Definition

Double Outlet Right Ventricle (DORV) is a type of ventriculoarterial connection in which both great vessels arise entirely or predominantly from the right ventricle. DORV is a type of conotruncal malformation with an incidence of .0003 to 0.2/1000 live births in infants and often associated with 22q11 deletion. If 50% or more of the circumference of the semilunar valve of the great vessel is committed to the ventricle, then the vessel is considered to arise from that ventricle. During transthoracic echocardiography, the short axis is preferred over the long axis view for diagnosing the overriding of the arterial valve because the motion of the heart can cause a change in the tangent within the long axis view.

Clinical presentation

The clinical presentation of a newborn with DORV can vary based upon the presence and location of ventricular septal defect, relationship of the ventricular septal defect to the great vessels, associated pulmonary or systemic outflow tract obstruction, other cardiac anomalies and the degree of pulmonary vascular resistance. Clinical continuum of DORV ranges from cyanosis (TOF type) due to obstruction in the right ventricular outflow tract to that of congestive heart failure and overcirculation (VSD type) in absence of pulmonary outflow tract obstruction. Perioperative management depends on the anatomical variants and the concomitant clinical presentation. Additionally, other cardiac defects associated with DORV (e.g.: AV-valve abnormalities, ASD, multiple VSDs, ventricular hypoplasia, arch anomalies) may influence the physiology or complicate the surgical repair. Surgical correction of DORV can be either univentricular or biventricular depending upon the degree of straddling and overriding of the valves, hypoplasia of the ventricle, and presence of other associated defects that preclude biventricular repair.
Role of echocardiography

Segmental approach is used in transthoracic echocardiography to assess the anatomy. Transthoracic echocardiography serves as the major imaging modality to diagnose DORV and for further surgical planning. Preoperative transesophageal echocardiography (TEE) is used to confirm the transthoracic findings, examine for the presence of any additional defects that were missed during the transthoracic imaging and identify any interval anatomical or physiological changes. Post-repair intraoperative TEE examination is done to confirm the adequacy of repair, identify residual defects, and assess heart function. This presentation primarily focuses on DORV with normal atrial situs and ventricular looping undergoing two-ventricle repair.

Classification

DORV can be classified based upon

1. Location of ventricular septal defect (VSD)
2. Relationship of the great vessels with each other

1. Location of VSD

This includes the relational anatomy between VSD and the great arteries. Based on the location, the VSD in DORV are characterized into four types:

I. Sub-Aortic
II. Sub-Pulmonary
III. Doubly-Committed
IV. Non-Committed

I. Sub-Aortic

This is the most common type of VSD in DORV and occurs in nearly 50% of cases. The ventricular septal defect is cradled between the limbs of the septomarginal trabeculation beneath the aortic valve, and to the right of the conal septum. The postero-inferior margin can be in fibrous continuity with the tricuspid valve or it may consist of muscular extension of the
posterior limb of septomarginal trabeculation. The outlet septum fuses with the anterior limb of the septomarginal trabeculation, thereby excluding the pulmonary valve from any relationship with the ventricular septal defect. There is absence of subaortic conus, so superior margin of the VSD is formed by the aortic annulus itself. In this case, the pulmonary trunk is usually anterior and leftward. This type of VSD anatomically and physiologically resembles the VSD of Tetralogy of Fallot (TOF). Some authors claim that the difference between the two is the presence of aortic mitral continuity in TOF and its absence in DORV. In such subaortic VSD, the flow of blood from the left ventricle is directed towards the aorta.

The following TEE views are used to evaluate the VSD. Magnitude and direction of shunting across the VSD can be assessed with color and spectral doppler

- Mid-esophageal four chamber view
- Mid-esophageal right ventricular inflow-outflow view
- Mid-esophageal aortic valve long axis view
- Deep transgastric long axis view of the left ventricular outflow tract
- Deep transgastric long axis view of the right ventricular outflow tract

The distance between the crest of the ventricular septum to the aortic valve can be assessed in the mid-esophageal four chamber view, mid-esophageal aortic valve long axis view, and deep transgastric long axis view. These views also help determine the relational anatomy between the VSD and the great vessels. The size of the VSD is measured in relation to the aortic valve.

The subaortic VSD in DORV can be associated with subpulmonary stenosis due to anterior displacement of the outlet septum or hypertrophy of the muscle bundles of the right ventricle. There may also be pulmonary valve hypoplasia.
The following TEE views are recommended to evaluate the morphology of pulmonary valve and to assess the cause and magnitude of RVOT obstruction. Color Doppler and spectral Doppler is used to identify the site and severity of obstruction

- Mid esophageal right ventricular inflow–outflow view
- Mid esophageal aortic valve long axis view.
- Mid esophageal ascending aortic short axis view
- Upper esophageal aortic arch short axis view
- Deep transgastric long axis view of the right ventricular outflow tract
- Deep transgastric long axis view of the left ventricular outflow tract

Sub aortic VSD in the setting of DORV can also be associated with straddling of Tricuspid valve. This can be evaluated in mid esophageal four chamber view and transgastric basal short axis view. Straddling is present when the chordae tendineae or papillary muscles of the valve cross the VSD and insert into the contralateral ventricle.

Coronary artery anomaly can also be present. The presence of any branch of coronary artery crossing the infundibulum can be evaluated either by counter clockwise rotation in mid-esophageal aortic valve long axis view or mid-esophageal aortic valve short axis view. The coronary arteries usually arise from the aortic sinuses facing the pulmonary valve. Absence of bifurcation of left coronary artery may indicate abnormal origin of its branches.

II. Sub-Pulmonary

Sub-pulmonary VSDs are present in approximately 30% of patients in surgical series of DORV. They lie anteriorly and superiorly beneath the pulmonary valve in the interventricular septum and are cradled within the limbs of the septomarginal trabeculation. The muscular outlet septum is attached to the mid-portion of the ventriculoinfundibular fold along with the posterior limb of the septomarginal trabeculation, thereby moving the aortic valve away from the defect although the pulmonary valve is closely associated with the VSD. Subaortic conus
is present. In this case, the anatomy and physiology resembles that of a transposition. If more than 50% of the pulmonary artery arises from the left ventricle, it is transposition; otherwise it is DORV. In case of transposition there is fibrous continuity between pulmonary and mitral valve which is absent in DORV. Left ventricular blood flow is directed to the pulmonary artery by the sub pulmonary VSD.

Taussig-Bing anomaly is described as DORV with sub pulmonary VSD. The sub-aortic and sub-pulmonary conus separate both aortic and pulmonary valves from atrioventricular valves (Bilateral Conus). The pulmonary and aortic valves lie side by side (parallel) and are at the same height.

The sub-pulmonary VSD in this setting can be associated with sub-aortic narrowing, aortic arch hypoplasia and interrupted arch. Sub-aortic narrowing can be assessed by TEE in the following views:

- Mid esophageal aortic valve short axis view
- Mid esophageal aortic valve long axis view
- Mid esophageal four-chamber view
- Deep transgastric long axis view of the left ventricular outflow tract
- Mid-esophageal ascending aortic short axis view

Muscular separation between the aortic and mitral valves will be evident in long axis views. PW Doppler can be used to evaluate different levels of narrowing. Continuous wave Doppler may be needed for high velocities across the obstruction. Use spectral Doppler to evaluate regurgitation across the valves. If arterial switch is planned, measure the pulmonary and aortic valve annular size in systole.
Transthoracic echocardiography is the best to evaluate aortic arch hypoplasia, interruption, coarctation. TEE views for evaluating coarctation are

- Upper-esophageal aortic arch short axis view
- Upper-esophageal aortic arch long axis view

Coronary artery anomaly should be assessed by 2D and color Doppler using very low Nyquist limit. There can also be presence of intramural coronary artery. The left main and left anterior descending artery may pass anterior to the pulmonary root.

These lesions may also be associated with straddling of mitral valve and sub-pulmonary stenosis.

III. Doubly-Committed

Doubly-committed VSDs occur in approximately 10% of DORV surgical series. The VSD lies within the divisions of septomarginal trabeculation superiorly in the interventricular septum and immediately beneath the leaflets of the aortic and pulmonary valves (juxtaarterial). The pulmonary and aortic valves generally are contiguous, because the infundibular septum is deficient or absent. The semilunar valves form the superior border of this typically large VSD. The septomarginal trabeculation, along with its anterior and posterior divisions, make up the anterior, inferior, and posterior borders of the VSD.

IV. Non-Committed

Non-committed (remote) VSDs occur in 10-20% of patients in surgical series. These defects are far removed from both the aortic and pulmonary valves, not nestled within the limbs of the septomarginal trabeculation, and located within the inlet septum without perimembranous extension or trabecular interventricular septum. Many of these DORV with non-committed (remote) VSDs are associated with CAVC (complete atroventricular septal defect or CAVSD).
It is difficult to surgically create an unobstructed pathway from the left ventricle to one of the great vessels in this setting, therefore it may need to be palliated to single ventricle physiology.

TEE evaluation of atrioventricular valves plays an important role intraoperatively as tricuspid valve leaflets may be crossing the VSD and great vessels in a way that makes it difficult to close the VSD. TEE views to evaluate the atrioventricular valves are

**Left AV Valve**
- Mid esophageal four chamber view
- Mid esophageal two chamber view
- Mid esophageal long axis view
- Mid esophageal mitral commissural view
- Transgastric basal short axis view
- Transgastric long axis view

**Right AV Valve**
- Mid esophageal four chamber view
- Mid esophageal right ventricular inflow–outflow view
- Mid-esophageal bicaval view
- Transgastric basal short axis view
- Transgastric RV inflow view

2. **Relationship of the great vessels with each other**

   There are two main types of arterial relationships found in double outlet right ventricle

   I. **Spiraling**

   II. **Parallel**

   I. **Spiraling**
In this case the aorta is usually rightwards and posterior to the pulmonary trunk. The two arteries spiral around each other as they emerge from the base of the heart. This type of pattern is usually associated with a sub-aortic VSD.

II. Parallel

The great vessels can also be parallel to each other and do not spiral. There can be four types of arrangements in parallel great vessels.

- Aorta is rightwards and side by side to pulmonary trunk. In this case the associated VSD is usually sub pulmonary
- Aorta is rightward and anterior to pulmonary trunk
- Aorta is directly anterior to pulmonary trunk
- Aorta is leftward and anterior to pulmonary trunk (L-malposition) – Least common type of relationship. Usually associated with sub-aortic VSD

TEE views to evaluate the relationship of great vessels

- Mid-esophageal four chamber view – pull up
- Mid-esophageal aortic valve long axis view
- Mid-esophageal right ventricular inflow–outflow view
- Deep transgastric long axis view
- Mid esophageal ascending aortic short axis view

Aorta is identified as the vessel giving rise to the coronaries in the absence of bifurcation. The pulmonary artery on the other hand bifurcates. Parallel arteries appear simultaneously in a cross section.

Associated anomalies

- ASD

DORV with transposition type of physiology can be associated with presence or absence of atrial
level shunt. The size, direction and magnitude of shunt across the ASD should be evaluated with color and spectral doppler. TEE views to evaluate the ASD are

- Mid-esophageal four-chamber view.
- Mid-esophageal bicaval view
- Mid-esophageal aortic valve short axis view
- Mid-esophageal right ventricular inflow–outflow view

**Additional muscular VSD**

Use color Doppler and contrast echocardiography if necessary to find additional VSD's.

**Ventricular hypoplasia**

**Right sided Aortic arch**

Arch sidedness is evaluated by transthoracic imaging

**Additional TEE examination**

SVC, IVC and PV should be examined to confirm the situs and rule out obstruction

TEE views to evaluate the valve in SVC

- Mid-esophageal bicaval view
- Deep transgastric long axis view.
- Upper-Mid-esophageal ascending aortic short axis view

TEE views to evaluate the valve in IVC

- Mid-esophageal bicaval view
- Lower-esophageal /Transgastric view

TEE views to evaluate the valve in Pulmonary veins

- Mid esophageal four chamber view
- Mid-esophageal aortic valve short axis view
- Mid-esophageal ascending aortic short axis view

**Post-operative TEE evaluation**

- Valvular stenosis or regurgitation
- Left ventricular outflow tract obstruction – In case of small VSD and prominent / elongated subaortic conus
- Right ventricular outflow tract obstruction
- Residual VSD
- Additional VSD’s
- Residual ASD
- Ventricular dysfunction
- PW Doppler in descending aorta can we used to examine repaired aortic arch. Low velocity laminar flow without a diastolic gradient is seen.
- Coronary flow is examined by Pulse wave and color doppler

Conclusion

Echocardiography plays an important role in diagnosing, identifying and understanding anatomy and physiology of various types of DORV defects as well as associated cardiac lesions. Echocardiography is an essential diagnostic modality for medical, surgical and long-term management of DORV.
References:

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2. Benjamin W. Eidem FC, Patrick W. O’Leary. Echocardiography in Pediatric and Adult Congenital Heart Disease: Lippincott Williams & Wilkins; October 28, 2009.