Essential Modifiers of Double Outlet Right Ventricle
Revisit With Endocardial Surface Images and 3-Dimensional Print Models

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Abstract —Hearts with double outlet right ventricle are a heterogeneous group of malformations in which a comprehensive diagnostic approach is required for tailored surgical management. This pictorial essay revisits essential modifiers of clinical and surgical importance in management of the patients with double outlet right ventricle using 3-dimensional volume-rendered endocardial surface images and 3-dimensional print models. Special emphasis is paid to the infundibular morphology, including the size and orientation of the outlet septum, relative to the margin of the ventricular septal defect, and the extent of the muscular infundibulum as an additional modifier of the distance between the ventricular septal defect margin and the arterial valve or valves. **(*Circ Cardiovasc Imaging*. 2018;11:e006891. DOI: 10.1161/CIRCIMAGING.117.006891.)**

Key Words: double outlet right ventricle ■ endocardium ■ heart septal defects, ventricular ■ printing, three-dimensional ■ ventricular septum

Double outlet right ventricle (DORV) is a type of ventriculoarterial connection in which both great arteries arise either entirely or predominantly from the right ventricle.[1–3] In defining in such a way, DORV is a descriptor of 1 particular aspect of the malformation. In segmental approach to congenital heart diseases, there are 3 facets to define: the morphology, the connections, and the relationships.[4] Although DORV defines the connection at the ventriculoarterial junction, it does not account for morphology and relationships. Hearts with DORV are an extremely heterogeneous group showing various morphological features, connections, and relationships at each level of the cardiac segments and intersegmental junctions.[2,3,5] As a consequence, the clinical manifestations and required surgical procedures for patients with DORV are variable. Accordingly, a comprehensive diagnostic approach is required in the assessment of the patients showing DORV.

Transthoracic echocardiography is the first-line investigation in the diagnosis of DORV and depiction of the surgical anatomy and associated abnormalities. In the majority of cases of DORV, transthoracic echocardiography provides sufficient information for surgical decision making and planning.[6] When the echocardiographic information is insufficient for this purpose, complementary cross-sectional imaging with computed tomography or magnetic resonance is performed. Contrast-enhanced computed tomography or magnetic resonance angiograms acquired in 3-dimensional volume allow for endocardial surface representation and 3-dimensional printing of physical replicas of the heart for further anatomic assessment and surgical planning, as well as surgical training and practice (Figure 1).[7,8] In addition, endocardial surface representation on a computer screen or 3-dimensional print models can be used for morphology-imaging correlation. This pictorial essay outlines the essential and controversial issues in the assessment of DORV with the aid of virtual and physical reconstruction of the heart using 3-dimensional magnetic resonance and computed tomographic data. Although DORV occurs with any combination of visceroatrial situs and atrioventricular connection, we limit our discussion to the cases occurring in the setting of situs solitus, levocarida, and concordant atrioventricular connection.

Essential Morphological Modifiers in DORV

As discussed, the hearts with DORV have no phenotypic uniformity.[5] There are several modifiers that are responsible for the large spectrum of heterogeneity of hearts with DORV. In almost all cases with DORV, a defect in the ventricular septum is an essential component of the malformation. Although it has been suggested that the defect in the ventricular septum should be called interventricular communication, we use the conventional and most widely used term ventricular septal defect (VSD) because of the reasons listed at the end of this

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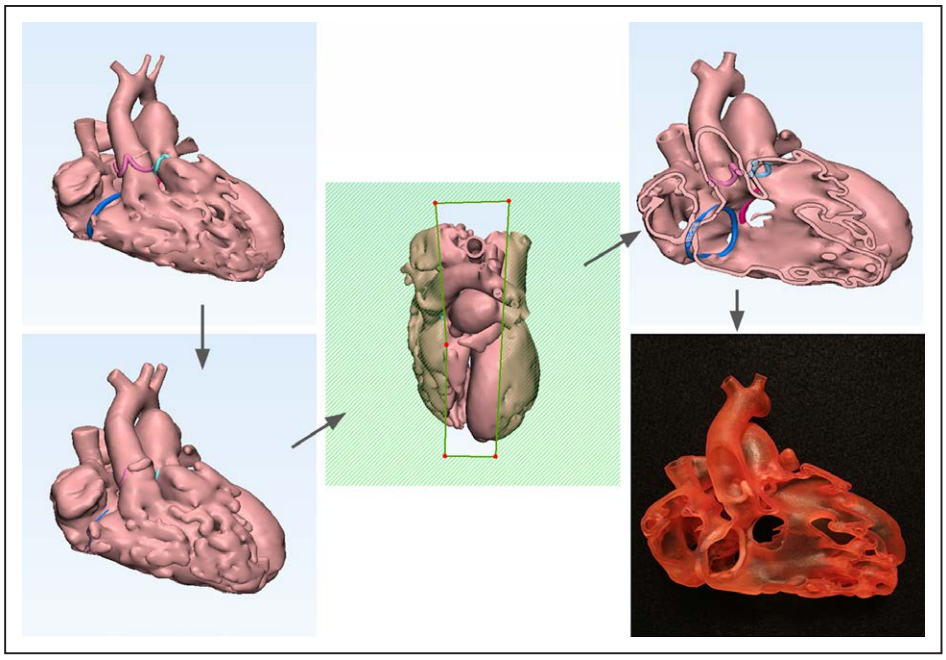

Figure 1. Process of endocardial surface representation and corresponding 3-dimensional print model in a case with double outlet right ventricle. A cast of the blood pool was segmented from electrocardiographically gated and respiration-navigated magnetic resonance angiograms (**top left**). The cardiac valve attachment sites are marked with thin bands. With computer graphic tool called hollow, a shell with a thickness of 1.7 mm was created (**left bottom**). The model is rotated, and the unnecessary parts are removed (**center**) to show the endocardial surface anatomy (**right top**). The model was saved as a standard tessellation language file, and a physical model was printed as shown in **right bottom**. In this endocardial surface representation, the outer surface does not represent the true anatomy.

paper. The VSDs in hearts with DORV have long been classified into 4 groups: subaortic, subpulmonary, doubly committed, and noncommitted or remote VSDs; after the seminal publication by Lev et al[9] in 1972 (Figure 2; Data Supplements 2A-2C). Although this convention seems simple and intuitive and has been widely adopted, it does have limitations

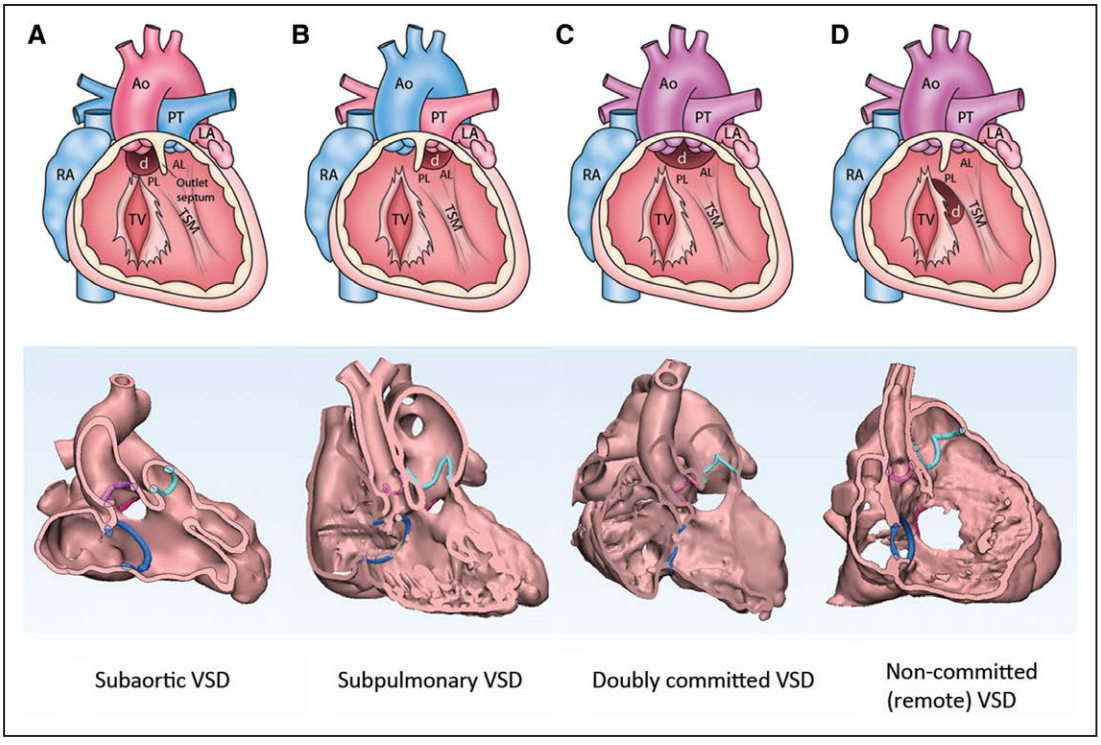

Figure 2. Lev et al's[9] classification of ventricular septal defects (VSDs) in double outlet right ventricle. The subaortic (**A**), subpulmonary (**B**), and doubly committed (**C**) VSDs classically involve the outlet part of the septum and therefore cradled between the anterior and posterior limbs of the trabecula septomarginalis (TSM). The classic noncommitted or remote VSD (**D**) involves the inlet part of the ventricular septum behind and below the posterior limb of the TSM. AL indicates anterior limb; Ao, aorta; d, ventricular septal defect; LA, left atrium; PL, posterior limb; PT, pulmonary trunk; RA, right atrium; and TV, tricuspid valve.

Table. List of Essential Modifiers of Surgical Anatomy of Double Outlet Right Ventricle

Features	Primary	Secondary
Relationship of the atrioventricular conduction axis to the VSD margin	Perimembranous VSD Nonperimembranous 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 3 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[9] classification: Subaortic Subpulmonary Doubly committed Noncommitted 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 Noncommitted VSD type AVSD
Atriovenricular valve abnormalities	Stenosis of the tricuspid or mitral valve Straddling or over-riding 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	

AEPC indicates Association of European Pediatric Cardiology; AVSD, atrioventricular septal defect; DORV, double outlet right ventricle; EACTS, European Association of Cardio-Thoracic Surgery; STC, Society of Thoracic Surgeons; TGA, transposition of the great arteries; and VSD, ventricular septal defect.

in fulfilling the requirements for surgical evaluation.[1] The important modifiers of clinical and surgical importance in patients with DORV include (Table):

1. Relationship of the atrioventricular conduction axis to the margin of the VSD (perimembranous versus non-perimembranous VSD)

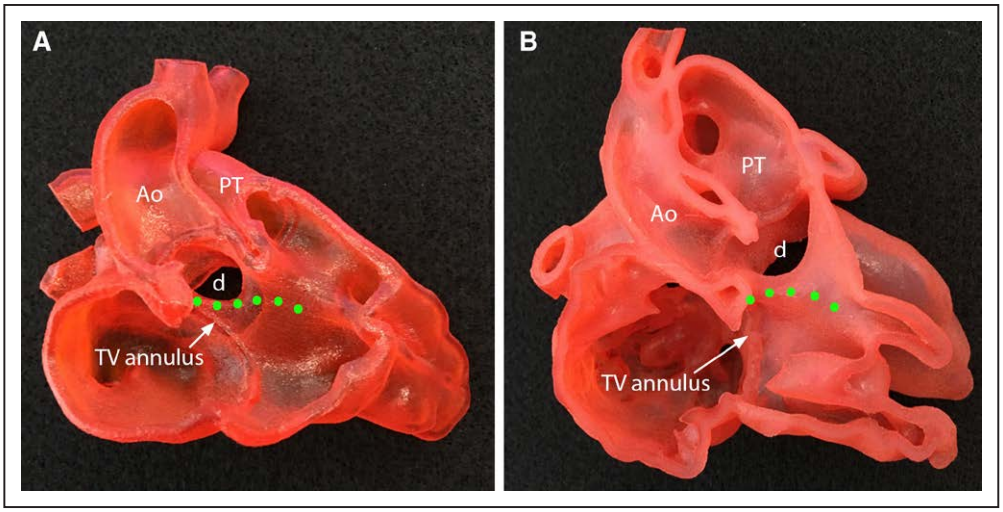

Figure 3. Perimembranous vs nonperimembranous ventricular septal defect (VSD). **A**, Perimembranous VSD extending predominantly toward the outlet of the right ventricle. **B**, Doubly committed VSD involving the outlet part of the ventricular septum. The perimembranous VSD in (**A**) shows direct contact of its posterior margin to the tricuspid valve (TV) annulus at the superior aspect of the septal leaflet while the doubly committed VSD in (**B**) is at a distance from the TV annulus. Note the expected course of the atrioventricular conduction axis (dotted line) in each case. Ao indicates aorta; d, ventricular septal defect; and PT, pulmonary trunk.

2. Location of the VSD in the ventricular septum seen from the right ventricle, and spatial relationship the VSD to the annulus along the septal leaflet of the tricuspid valve
3. Size of the VSD; unrestrictive versus restrictive
4. Orientation of the outlet septum in relation to the VSD margin
5. Presence and extent of the muscular infundibulum
6. Great arterial relationship
7. Presence or absence of aortic or pulmonary outflow tract obstruction
8. Relationship of the VSD to the arterial valves (Lev et al's[9]) and type of DORV (classification of Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery–Association for European Paediatric and Congenital Cardiology)[1]
9. Atrioventricular valve abnormalities
10. Ventricular cavity sizes
11. Other associated abnormalities

Perimembranous Versus Nonperimembranous VSD

The relationship of the VSD margin with the membranous septum is important because the atrioventricular conduction axis courses along the posteroinferior margin of the membranous septum regardless of whether it is intact or defective.[10] VSDs involving the membranous septum and the adjacent muscular septum are called perimembranous VSDs in which the atrioventricular conduction axis courses along the posteroinferior margin of the VSD. When the VSD does not involve the membranous septum (nonperimembranous VSDs), the atrioventricular conduction axis courses along the intact membranous septum at a distance from the margin of the VSD. In defining whether the VSD is perimembranous or nonperimembranous, it is worthwhile to note that the membranous septum is the septal extension of the central fibrous body that normally exists in the triangular space between the tricuspid, mitral, and aortic valves. When neither aortic nor pulmonary valve completes this triangular space because of the intervening muscular infundibulum, the central fibrous body keeps its location between the anterosuperior aspects of the tricuspid and mitral valve annuli. In imaging and pathological observation, the perimembranous or nonperimembranous nature of the defect can be determined by observing the

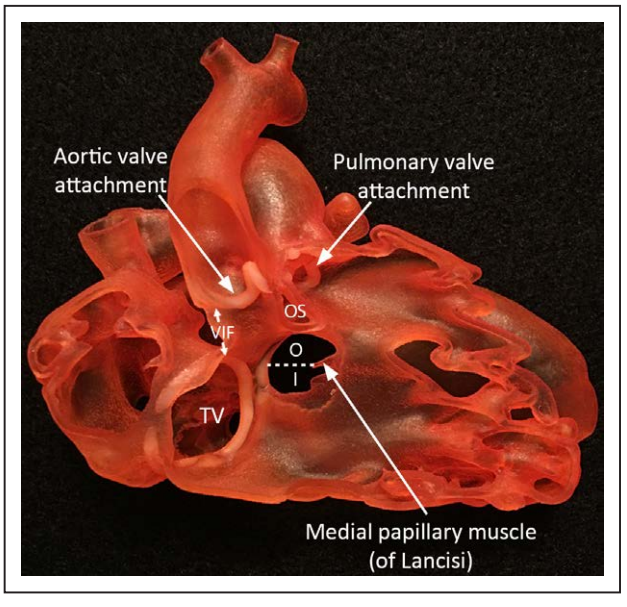

Figure 4. Double outlet right ventricle with a perimembranous ventricular septal defect (VSD) extending toward the inlet and outlet. A dotted line connecting the top of the tricuspid valve annulus and the medial papillary muscle of Lancisi demarcates the inlet (I) and outlet (O) components of the VSD. The VSD is at some distance from the aortic valve with a long subaortic muscular infundibulum. Note that the ventriculoinfundibular fold (VIF) separates the aortic and tricuspid valves. The VSD is aligned with the subaortic outflow tract with the outlet septum (OS) inserted to the left superior margin of the VSD. TV indicates tricuspid valve.

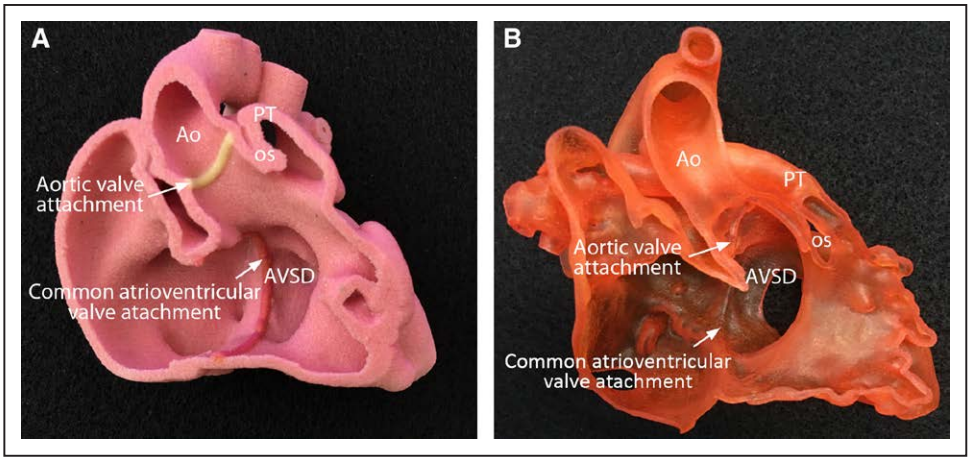

Figure 5. Two cases showing double outlet right ventricle (DORV) with an atrioventricular septal defect (AVSD) and subpulmonary outflow tract obstruction. **A**, AVSD remote from the semilunar valves. The defect is confined mostly to the inlet part of the ventricular septum. However, the defect is aligned with the subaortic outflow tract. This case was associated with right isomerism. **B**, AVSD extending toward the aortic valve (tetralogy type of DORV with AVSD). The cranial extent of the AVSD reaches the aortic valve. Both cases show subpulmonary outflow tract obstruction because of deviated outlet septum (OS). The outlet septum is free floating at a distance above the AVSD in (**A**) while it is attached to the left margin of the cranially extended part of the AVSD in (**B**). Ao indicates aorta; and PT, pulmonary trunk.

presence or absence of direct contact of the defect margin to the tricuspid valve annulus along the most cranial aspect of its septal leaflet (Figure 3).

Location of the VSD in the Ventricular Septum

Because the surgical repair of DORV is almost always performed from the right side of the heart, the location of the VSD

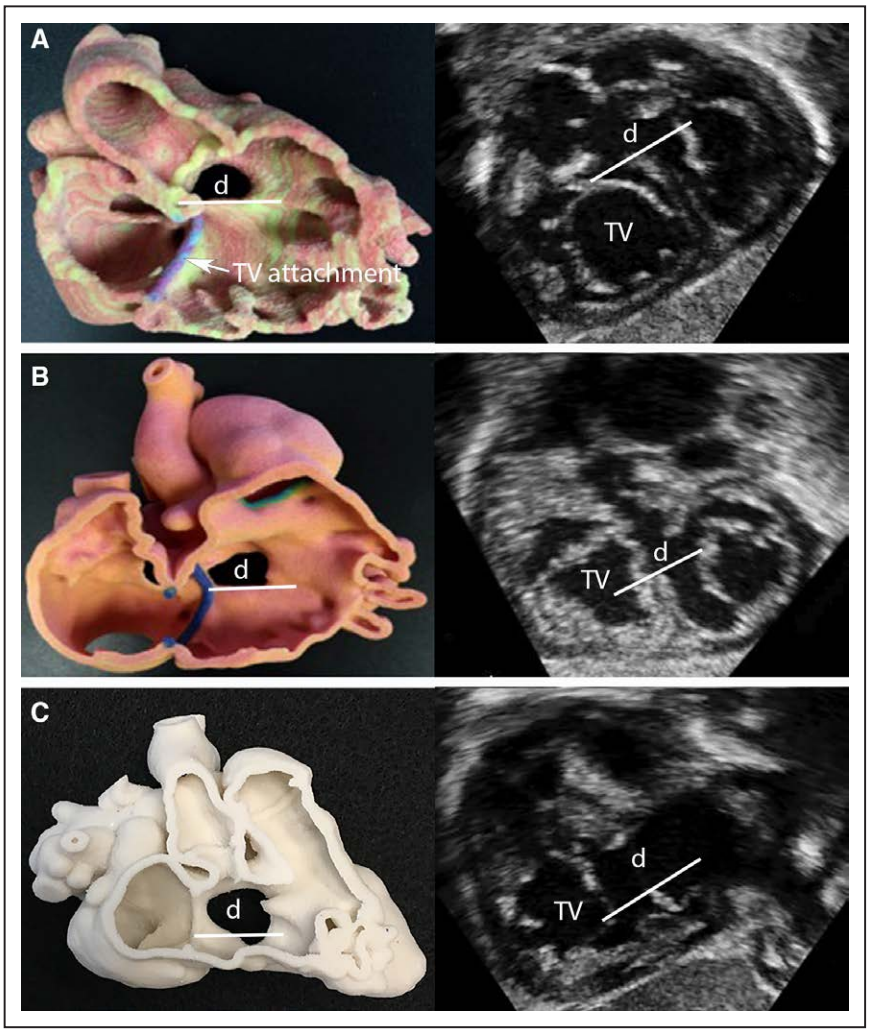

Figure 6. Relationship between the ventricular septal defect (VSD) and septal leaflet of the tricuspid valve (TV). Three different cases showing the VSD not directly related to the tricuspid valve attachment (**top**; **A**), the VSD related to the upper one third of the tricuspid valve attachment (**middle**; **B**), and the VSD related to the two thirds of the tricuspid valve attachment (**bottom**; **C**). d indicates ventricular septal defect.

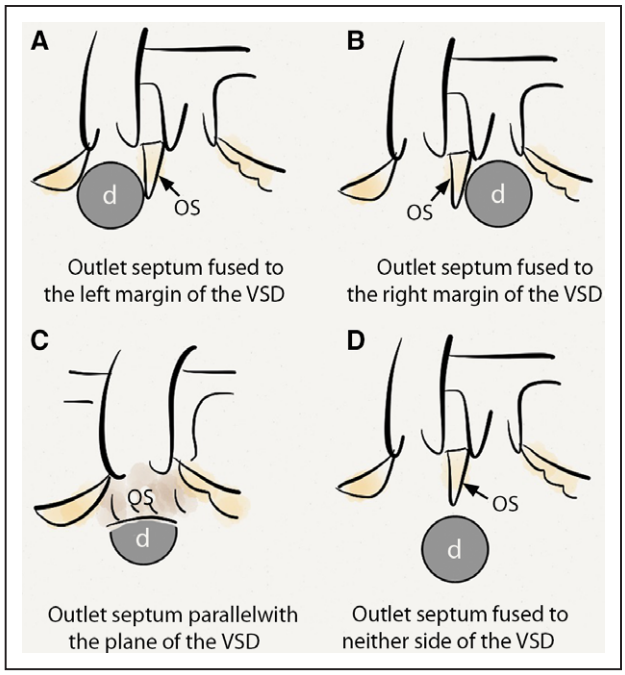

Figure 7. Orientation of the outlet septum (OS) in relation to the margin of the ventricular septal defect (VSD). d indicates ventricular septal defect.

is described as it is seen from the right ventricle. The VSD commonly involves the outlet part of the septum between the anterior and posterior limbs of the trabecula septomarginalis (Figure 2A–2C; Data Supplements 2A-2C). Less frequently, the VSD involves predominantly the inlet part of the septum along the septal leaflet of the tricuspid valve (Figures 2D). However, the location of the VSDs is not limited to these classic locations. Not infrequently, the VSD involves both outlet and inlet parts of the septum with the posterior limb of the trabecula septomarginalis being deficient (Figure 4; Data Supplements 4A-4C). Rarely, the VSD occurs in the apical trabecular part of the septum. DORV may also occur with atrioventricular septal defect that often extends toward one or both outflow tracts (Figure 5).

Extent of the Tricuspid Valve in Proximity to the VSD

Intraventricular baffling of the VSD to an arterial valve may compromise the size and function of the tricuspid valve when the VSD involves the inlet part of the ventricular septum. Therefore, it is important to define the extent of direct contact or proximity between the septal leaflet of the tricuspid valve and the VSD margin (Figure 6; Data Supplement 6A-6C).

Size and Multiplicity of the VSD

The VSD is the only or at least the major outlet of the left ventricle in DORV. Usually, the VSD is single and large with its diameter greater than the age-matched diameter of the normal aortic valve. When the VSD is smaller than the size of the normal aortic valve, the VSD is regarded as restrictive.[11–13] Not infrequently, additional muscular VSDs can be found in the apical trabecular septum. Exceedingly rarely, no VSD is identifiable. Those cases are considered to have the VSD spontaneously closed in fetal or postnatal life.[3]

Orientation of the Outlet Septum

The outlet septum is exclusively a right ventricular structure in DORV.[2,3] It is usually perpendicular to the ventricular septum that is fused to the right or left margin of the VSD when the defect involves predominantly the outlet part of the septum (Figures 2A, 2B, 7A, and 7B).[14] When the outlet septum is fused to the left margin of the VSD, the VSD is solely committed to the right-sided arterial trunk, which is usually the aorta (Figures 2A and 7A). When the outlet septum is fused to the right margin of the VSD, the VSD is committed solely to the left-sided arterial trunk, which is usually the pulmonary trunk (Figures 2B, 7B, and 8A; Data Supplement 8A). Less frequently, the outlet septum can be parallel with the ventricular septum without attachment to the right or left margin of the VSD. When the VSD involves predominantly the outlet part of the septum and the outlet septum is parallel with the ventricular septum, the VSD is committed to the posteriorly located arterial valve (Figures 7C and 8B; Data

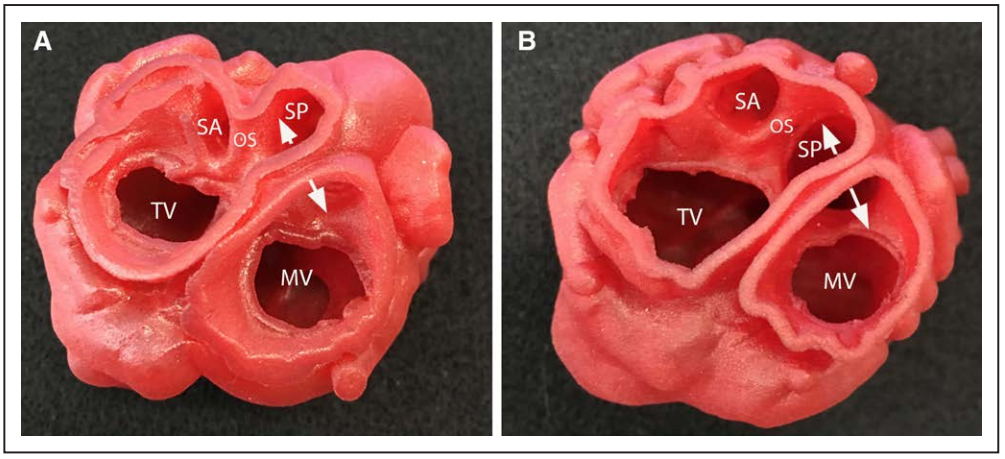

Figure 8. Double outlet right ventricle (DORV) with subpulmonary ventricular septal defect (VSD; so-called Taussig–Bing malformation) with different orientation of the outlet septum. **A**, DORV with the outlet septum (OS) fused to the right margin of the VSD leaving the VSD exclusively committed to the left-sided pulmonary valve. **B**, DORV with the outlet septum parallel to the rest of the ventricular septum without fusion to either side of the VSD, leaving the VSD exclusively committed to the posteriorly located pulmonary valve. Double-headed arrows are through the VSD. MV indicates mitral valve orifice; SA, subaortic outflow tract; SP, subpulmonary outflow tract; and TV, tricuspid valve orifice.

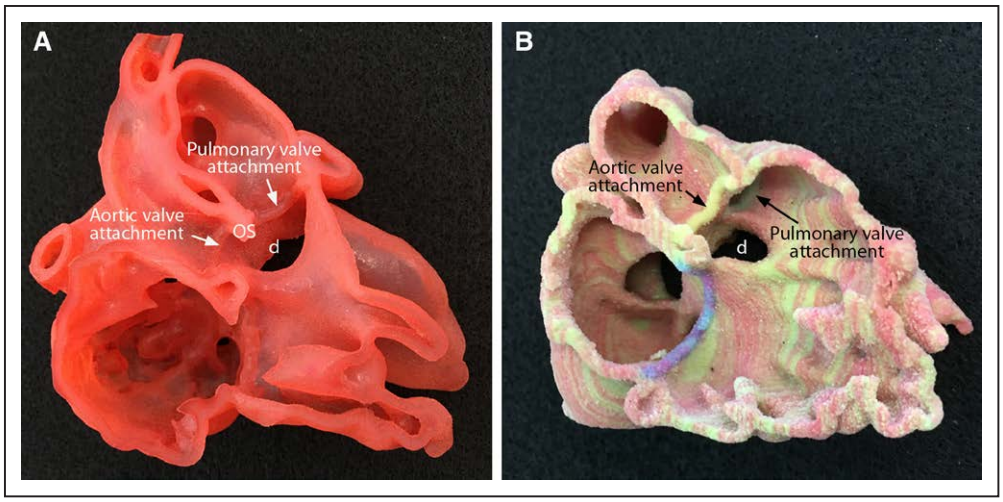

Figure 9. Deficient outlet septum vs double commitment of the ventricular septal defect (VSD) to the arterial valves. **A**, In this case with a doubly committed VSD (d), small free-floating outlet septum (OS) is seen above the VSD. **B**, In this case with deficient outlet septum having the aortic and pulmonary valves in direct contact, the VSD is located exclusively below the aortic valve. Deficient outlet septum does not always define that the VSD is doubly committed.

Supplement 8B). When the VSD involves the inlet part of the septum, the lower margin of the outlet septum seems free standing in the right ventricular outlet (Figures 2D and 7D).

Infrequently, the outlet septum is deficient or vestigial. The absence of the outlet septum with the 2 arterial valves in fibrous continuity has been regarded as the key finding of the doubly committed VSD[2] (Figure 2C). However, in cases where the outlet septum is completely absent or vestigial, the VSD is often not equally committed to the aortic and pulmonary valves. Rather, VSD commitment is a continuum and may even be solely committed to the aortic valve or less frequently to the pulmonary valve (Figure 9B). On the contrary, the presence of the outlet septum does not preclude the diagnosis of a doubly committed VSD (Figure 9A). The extent of the outlet septum and the VSD commitment to an arterial valve or valves are 2 independent morphological features.

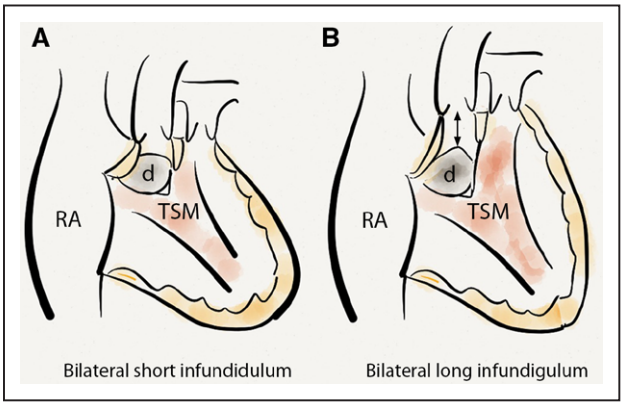

Figure 10. Extent of muscular infundibulum versus distance between the ventricular septal defect (VSD) and semilunar valves. The VSD is aligned with the aortic valve in both cases. The VSD is close to the aortic valve in (**A**) while it is remote from the aortic valve in (**B**). **B**, the VSD is at a distance from the aortic valve because its subaortic infundibulum (double-headed arrow) is much longer than the infundibulum shown in (**A**). Similar difference can be appreciated between the cases shown in Figures 3A and 4. d indicates ventricular septal defect; RA, right atrium; and TSM, trabecula septomarginalis.

Extent of Muscular Infundibulum

The aortic or pulmonary valve is defined to be supported by a muscular infundibulum or simply the infundibulum or conus when the whole circumference of the outflow tract is a muscular structure. The presence of the infundibulum implies that the aortic or pulmonary valve is separated from the tricuspid valve by the intervening muscle called the ventriculoinfundibular fold. The muscular infundibulum pushes its supporting semilunar valve forward and upward away from the tricuspid valve. The extent of the infundibulum also contributes to the remoteness of the VSD from the semilunar valve. For a given location of the VSD in relation to the trabecula septomarginalis and the tricuspid valve, the longer the infundibulum, the further the VSD from the semilunar valve (Figure 10).[3]

Remoteness of the VSD From the Arterial Valves

Noncommitment or remoteness of the VSD to the arterial valves is perhaps the most controversial issue not only in the literature but also in our daily practice. The defects involving the inlet or trabecular parts are regarded as noncommitted[9] or remote[15] VSDs. Because it is an arbitrary distinction between the committed and noncommitted VSDs in borderline cases, it was suggested that the VSD should be defined as noncommitted when the distance between the VSD and both aortic and pulmonary valve is greater than the age-matched aortic valve diameter.[16,17] Although this definition seems clear, it should be understood that the arterial valve can be at a significant distance from the upper margin of the VSD involving the outlet part of the septum if there is an excessively long infundibulum (Figure 10B).[3,18,19] On the contrary, the VSD involving predominantly the inlet part of the septum can be close to an arterial valve if the arterial valve is not supported by a substantial muscular infundibulum. Therefore, the remoteness of the VSD from the arterial valves is determined not only by the location of the VSD within the septum but also by the extent of

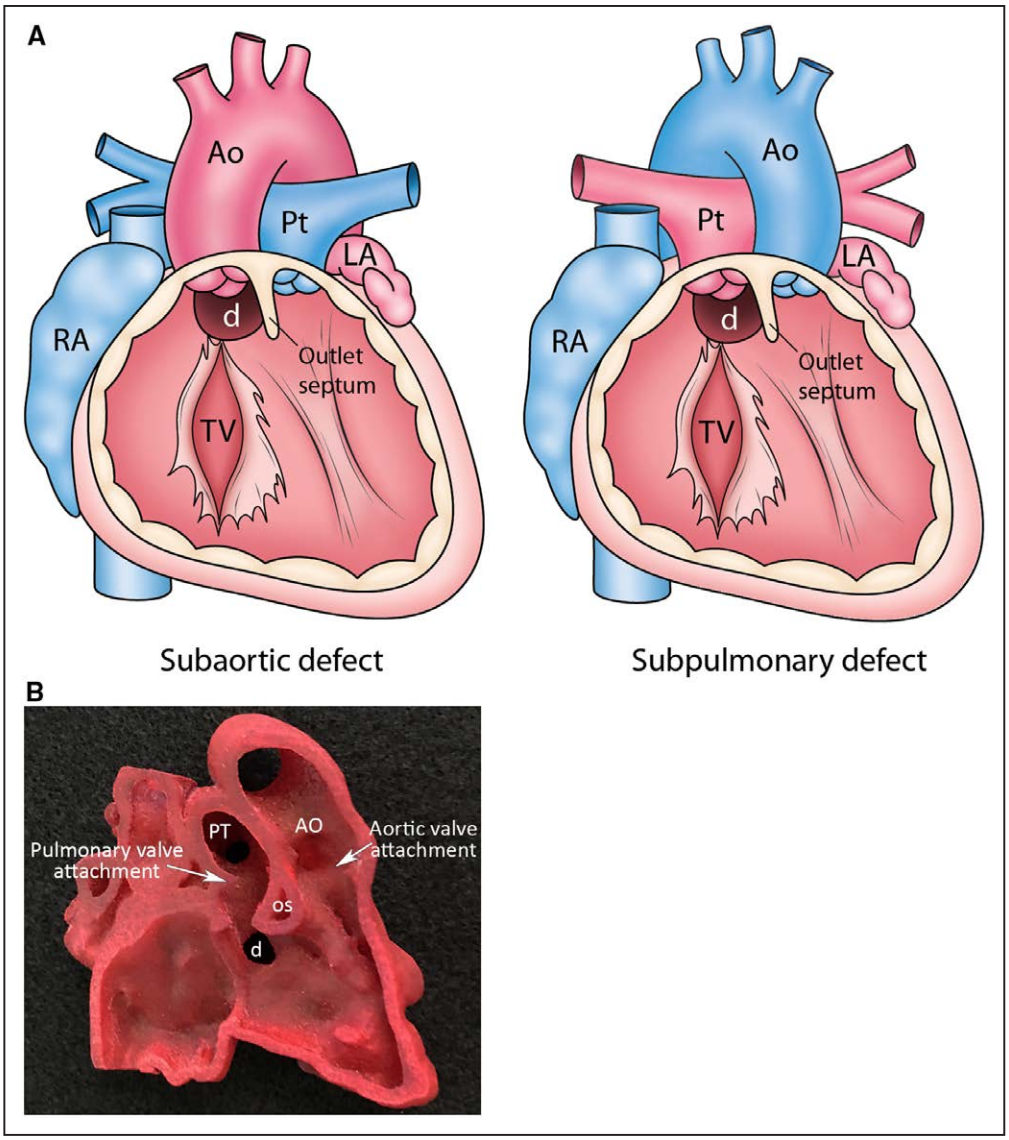

Figure 11. Great arterial relationship and ventricular septal defect (VSD). **A**, Illustrations showing the same intracardiac anatomy and flipped position of the great arterial trunks. The VSD commitment to the semilunar valve in left hand and right hand is opposite. **B**, Three-dimensional print model showing the VSD location similar to Figure 9B. It is committed to the right-sided pulmonary arterial trunk (PT). Ao indicates aorta; d, ventricular septal defect; LA, left atrium; OS, outlet septum; RA, right atrium; and TV, tricuspid valve.

the infundibulum. In addition, it should be taken into account that the VSDs that are committed to an arterial valve by anatomic definition may be rendered surgically noncommitted when there are the factors that do not allow baffling the VSD to an arterial valve.[2,3,18] Furthermore, it is important that the remoteness of the VSD to the arterial valves does not preclude intraventricular baffling. Therefore, so-called noncommitted or remote VSDs should be further divided into subcategories according to alignment of the VSD to the subaortic and subpulmonary outflow tracts.

Great Arterial Relationship

Two hearts showing almost identical intracardiac anatomy would present completely differently if the great arterial relationship is reversed. DORV showing otherwise typical intracardiac anatomy for cases with subaortic VSD would have the VSD in a subpulmonary location if the great arterial relationship is reversed from the usually right-sided aortic location (Figure 11). Conversely, DORV showing otherwise typical intracardiac anatomy for cases with a subpulmonary VSD will have the VSD in a subaortic location if the great arterial relationship is reversed from the usually right-sided aortic location.

Ventricular Outflow Tract Obstruction

When the VSD is committed to an arterial valve, the outflow tract leading to the noncommitted arterial valve tends to be narrowed between the outlet septum and the free wall of its infundibulum. In other words, subpulmonary outflow tract obstruction is common in DORV with subaortic VSD while subaortic outflow tract obstruction with or without obstructive lesions of the aortic arch is common in DORV with a

subpulmonary VSD. However, there are ample exceptions to this general tendency.

Right and Left Ventricular Cavity Volumes

Biventricular repair of DORV requires adequate volumes of both ventricles after surgery. Intraventricular baffling or tunneling compromises the right ventricular volume as a part of the right ventricle is incorporated into the left ventricular outflow tract. Therefore, it is important to assess the right ventricular cavity size and estimate the volume of the remaining right ventricle after the intended surgery.

Other Issues of Surgical Importance

DORV often occurs with anomalous systemic or pulmonary venous connections, especially when there is visceral heterotaxy with right or left isomerism. As a form of conotruncal abnormalities, left juxtaposition of the atrial appendages is not uncommon. When arterial switch operation is required, coronary arterial origins and proximal courses should be recorded.

Ventricular Septal Defect Versus Interventricular Communication

Recently, it has been suggested that the channel between the ventricles that has been called a VSD should be named an interventricular communication presuming that most cardiac imagers and surgeons define the VSD as the plane for application of a surgical patch or baffle to reconnect the aortic root with the left ventricle.[2,3,6,20] As far as we know, this is not the case of most institutions as well as our own. We describe the plane that is proposed to be called the interventricular communication simply and unanimously using the common term, VSD. We do not define the plane for surgical patch or baffle with any name but describe how the VSD could be baffled to the aorta or pulmonary trunk with or without enlarging the VSD or resecting all or a part of the outlet septum. By defining the VSD in such a way, the outlet septum never forms a margin of the VSD because it is exclusively a right ventricular structure in DORV, as well as in tetralogy of Fallot. What we call VSD is identical to what is called interventricular communication by others.[2,3,6,20]

Disclosures

Dr Yoo is the owner of a 3-dimensional (3D) printing company 3D Human Organ Printing and Engineering Medical and the chief officer of the not-for-profit organization International Medical Image Bank for Congenital Heart Diseases. The other authors report no conflicts.

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