the outlet with deviation of the outlet septum encroaching on the pulmonary outflow tract. The repair of Fallot-type of DORV consists of relief of subpulmonary outflow tract obstruction through resection of the outlet septum and obstructing muscle bundles when present, and intraventricular tunneling of the VSD to the aorta. Although pulmonary valvotomy or augmentation with an outflow tract patch can be required, preservation of the native pulmonary valve function or restricted enlargement of the pulmonary valve is considered desirable [13-17]. When the right coronary artery or left anterior descending coronary artery takes an anomalous course across the subpulmonary outflow tract, a valved extracardiac conduit needs to be inserted from the right ventricle to the pulmonary artery. Although an early one-stage surgery is preferred, severe pulmonary arterial hypoplasia may need early palliation with a systemic-to-pulmonary arterial shunt.

3. DORV, TGA-type (Taussig-Bing malformation)

This type is characterized by a subpulmonary VSD and is often associated with a degree of subaortic narrowing and tubular hypoplasia, coarctation, or interruption of the aortic arch. The patients present with severe cyanosis and heart failure in the immediate postnatal period as in complete TGA. If applicable, one-stage operation is required in the first or second week of life. When a one-stage surgery is considered to be associated with a high risk, delayed definitive repair after initial palliation with pulmonary artery banding and correction of the aortic arch obstruction and subaortic stenosis has been suggested [18].

Most cases are not associated with pulmonary stenosis and are amenable to either arterial switch procedure with baffling of the VSD to the neo-aorta or intraventricular repair with construction of a direct tunnel from the VSD to the aorta (Kawashima procedure) [1, 19-25]. The Kawashima's intraventricular repair is applicable when the tricuspid and pulmonary valves are positioned wide apart, leaving enough space for the unobstructed tunnel [2, 3, 25, 26]. The Kawashima procedure requires resection of the outlet septum to secure the unobstructed left ventricular outflow tract. Both arterial switch and intraventricular repair requires enlargement of the VSD when the size of the defect is less than the size of the aortic valve. The incision for enlargement should be on the anterior rim of the VSD as the atrioventricular conduction axis is on or at a short distance from the posteroinferior rim of the VSD. In general, the cases with the great arteries in an antero-posterior relationship tend to have a shorter tricuspid-pulmonary valve distance and are more suitable for an arterial switch procedure, while those with the great arteries in a side-by-side relationship tend to have a wider tricuspid-pulmonary valve distance and are suitable for intraventricular repair [19, 23]. If achievable, the intraventricular repair is advantageous compared to arterial switch operation because there is no need for reimplantation of the coronary arteries, and the risk of later development of aortic regurgitation is avoided [23]. This is especially the case when an unusual coronary artery anatomy is a risk

factor or the pulmonary valve is not considered adequate to function as a systemic valve, such as in patients with prior pulmonary artery banding.

When pulmonary stenosis precludes an arterial switch operation with a VSD closure and a standard intraventricular tunnel repair is not applicable, an REV (*Réparation à l'Etage ventriculaire)* procedure can be performed [28-31]. The REV procedure consists of extensive excision of the outlet septum when present, translocation of the pulmonary artery through the gap in the dissected ascending aorta, tunneling of the VSD to the aortic valve and direct anastomosis of the pulmonary artery to the right ventricle using a pericardial patch or monocuspid dacron patch. The REV procedure is a modification of the Rastelli operation which is often complicated by both left and right ventricular outflow tract obstruction due to the long curved nature of the intraventricular tunnel and the lack of growth potential of the external conduit [32, 33]. The REV procedure provides a straighter and wider tunnel from the left ventricle to the aorta and the growth potential for the right ventricular outflow tract as compared to the original Rastelli procedure [28, 29, 33]. It also allows complete repair in infancy. An external valved conduit between the right ventricle and the pulmonary arterial trunk is used only when the pulmonary arterial trunk cannot be directly anastomosed to the right ventricle.

Recently, there has been revival of the Nikaido procedure which consists of mobilization of the aortic root with its valve from the right ventricle, resection of the outlet septum and excision of the pulmonary valve, implantation of the mobilized aortic root in the pulmonary location, patch closure of the VSD, and reconstruction of the right ventricular outflow tract to the pulmonary artery using a pericardial patch [34]. When the stenotic or hypoplastic pulmonary valve is still usable, the pulmonary root can also be mobilized and translocated to the aortic position and the pulmonary outflow tract is augmented by using a transannular pericardial patch. This procedure is called double-root translocation [35].

4. DORV, non-committed VSD-type

The non-committed location of the VSD is defined as a distance of the VSD from both aortic and pulmonary valve greater than the diameter of the matched (i.e, normal) aortic valve [8, 36, 37]. The remoteness of the VSD may preclude biventricular repair and instead requires multiple staged operations toward a Fontan circuit [38-40]. However, biventricular repair is often achievable with a long tunneling of the VSD to the aorta or to the pulmonary artery [8, 36, 37, 41]. When the VSD is tunneled to the pulmonary artery, the aorta and pulmonary arteries should be translocated by using an arterial switch or Nikaidoh operation or double-root translocation procedure depending on the size and healthiness of the pulmonary valve and the absence or presence of subpulmonary stenosis [35]. The surgical procedure for DORV with remote VSD is usually undertaken through a right ventriculotomy, especially when the VSD involves the apical trabecular septum. In the majority of the cases, the VSD is restrictive and needs to be enlarged anteriorly and superiorly [41]. More than one VSD patch is often required to avoid obstruction of the right ventricular inlet or injury to the tricuspid valve tension apparatus [41]. Because the intraventricular tunnel occupies a substantial space in the right ventricle, the right ventricular outflow tract often needs to be enlarged or reconstructed using a conduit. As the corrective surgery involves complex intraventricular repair requiring a long cross-clamp time, staged operation with a Blalock-Taussig shunt or pulmonary artery banding in early infancy and delayed corrective repair in later infancy is often preferred [8].

When biventricular repair cannot be undertaken, univentricular repair is indicated. In addition to the remoteness of the VSD to an arterial valve, straddling of an atrioventricular valve or valves, severe hypoplasia of one ventricle and multiple VSDs are the indications for univentricular repair. Univentricular repair consists of Blalock-Taussig shunt or pulmonary artery banding in the first few weeks of life, bidirectional cavopulmonary connection at 4-9 months of age and modified Fontan operation at 2-4 years. Although biventricular repair is a preferred option, it should be emphasized that biventricular repair requiring a complex intraventicular procedures is associated with a higher requirement for reintervention than univentricular repair [41, 42, 43].

DORV with discordant atrioventricular connection

This type is a variant of congenitally corrected TGA. The clinical presentation may vary according to the relationship of the VSD to the arterial valves and the presence or absence of obstruction of the aortic and pulmonary outflow tracts. Until the ate 1980's, TGA or DORV in the setting of discordant atrioventricular connection had been managed by tunneling the VSD to the pulmonary valve so that the morphologically right ventricle supports the systemic circulation and the morphologically left ventricle supports the pulmonary circulation [44]. With increasing incidence of failure of the systemic right ventricle and development of tricuspid regurgitation, the concept of "anatomical repair" using the left ventricle as the systemic ventricle has been introduced. The "anatomic repair" includes combined Mustard/Senning atrial switch procedure, tunneling of the VSD to the pulmonary valve and arterial switch operations [45-48]. Although the "anatomical repair" appears ideal, its long-term outcome is still debated [49]. When an anatomical repair is opted, an elective anatomic repair beyond 6 months of age is recommended since the intracardiac procedure is complex [50-52]. In neonates with severe cyanosis due to severe pulmonary stenosis or hypoplastic pulmonary arteries, a palliative shunt is placed to allow growth of the cardiovascular structures. In neonates with unobstructed pulmonary outflow tract and heart failure, pulmonary artery banding is required until the optimum age for complete repair.

DORV in Heterotaxy syndrome

Heterotaxy syndrome, especially the right isomerism is often associated with DORV. The interventricular communication is usually through an AVSD that often extends toward the outlet [9, 37]. In this particular setting, subpulmonary stenosis due to deviated outlet septum is a rule rather than an exception. Some surgeons regard the cases with an AVSD extending toward the aortic outflow tract and pulmonary outflow tract stenosis as DORV of tetralogy type [8, 9, 37). Traditionally univentricular repair has been favored because of the proximity of the atrioventricular valve leaflets and their tension apparatuses to the VSD, as well as the frequent association with total anomalous pulmonary venous connection [52]. A more recent study showed that biventricular repair was feasible in the majority of the cases with a low surgical mortality rate [8, 9]. The intraventricular tunneling of the VSD to the aorta requires division of the anterior bridging leaflet of the common atrioventricular valve. Both biventricular and univentricular repairs are associated with

high risks when there is atrioventricular valve regurgitation or pulmonary venous obstruction and when the patient presents in the early neonatal period [52].

DORV with Abnormal Chordal Insertion or Straddling of the Atrioventricular Valves

When the tricuspid valve has an abnormal chordal attachment to the outlet septum, the outlet septum is not resected, but mobilized with the chordal insertions [3, 29, 54-56]. After the intraventricular tunnel is constructed using a patch, the mobilized outlet septum is sutured to the patch. A similar technique can also be used for those with tricuspid chordal attachment to the subpulmonary and/or subaortic outflow tract [56]. More extensive chordal attachment of the tricuspid or mitral valve all around the VSD margin precludes biventricular repair [56]. When there is straddling of the tricuspid or mitral valve, the abnormal chordae or papillary muscle can be retracted toward the ventricle that the straddling valve belongs to and the VSD is closed on the opposite side of the septum [56].

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CHAPTER 4. CASE SERIES

CASE 1. DORV with a subaortic VSD and mild subpulmonary stenosis (so-called VSD type)

CASE 2. DORV with a subaortic VSD and subpulmonary stenosis (so-called tetralogy-type)

CASE 3. DORV with a restrictive subaortic VSD and subpulmonary stenosis

CASE 4. DORV with a subpulmonary VSD (so-called TGA-type or Taussig-Bing anomaly)

*CASE 5.*DORV with a subpulmonary VSD (so-called TGA-type or Taussig-Bing anomaly)

CASE 6. DORV with a doubly-committed non-perimembranous VSD

CASE 7. DORV with a subaortic non-perimembranous VSD with absent outlet septum

CASE 8. DORV with a non-committed VSD

CASE 9. DORV with a non-committed VSD related to subaortic outflow tract

CASE 10. DORV with a VSD related to subaortic outflow tract at a distance from aortic valve

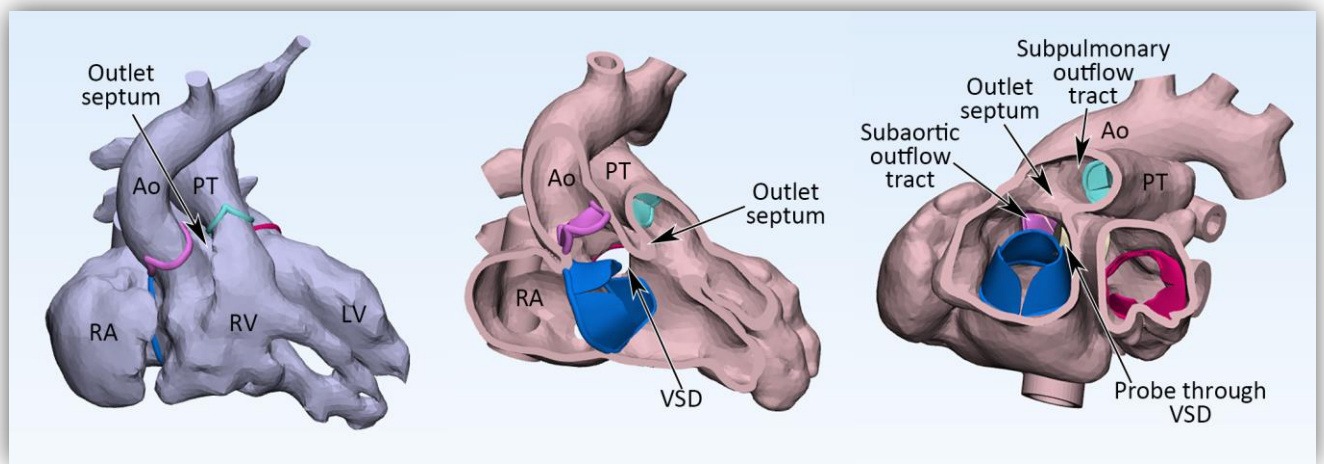
CASE 11. DORV with a non-committed VSD related to subpulmonary outflow tract

CASE 12. DORV with a VSD related to subpulmonary outflow tract at a distance from pulmonary valve.

CASE 1. **DORV with a subaortic VSD and mild subpulmonary stenosis (so-called VSD type)**

- ❖ Source images: Contrast-enhanced MR angiogram.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with long subpulmonary and no subaortic infundibulum
- ♥ Normally related arterial trunks
- ♥ Subaortic perimembranous VSD
- ♥ Left pulmonary artery sling



- Both aorta and pulmonary arterial trunk arise from the morphologically right ventricle.
- The ascending aorta is located more anteriorly than it is normally seen. However, the ascending aorta and pulmonary arterial trunk show a normal spatial relationship.
- The right ventricular outflow tract is divided into the right posterior subaortic and left anterior subpulmonary outflow tracts by the outlet septum. The subpulmonary outflow tract is a long muscular infundibulum. The subaortic outflow tract is short with fibrous continuity between the aortic and tricuspid valve attachments. The subpulmonary infundibulum is slightly narrower than the subaortic infundibulum.
- The outlet septum is exclusively a right ventricular structure, appearing deviated forward and leftward from its normal position.
- A large VSD involves the superior and posterior part of the ventricular septum. Its posterior margin is in direct contact with the uppermost part of the tricuspid valve where the septal and anterior leaflets form a commissure. This location indicates that the defect is a perimembranous defect. The atrioventricular conduction axis is considered to be disposed on the posterior inferior rim of the VSD.
- The VSD extends leftward and is located below the aortic valve. As the prominent outlet septum is fused to the left anterior margin of the VSD, the defect is committed exclusively to the right-sided aortic valve. The left-sided pulmonary valve is isolated from the defect by the outlet septum.
- As an incidental finding, the left pulmonary artery arises distally from the right pulmonary artery forming a sling on the right side of the trachea (not shown).

- ❖ *Compare this case with Case 2 and 3.*

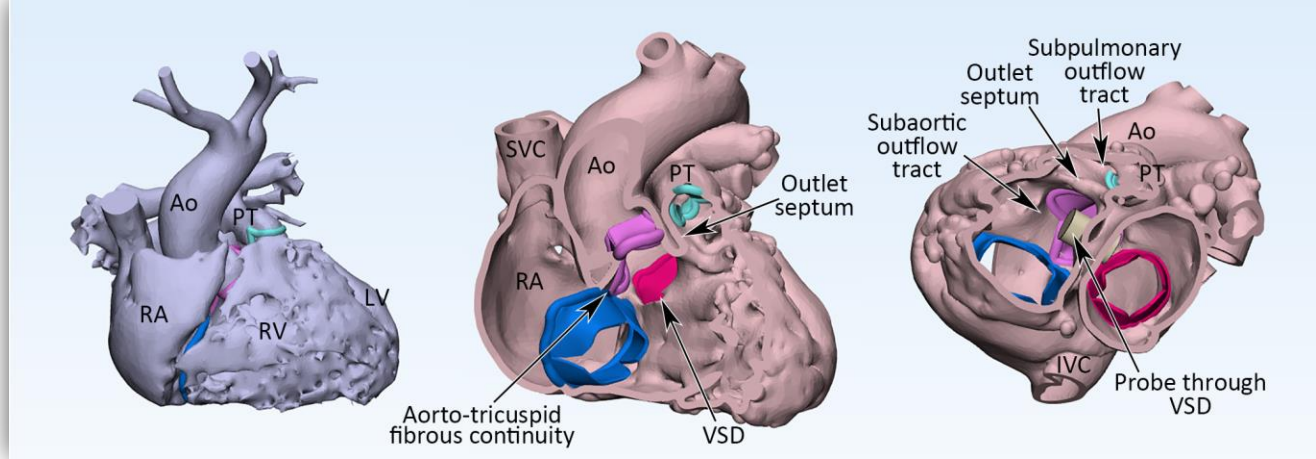
Surgical Consideration:

This heart has normally related great arteries with slightly small pulmonary artery compared to the aorta and a moderate to large subaortic VSD. The VSD locates just beneath the aortic valve. The right ventricular outflow tract and the pulmonary valve are somewhat small but appear to be unobstructed. The location of the VSD is favorable for intra-ventricular baffling. The VSD probably has to be enlarged given that it is smaller than the size of the aortic valve. The baffle does not seem to compromise the tricuspid valve inflow or the right ventricular outflow tract. In addition, the left pulmonary artery sling has to be repaired by a standard reimplantation technique. Overall this heart is suitable for primary biventricular repair with an intra-ventricular baffle.

***CASE 2*. DORV with a subaortic VSD and subpulmonary stenosis (so-called tetralogy-type)**

- ❖ Source images: Contrast-enhanced CT angiograms.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with subpulmonary infundibulum and no subaortic infundibulum
- ♥ Normally related arterial trunks
- ♥ Subaortic perimembranous VSD
- ♥ Subpulmonary stenosis
- ♥ Small patent ductus arteriosus



- Entire pulmonary arterial trunk and > 75% of aorta arise from the morphologically right ventricle.
- The ascending aorta is located more anteriorly than it is normally seen. However, the ascending aorta and pulmonary trunk show a normal spatial relationship.
- The right ventricular outflow tract is divided into the right-sided subaortic and left-sided subpulmonary outflow tracts by the outlet septum
- The outlet septum is exclusively a right ventricular structure, appearing deviated forward and leftward from its normal position.
- A large VSD involves the superior and posterior part of the ventricular septum. Its posterior margin is in direct contact with the uppermost part of the tricuspid valve where the septal and anterior leaflets form a commissure. The aortic valve is in direct contact with the tricuspid valve. This location indicates that the defect is a perimembranous defect. The atrioventricular conduction axis is considered to be disposed on the ventricular septal crest.
- The VSD extends leftward and is located below the aortic valve. As the prominent outlet septum is fused to the left anterior margin of the VSD, the defect is committed exclusively to the right-sided aortic valve. The left-sided pulmonary valve is isolated from the defect by the outlet septum.
- The subpulmonary outflow tract is a narrow muscular infundibulum. The outlet septum isolates the subpulmonary infundibulum from the defect. There are small muscle bundles in the subpulmonary outflow tract. The main pulmonary artery is mildly hypoplastic and the branch pulmonary arteries are of good size.

- ❖ *Compare this case with Case 1 and Case 3.*

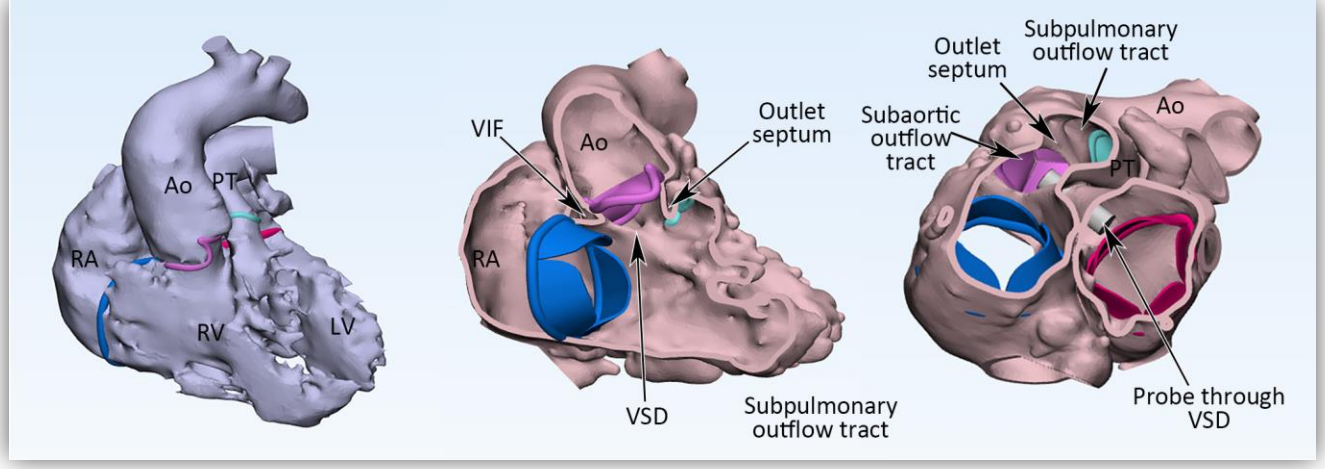
Surgical Consideration:

This heart is so-called tetralogy type of DORV characterized by >75% aortic override, significant right ventricular outflow tract obstruction, and a hypoplastic pulmonary valve. There is a large unrestrictive subaortic VSD. The main pulmonary artery is mildly hypoplastic, and the branch pulmonary arteries are relatively well developed. There is a tiny patent ductus arteriosus. Surgical approach to this VSD is either an intra-ventricular baffle with an open tube graft. The size of VSD appears to be adequate so VSD enlargement is less likely to be needed. Extensive right ventricular muscle bundle resection, pulmonary valvotomy, and main pulmonary artery patch plasty are required. A transannular patch may not be needed.

***CASE 3*. DORV with a restrictive subaortic VSD and subpulmonary stenosis**

- ❖ Source images: Contrast-enhanced CT angiograms.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with subpulmonary and subaortic infundibulum
- ♥ Parallel arterial trunks with the aorta on the right anterior to the pulmonary arterial trunk
- ♥ Subaortic restrictive non-perimembranous VSD
- ♥ Subpulmonary stenosis



- Both aorta and pulmonary arterial trunk arise from the morphologically right ventricle.
- The arterial trunks show a parallel relationship with the ascending aorta located rightward and forward. Note that the anterior location of the aorta breaks the normal relationship of the arterial trunks.
- The right ventricular outflow tract is divided into the right-side anterior subaortic and left-side posterior subpulmonary outflow tracts by the outlet septum
- The outlet septum is exclusively a right ventricular structure.
- A small VSD involves the outlet part of the ventricular septum. The VSD is separated from the tricuspid valve annulus by a muscular rim (the posterior limb of the trabecula septomarginalis) and does not involve the membranous septum. The atrioventricular conduction axis is not considered disposed directly on the ventricular septal crest.
- As the prominent outlet septum is fused to the left anterior margin of the VSD, the defect is committed exclusively to the right-sided aortic valve. The left-sided pulmonary valve is isolated from the defect by the outlet septum.
- The subpulmonary outflow tract is a narrow muscular infundibulum behind the outlet septum. The outlet septum isolates the subpulmonary infundibulum from the defect.

❖ *Compare this case with Case 1 and Case 2.*

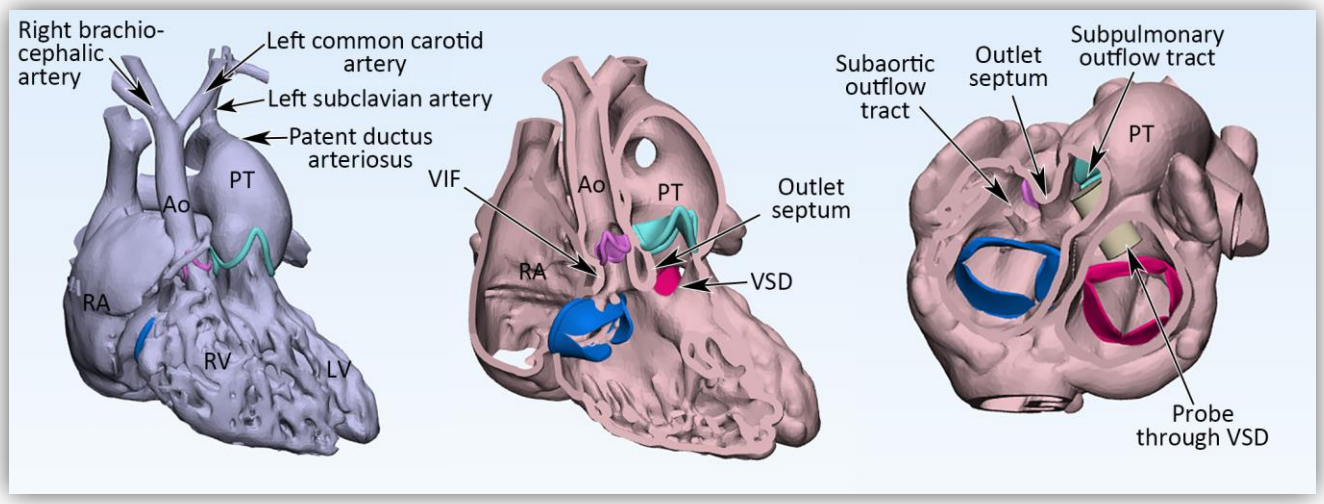
Surgical Consideration:

This heart has right ventricular outflow tract obstruction with hypoplastic pulmonary valve and main pulmonary artery. More importantly the subaortic VSD is very restrictive. Although it is not clearly shown in the models, it is quite possible that left ventricle has pathologic hypertrophy due to outflow tract obstruction at the VSD level. This heart needs an urgent operation in a neonatal period to release the left ventricular outflow tract obstruction. The decision on staged operations vs. primary full repair will be dictated by the degree of left ventricular hypertrophy, and age and body weight of the patient. If there is considerable left ventricular hypertrophy, it may be a wiser option to palliate this heart with VSD enlargement and additional right ventricular outflow tract surgery. If left ventricular hypertrophy is not significant and the patient is good size, primary full repair, consisting of VSD enlargement, intra-ventricular baffling, and reconstruction of the right ventricular outflow tract, could be performed.

***CASE 4*. DORV with a subpulmonary VSD and subaortic stenosis (so-called TGA-type or Taussig-Bing anomaly)**

- ❖ Source images: Contrast-enhanced CT angiograms.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with subaortic and subpulmonary infundibulum
- ♥ Side-by-side relationship of the arterial trunks with the right-sided aorta
- ♥ Subpulmonary non-perimembranous VSD
- ♥ Subaortic stenosis
- ♥ Interruption of the aortic arch distal to the origin of the left common carotid artery



- The aorta and 80% of the pulmonary arterial trunk arise from the morphologically right ventricle.
- The arterial trunks show a side-by-side parallel spatial relationship with the aorta on the right.
- The right ventricular outflow tract is divided into the right-sided subaortic and left-sided subpulmonary outflow tracts by the outlet septum, which is exclusively a right ventricular structure.
- Both aortic and pulmonary valves are supported by the muscular infundibulum.
- The VSD involves the uppermost aspect of the septum below the pulmonary valve that overrides the ventricular septum. The VSD is separated from the tricuspid valve annulus and therefore from the membranous septum by a muscular rim. Therefore, the atrioventricular conduction axis is not disposed on the ventricular septal crest.
- The aortic valve is located remote from the VSD and isolated from the defect by the outlet septum.
- The muscular subaortic outflow tract is encroached on between the outlet septum and the ventriculoinfundibular fold (VIF). The aortic valve is small.
- Noticeably the muscular outlet septum is perpendicular to the ventricular septum and fused to the right margin of the defect, leaving the defect solely committed to the overriding pulmonary valve and isolating the subaortic outflow tract from the VSD.
- The aortic arch shows type B interruption. The ascending aorta bifurcates into the right brachiocephalic and left common carotid arteries and does not connect to the descending aorta. The pulmonary arterial trunk connects to the descending aorta through the narrow ductus arteriosus. The left subclavian artery arises from the top of the descending aorta.

n *Compare this case with Case 5.*

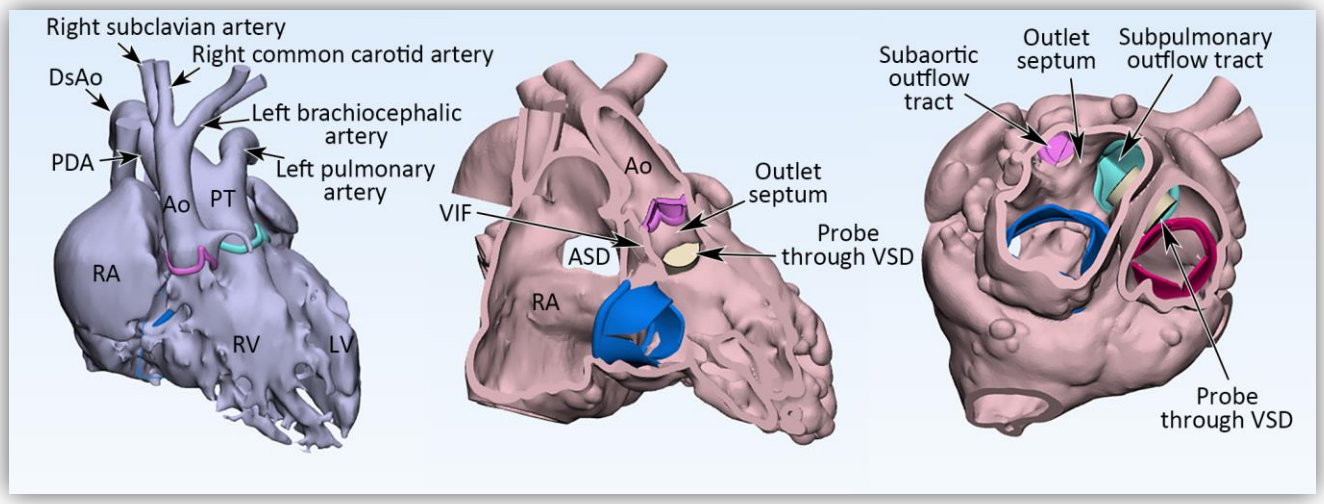
Surgical Consideration:

This heart has features of Taussig-Bing anomaly, consisting of a subpulmonary VSD, transposed and side-by-side great arteries arrangement with hypoplastic aorta, and bilateral conus. In addition, there is an interruption of the aortic arch after the second head and neck branch, and a moderate sized arterial duct connects to the distal arch, maintaining lower body perfusion. There are 2 coronary artery systems arising from the facing sinuses (1L, 2R) although the take-off of the circumflex coronary artery is not shown. The anatomy is suitable for neonatal primary repair, consisting of arterial switch operation, aortic arch reconstruction, and intra-ventricular baffling to the neo-aortic root. The entire aortic arch has to be reconstructed with a generous patch to match the size of the neo aortic root, which is quite dilated. Additional right ventricular muscle resection is almost always necessary. The coronary artery pattern appears to be favorable for coronary transfer.

CASE 5. DORV with a subpulmonary VSD and subaortic stenosis (so-called TGA-type or Taussig-Bing anomaly)

❖ Source images: Contrast-enhanced CT angiograms.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with subaortic infundibulum and no subpulmonary infundibulum
- ♥ Aorta on the right anterior aspect of the pulmonary arterial trunk
- ♥ Subpulmonary perimembranous VSD
- ♥ Subaortic stenosis
- ♥ Right aortic arch with Interruption of the aortic arch distal to the origin of the right subclavian artery



- The aorta and 60% of the pulmonary arterial trunk arise from the morphologically right ventricle.
- The ascending aorta is located rightward and anterior to the pulmonary arterial trunk.
- The right ventricular outflow tract is divided into the right anterior subaortic and left posterior subpulmonary outflow tracts by the outlet septum, which is exclusively a right ventricular structure.
- The aortic valve is supported by a narrow muscular infundibulum. The pulmonary valve is in direct contact with the mitral valve without intervening subpulmonary infundibulum.
- The VSD involves the uppermost part of the septum below the pulmonary valve that overrides the ventricular septum. The aortic valve is located remote from the VSD and further isolated from the defect by the outlet septum. The posterior margin of the defect reaches the commissure between the anterior and septal leaflets of the tricuspid valve and therefore the membranous septum. The defect is a large perimembranous VSD extending toward the outlet of the right ventricle. The atrioventricular conduction axis is considered disposed on the posteroinferior margin of the defect.
- The subaortic infundibulum is surrounded by the ventriculoinfundibular fold on the right, the free wall anteriorly and leftward and the outlet septum posteriorly.
- Noticeably the outlet septum does not directly insert to either side of the VSD but is parallel to the rest of the ventricular septum. It is in contrast to Case 4 in which the outlet septum is perpendicular to the septum and inserts to the right margin of the VSD.
- The aortic arch is right sided and interrupted distal to the origin of the right subclavian artery (Type A interruption). The pulmonary trunk connects to the descending aorta (DsAo) on the right through the mildly restrictive patent ductus arteriosus (PDA).

❖ *Compare this case with Case 4.*

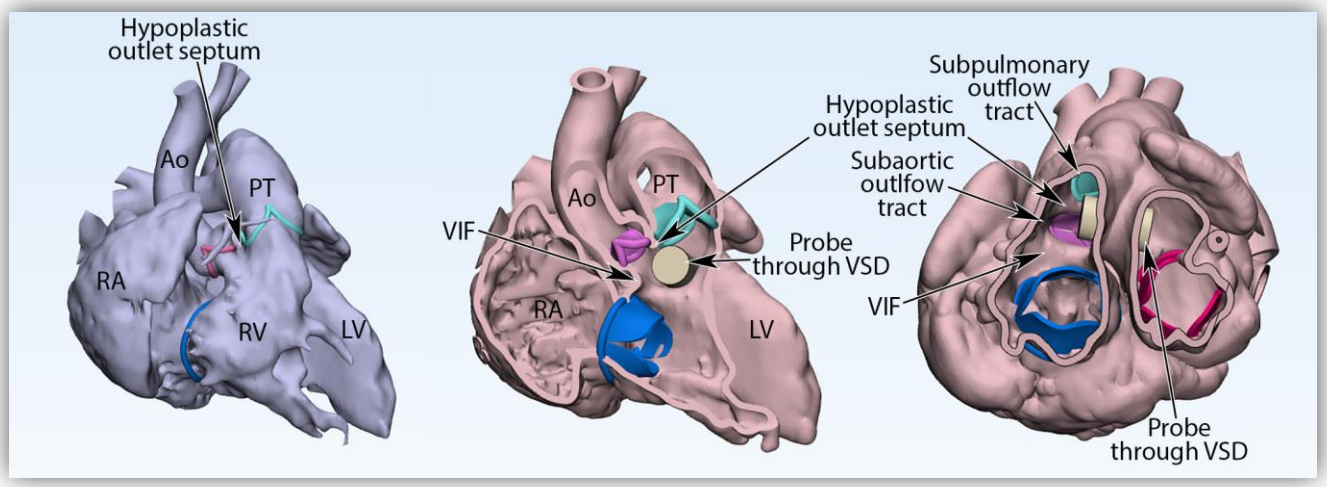
Surgical Consideration:

This heart has features of Taussig-Bing anomaly with a large subpulmonary VSD and the aortic valve located right and anterior to the pulmonary valve. The ascending aorta is hypoplastic aorta and the aortic arch is interrupted after giving rise to all head and neck branches. Approximately two thirds of the pulmonary artery is committed to the right ventricle. There are two coronary artery systems coming off facing sinuses (1L, 2R). The status of the circumflex coronary artery is not shown. This heart is suitable for neonatal primary repair consisting of arterial switch operation, aortic arch reconstruction, and intra-ventricular baffling. Given the location of the pulmonary artery in relation to the VSD, extensive and long intra-ventricular baffling is unnecessary and the shape of the patch can be relatively flat. The aortic arch should be augmented with a patch to match the size of the neo-aortic root.

CASE 6. DORV with a doubly-committed non-perimembranous VSD

- ❖ Source images: Contrast-enhanced CT angiograms.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with short subaortic and long subpulmonary infundibulum
- ♥ Side-by-side relationship of the arterial trunks with the right-sided aorta
- ♥ Doubly committed, non-perimembranous VSD
- ♥ Small size of the aortic valve as compared to pulmonary valve
- ♥ Tubular hypoplasia of the aortic arch and restrictive patent ductus arteriosus
- ♥ Single coronary artery arising from Sinus 1.



- The aorta and 80% of the pulmonary arterial trunk arise from the morphologically right ventricle.
- The arterial trunks show a side-by-side parallel spatial relationship with the aorta on the right.
- The right ventricular outflow tract is a common outflow tract supporting the right-sided aortic valve and left-sided pulmonary valve. The outlet septum is very hypoplastic.
- The VSD involves the uppermost aspect of the ventricular septum below the pulmonary valve and extends downward along the aortic valve. Although the VSD is doubly-committed, it appears committed more to the pulmonary valve than the aortic valve. The VSD does not reach the tricuspid valve annulus. A muscular rim separates the defect from the tricuspid valve annulus and therefore from the membranous septum. The atrioventricular conduction axis is not considered disposed directly on the ventricular septal crest.
- The hypoplastic outlet septum is located above the central part of the upper margin of the VSD.
- The subaortic outflow tract is narrow between the hypoplastic outlet septum and the ventriculoinfundibular fold (VIF). The aortic valve is smaller than the pulmonary valve.
- The aorta shows diffuse tubular hypoplasia of its arch. The aortic isthmus is long and narrow. The aortic isthmus and restrictive patent ductus arteriosus form a confluence at the descending aorta with a shallow posterior shelf.
- A single coronary artery arises from the anterior facing sinus (Sinus 1).

- ❖ *Compare this case with Case 7.*

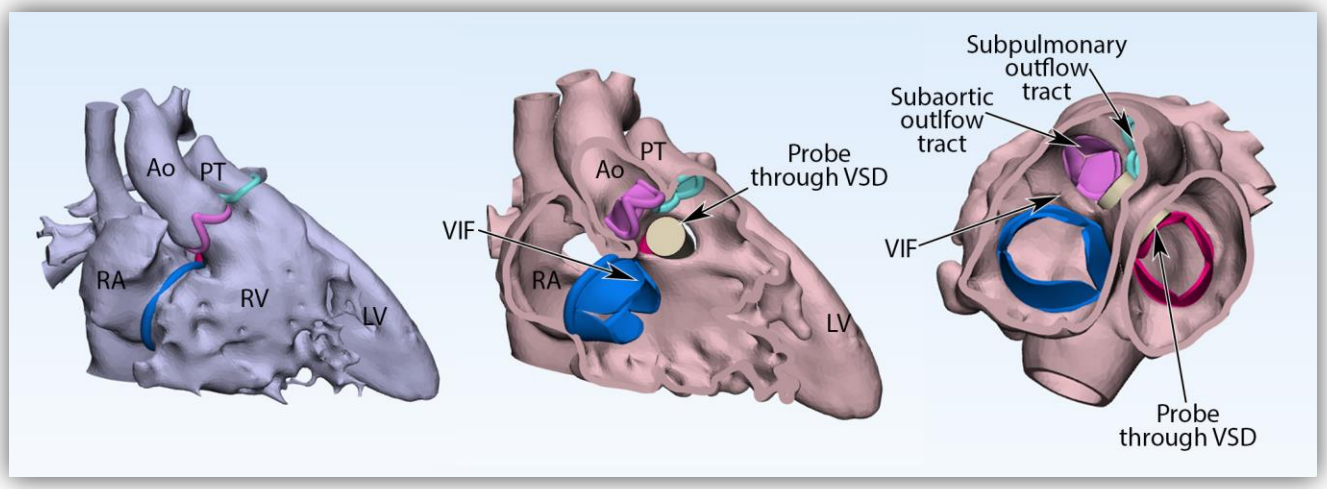
Surgical Consideration:

This heart has a large doubly-committed VSD and side-by-side relationship of the great arteries with the aorta on the right. The VSD appears to be more committed to the pulmonary artery, which reflects much larger pulmonary artery, and smaller aorta with some subaortic and aortic arch obstruction. A single coronary artery system arises from Sinus 1. The distal aortic arch is quite hypoplastic. Although it may be possible to create a baffle from the VSD to the aorta, the baffle probably obstruct the subpulmonary area. On the other hand, baffling from the VSD to the pulmonary artery seems quite straightforward. Therefore a similar strategy for Tassing-Bing anomaly would be an appropriate choice, although there is some room for intraoperative judgement. The aortic arch has to be augmented with a patch to match the size of the neo aortic root. The right ventricle may require some muscle resection. The neo pulmonary valve appears to be large enough although it is possible to require a transannular patch depending on the residual pressure gradient across the neo right ventricular outflow tract after repair.

***CASE 7*. DORV with a non-perimembranous subaortic VSD in the absence of outlet septum**

- ❖ Source images: Contrast-enhanced CT angiograms.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with a common subaortic and subpulmonary infundibulum without intervening outlet septum
- ♥ Side-by-side relationship of the arterial trunks with the right-sided aorta
- ♥ Non- perimembranous VSD involving outlet part of the septum, exclusively related to the aortic valve



- Both arterial trunks arise from the morphologically right ventricle.
- The arterial trunks show a side-by-side parallel spatial relationship with the aorta on the right.
- The right ventricular outflow tract is a common channel for right-sided subaortic and left-sided subpulmonary outflow tracts. There is no sizable outlet septum between the two outflow tracts.
- The VSD involves the outlet part of the ventricular septum. The VSD does not reach the tricuspid valve annulus leaving the VSD margin free of the atrioventricular conduction axis.
- The aortic valve is supported by a short muscular infundibulum, while the pulmonary valve is supported by a long infundibulum.
- This case shows that the absence of the outlet septum does not define the VSD to be a doubly-committed defect.

- ❖ *Compare this case with Case 6.*

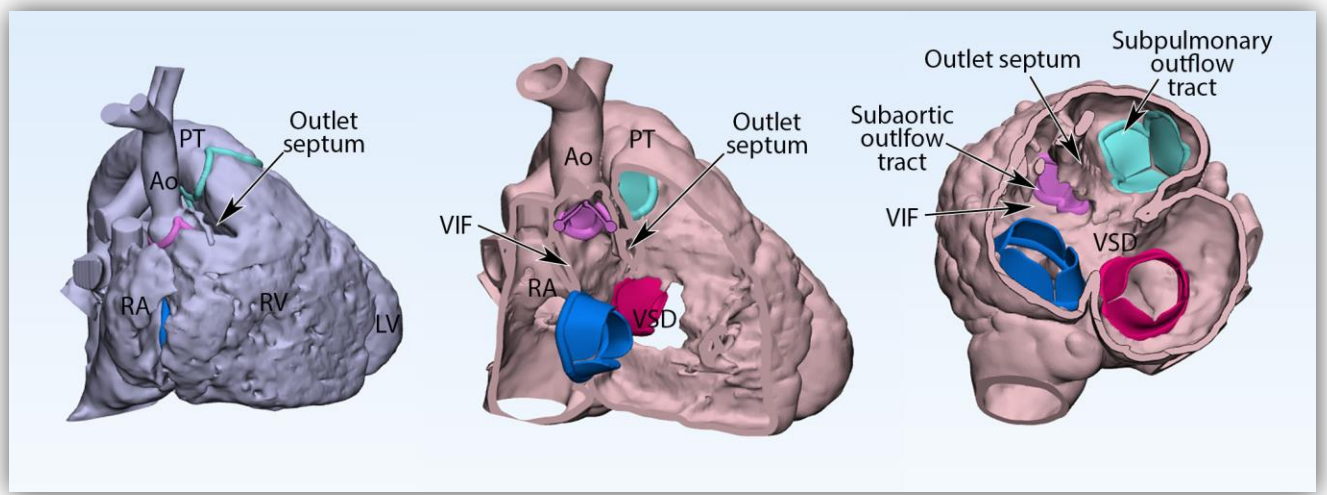
Surgical Consideration:

This is DORV with a common channel for subaortic and subpulmonary outflow tracts without intervening outlet septum. As the VSD is solely committed to the aortic valve, a rounded patch can be easily constructed from the ventricular septal crest to the anterior margin of the aortic valve annulus. Neither suaboartic nor subpulmonary outflow tract is expected to be narrow.

CASE 8. DORV with a remote non-committed VSD

- ❖ Source images: Contrast-enhanced MR angiograms.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with subaortic and subpulmonary infundibulum
- ♥ Side-by-side great arterial trunks with aorta on the right of the pulmonary arterial trunk
- ♥ Non-committed perimembranous VSD extending mostly toward the inlet part of the right ventricle
- ♥ Interrupted aortic arch after origin of the left subclavian artery



- Both aorta and pulmonary arterial trunk arise entirely from the morphologically right ventricle.
- The arterial trunks show a parallel spatial relationship with the aorta on the right.
- The right ventricular outflow tract is divided into the right-sided subaortic and left-sided subpulmonary outflow tracts by the outlet septum, which is exclusively a right ventricular structure above the VSD. Both aortic and pulmonary valves are supported by a long muscular infundibulum with a prominent ventriculoinfundibular fold (VIF).
- The VSD involves the ventricular septum along the septal leaflet of the tricuspid valve, characterizing it to be a perimembranous inlet VSD. The defect is remote from both aortic and pulmonary valves. The outlet septum is oriented in such a way to make the subaortic outflow tract mildly narrowed.
- The atrioventricular conduction axis is considered to course along the posteroinferior margin of the ventricular septal crest.

n *Compare this case with Cases 9-12.*

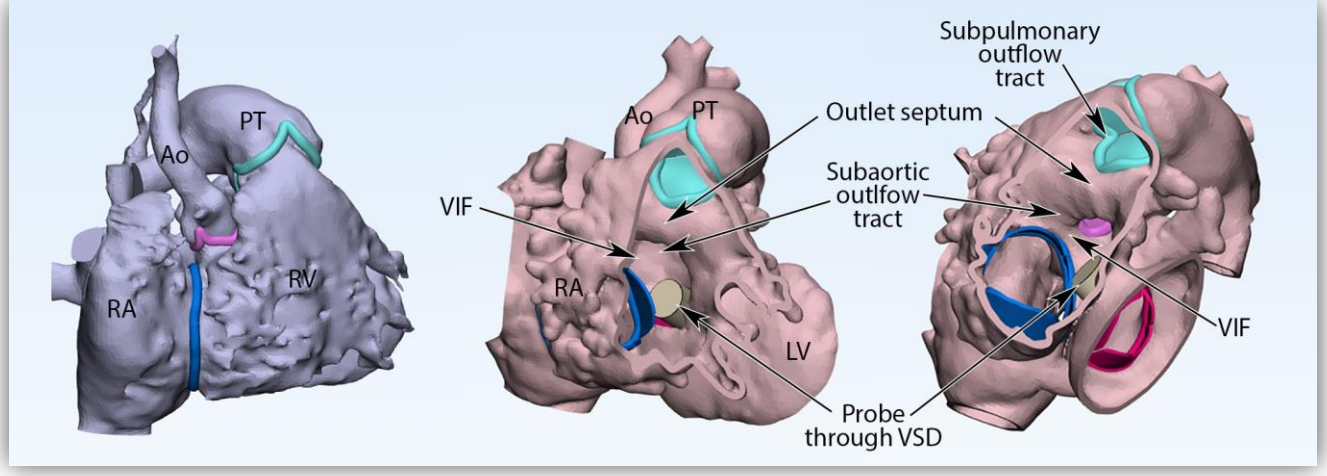
Surgical Consideration:

This is DORV with a perimembranous VSD that extends mainly toward the inlet of the right ventricle. The VSD is remote from both arterial valves not only because of its predominant inlet position but also due to long subaortic and subpulmonary infundibulum. A prominent outlet septum divides the subaortic and subpulmonary outflow tracts. Although the VSD is remote from the aortic valve, intraventricular baffling with extensive resection of the outlet septum will be feasible. The orifice size and function of the tricuspid valve would not be significantly compromised by the baffle. The volume of the functional part of the right ventricle after baffling would be large enough. The aortic arch should be augmented with a patch to match the size of the aortic root.

CASE 9. DORV with a remote VSD aligned with subaortic outflow tract

❖ Source images: Contrast-enhanced MR angiograms.

♥ Situs solitus / Levocardia / Concordant atrioventricular connection
♥ DORV with short subaortic and long subpulmonary infundibulum
♥ Normally related arterial trunks
♥ Remote perimembranous VSD extending mostly toward the inlet part of the right ventricle
♥ VSD aligned exclusively with the subaortic outflow tract
♥ Subaortic stenosis



- Both aorta and pulmonary arterial trunk arise entirely from the morphologically right ventricle.
- The arterial trunks show a normal relationship with the aortic root located right and posterior to the pulmonary arterial trunk.
- The right ventricular outflow tract is divided into the right posterior subaortic and left anterior subpulmonary outflow tracts by the large outlet septum, which is exclusively a right ventricular structure. The pulmonary valve is supported by a long infundibulum, while the aortic valve is supported by a short infundibulum. Note the ventriculoinfundibular fold (VIF) between the tricuspid valve annulus and the aortic and pulmonary valve attachments.
- The VSD abuts to the top of the tricuspid valve annulus on the septum, characterizing it to be a perimembranous VSD. The VSD mainly involves inlet part of the septum behind and below the attenuated upper part of the trabecula septomarginalis. Although the VSD involves the inlet septum and is not directly committed to either arterial valve, it is aligned exclusively with the subaortic outflow tract.
- The atrioventricular conduction axis is considered to course along the posteroinferior margin of the ventricular septal crest.
- The aorta shows severe tubular hypoplasia of its distal arch. The narrow aortic isthmus and restrictive patent ductus arteriosus form a confluence at the descending aorta with a shallow posterior shelf.

❖ *Compare this case with Cases 8 and 10. Also compare this case with Cases 2 and 3.*

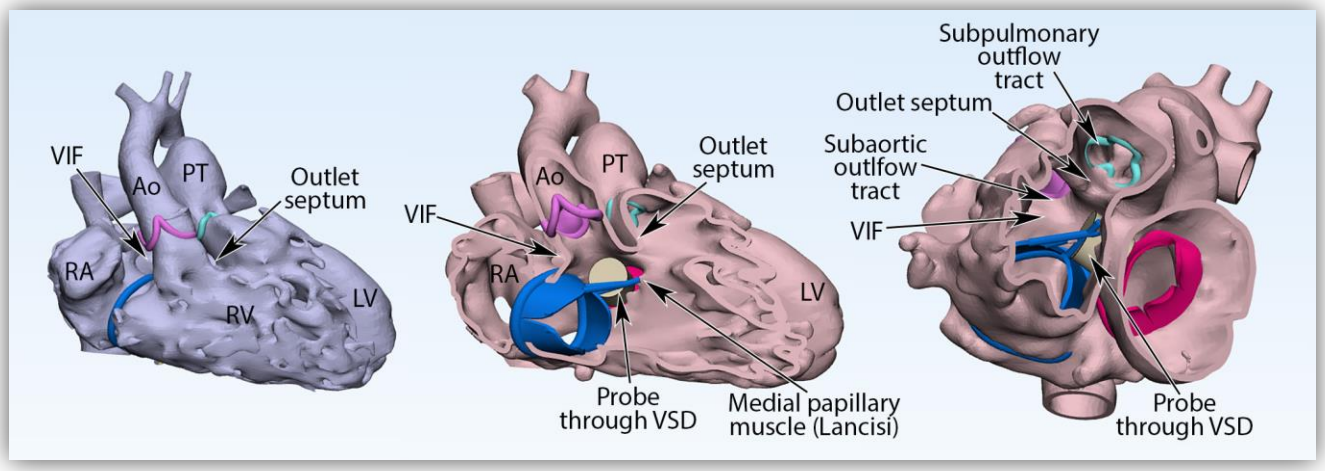
Surgical Consideration:

This heart has a large non-committed VSD and normally related great arteries with subaortic and aortic arch obstruction. The pulmonary trunk is more anterior and much larger than the aorta. The VSD is located in the inlet septum along the tricuspid valve and has significant trabecular extension. Although it is remote, there may be a pathway from the VSD to the aorta through a long subaortic outflow tract. Feasibility of biventricular repair would be dictated by quality and size of the aortic valve, and relationship between the intra-ventricular baffle and the tricuspid valve inflow. The alternative would be Norwood type procedure with DKS anastomosis and aortic arch reconstruction. Regarding tricuspid valve inflow, it appears to be enough inflow space even through an intra-ventricular baffle is created.

***CASE 10*. DORV with a VSD aligned with subaortic outflow tract at some distance from aortic valve**

- ❖ Source images: Contrast-enhanced MR angiograms.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with subaortic and subpulmonary infundibulum
- ♥ Parallel arterial trunks with the aorta located anterior and slightly right to the pulmonary arterial trunk
- ♥ VSD involving both the outlet and inlet parts of the right ventricle
- ♥ VSD is at a distance from the aortic valve due to a long subaortic infundibulum but aligned exclusively with subaortic outflow tract



- Both aorta and pulmonary arterial trunk arise entirely from the morphologically right ventricle.
- The arterial trunks show a parallel spatial relationship with the aorta located right and slightly anterior to the pulmonary arterial trunk.
- The right ventricular outflow tract is divided into the right anterior subaortic and left posterior subpulmonary outflow tracts by the outlet septum which is exclusively a right ventricular structure. Both aortic and pulmonary valves are supported by a long muscular infundibulum. A prominent ventriculoinfundibular fold (VIF) separates the aortic and tricuspid valve attachments.
- The VSD is a confluent defect involving both outlet and inlet parts of the ventricular septum with the posterior limb of the trabecula septomarginalis deficient. The medial papillary muscle of the tricuspid valve inserts to the left margin of the VSD. The outlet septum fuses to the left and superior margin of the VSD. As the aortic valve is at a distance from the VSD margin, one may describe this VSD a remote type. However, the VSD is exclusively aligned with the subaortic outflow tract.
- A thin muscular rim separates the VSD from the tricuspid valve annulus and therefore the VSD is not a perimembranous type. However, the atrioventricular conduction axis is considered to course close to the posteroinferior margin of the ventricular septal crest.

- ❖ *Compare this case with Cases 8 and 9. Also compare this case with Cases 2 and 3.*

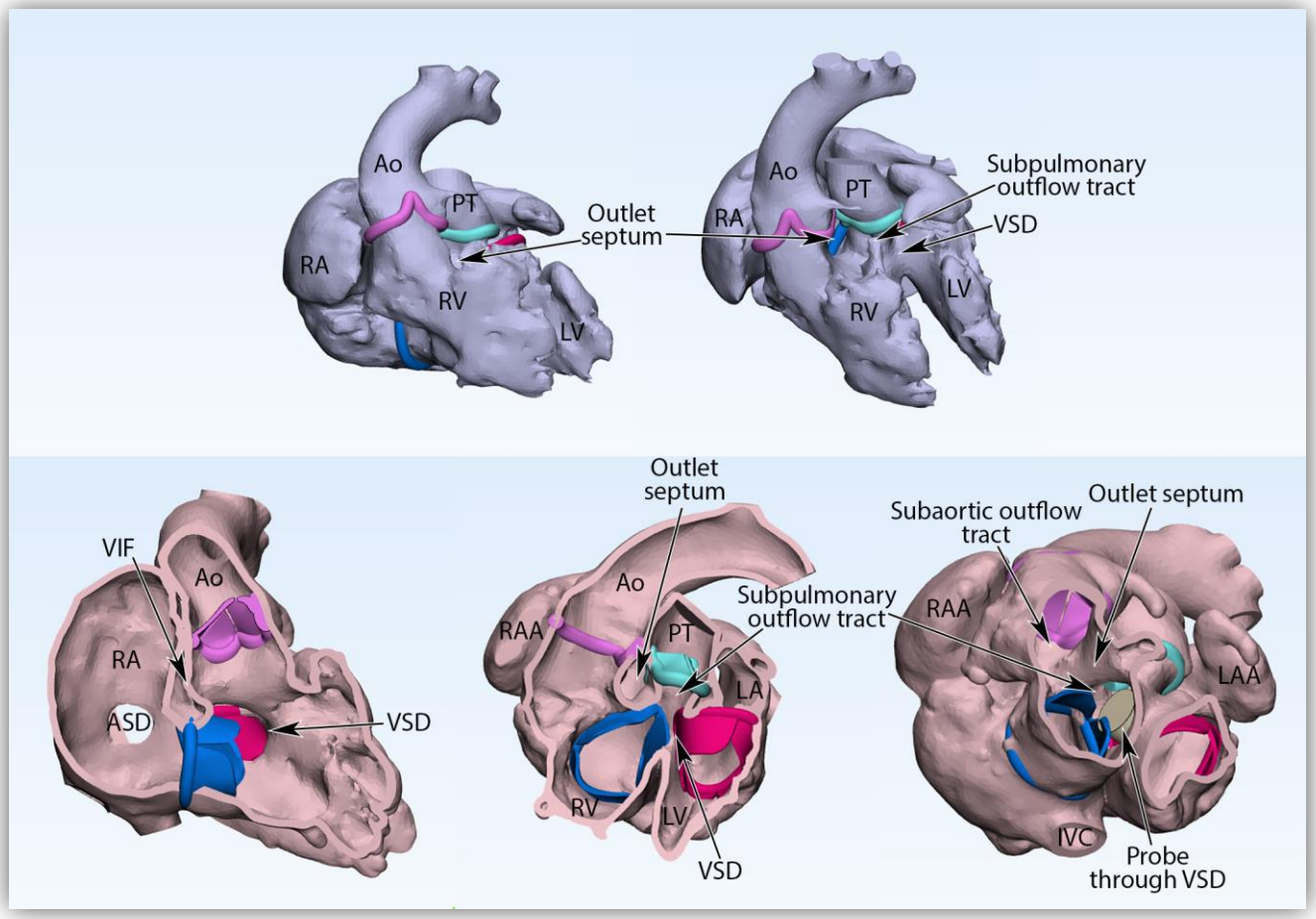
Surgical Consideration:

This is a 200% DORV with a side-by-side great artery arrangement where the aorta locates anterior and rightward to the pulmonary artery. Both outflow tracts are unobstructed although there is pulmonary valve stenosis. There is a large VSD in the perimembranous area, which is rather remote but relatively close to the aorta. This is going to be a long intra-ventricular baffle from the VSD to the anteriorly-located aorta, although there is no anatomic structure to preclude intra-ventricular baffling. As the medial papillary muscle and its chords overlay the lower part of the VSD, they should be dissected and reimplanted on the right ventricular aspect of the baffle. However, the baffle does not seem to obstruct the tricuspid valve inflow in a significant degree. The right ventricular outflow tract is unobstructed but the mid-right ventricular patch may be necessary if the intra-ventricular baffle bulges too much into the right ventricular mid cavity.

***CASE 11*. DORV with a remote VSD aligned with subpulmonary outflow tract**

- ❖ Source images: Contrast-enhanced MR angiograms.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with subaortic and subpulmonary infundibulum
- ♥ Parallel arterial trunks with the aorta located right and anterior to the pulmonary arterial trunk
- ♥ A perimembranous VSD extending mainly toward the inlet part and partly toward the outlet of the right ventricle
- ♥ VSD aligned exclusively to the subpulmonary outflow tract
- ♥ Subpulmonary stenosis



- Both arterial trunks arise from the morphologically right ventricle.
- The arterial trunks show a parallel relationship with the aortic root right and anterior to the pulmonary arterial trunk.
- The right ventricular outflow tract is divided into the anterior slightly right-sided subaortic outflow tract and the posterior and slightly left-sided subpulmonary outflow tract by the thick outlet septum, which is exclusively a right ventricular structure. The pulmonary valve is supported by the short infundibulum, while the aortic valve is supported by the long infundibulum. Note the ventriculoinfundibular fold (VIF) between the tricuspid valve annulus and the aortic and pulmonary valve attachments.
- The VSD involves mainly the inlet part and partly the outlet part of the septum with the posterior limb of the trabecula septomarginalis deficient. The VSD abuts to the top of the tricuspid valve annulus on the

septum, characterizing it to be a perimembranous VSD. Although the pulmonary valve is at a distance from the VSD margin, the VSD is exclusively committed to the subpulmonary outflow tract.

- The atrioventricular conduction axis is considered to course along the posteroinferior margin of the ventricular septal crest.
- The outlet septum is oriented parallel to the rest of the ventricular septum. The thick outlet septum encroaches on the subpulmonary outflow tract.

n *Compare this case with Cases 8-10 and 12. Also compare this case with Cases 4 and 5.*

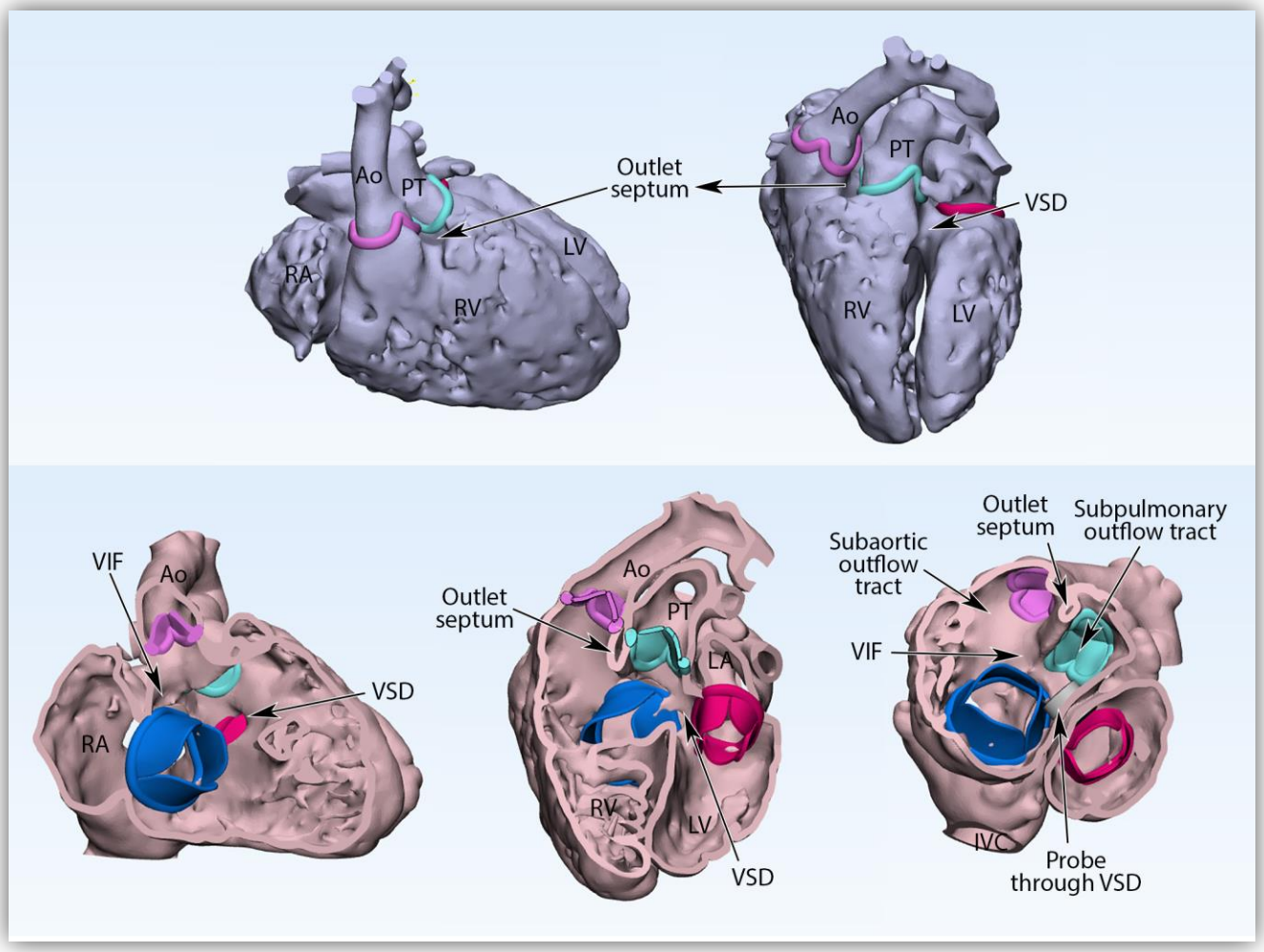
Surgical Consideration:

This heart has a complex problem of the presence of a perimembranous VSD involving the inlet part of the right ventricle. The VSD is related to the subpulmonary outflow tract. There is a parallel great artery arrangement with the aorta located anterior and rightward to the pulmonary artery. The subpulmonary outflow tract is narrow between the ventriculoinfundibular fold (VIF) and the hypertrophic outlet septum. It appears to be a single coronary system coming off from Sinus 1. The primary question is whether or not subpulmonary stenosis can be released effectively enough in order to make it the systemic outflow tract. Intracardiac exploration is essential in this case. If subpulmonary stenosis can be effectively released, the best option is to perform arterial switch operation and intra-ventricular baffling to the neo aortic root. If the subpulmonary outflow tract is deemed unsuitable as the systemic outflow tract, a potential option is to perform Nikaidoh procedure or double-root translocation procedure. The presence of the single coronary artery system may preclude aortic translocation since one of the coronary arteries that need to reach the other side of ventricle often loops around the aorta, which makes detachment of the aortic root impossible. The last option is to perform single ventricle palliation.

***CASE 12*. DORV with a VSD aligned with subpulmonary outflow tract at some distance from pulmonary valve**

- ❖ Source images: Contrast-enhanced MR angiograms.

- ♥ Situs solitus / Levocardia / Concordant atrioventricular connection
- ♥ DORV with subaortic and subpulmonary infundibulum
- ♥ Parallel arterial trunks with the aorta located right and anterior to the pulmonary arterial trunk
- ♥ Perimembranous VSD extending toward the outlet and inlet parts of the right ventricle
- ♥ VSD aligned exclusively with the subpulmonary outflow tract



- Both aorta and pulmonary arterial trunk arise entirely from the morphologically right ventricle.
- The arterial trunks show a parallel relationship with the aortic root right and anterior to the pulmonary arterial trunk.
- The right ventricular outflow tract is divided into the anterior slightly right-sided subaortic outflow tract and the posterior and slightly left-sided subpulmonary outflow tract by the outlet septum, which is exclusively a right ventricular structure. The pulmonary valve is supported by the short infundibulum, while the aortic valve is supported by the long infundibulum. Note the ventriculoinfundibular fold (VIF) between the tricuspid valve annulus and the aortic and pulmonary valve attachments.
- The VSD is a confluent defect involving both outlet and inlet parts of the ventricular septum with the posterior limb of the trabecula septomarginalis deficient. The VSD abuts to the top of the tricuspid valve

annulus on the septum, characterizing it to be a perimembranous VSD. Although the pulmonary valve is at a distance from the VSD margin, the VSD is exclusively aligned with the subpulmonary outflow tract.

- The atrioventricular conduction axis is considered to course along the posteroinferior margin of the ventricular septal crest.
- The outlet septum is oriented parallel to the rest of the ventricular septum.
- The aorta shows mild tubular hypoplasia of its distal arch.

❖ *Compare this case with Cases 8-10 and 11. Also compare this case with Cases 4 and 5.*

Surgical Consideration:

This is a 200% DORV with a parallel relationship of the great arteries with the aorta on the right. There is a moderate-sized VSD in the perimembranous area, which is closer to the pulmonary artery. There is a quite distance between the VSD and the aorta. Although it is going to be a relatively long pathway to get the pulmonary artery from the VSD, baffling appears to be feasible. Therefore, a proposed procedure would be arterial switch operation and intra-ventricular baffling to the neo aortic root. A baffle does not seem to obstruct the tricuspid valve inflow or mid cavity of the right ventricle.

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