16 Double-Outlet Right Ventricle

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Double-outlet right ventricle (DORV) is a heterogeneous congenital cardiovascular malformation defined by both great arteries arising mainly (>50%) from the RV. In almost all cases a VSD is present. In general, there is also discontinuity between the AV valves and semilunar valves with presence of a small muscular apron (conus) underneath the semilunar valves. DORV fits within the spectrum of conotruncal anomalies that includes on one end normally related great arteries and tetralogy of Fallot (TOF), and on the other end, transposition of the great arteries (TGA). DORV can also occur within the context of a functional single ventricle and other complex malformations, such as heterotaxy syndromes. The discussion that follows addresses mainly DORV in a biventricular circulation.

Classification

DORV can be anatomically classified based on the relationship of the VSD to the great arteries into (Figure 16-1):

- **Subaortic VSD.** The VSD is mainly related to the aortic valve.
- **Subpulmonary VSD (Taussig-Bing).** The VSD is mainly related to the pulmonary valve. The great vessels are usually malposed with the aortic valve anterior and rightwards to the pulmonary valve or a side-by-side arrangement is present.
- **Doubly committed VSD.** There is a lack of infundibular septum, both aortic and pulmonary valves are at the same level and in continuity, and the VSD is related to both great vessels.
- **Noncommitted VSD.** The VSD is remote to the great arteries.

Pathophysiology and Clinical Presentation

The clinical presentation in DORV is extremely variable, reflecting the underlying pathology. The physiology and clinical features are mainly determined by the anatomic relationship of the VSD to the great arteries and the presence or absence of outflow tract obstruction. These factors account for the classification of the morphologic variants into four physiologic types, each displaying clinical features that resemble those of other malformations as follows:

- **VSD-type:** mimics VSD (Chapter 11)
- **TOF-type:** mimics TOF (Chapter 13)
- **TGA-type:** mimics TGA (Chapter 14)
- **Remote VSD:** mimics VSD or AVSD (Chapters 11 and 12)

The relationship between the physiologic and anatomic classifications is shown in Table 16-1.

Coexistent pathology in DORV can also influence the physiology. Associated defects include pulmonary stenosis (PS) (common finding occurring at the valvar and/or subvalvar levels), secundum ASD, PDA, additional VSDs, AV valve anomalies, subaortic

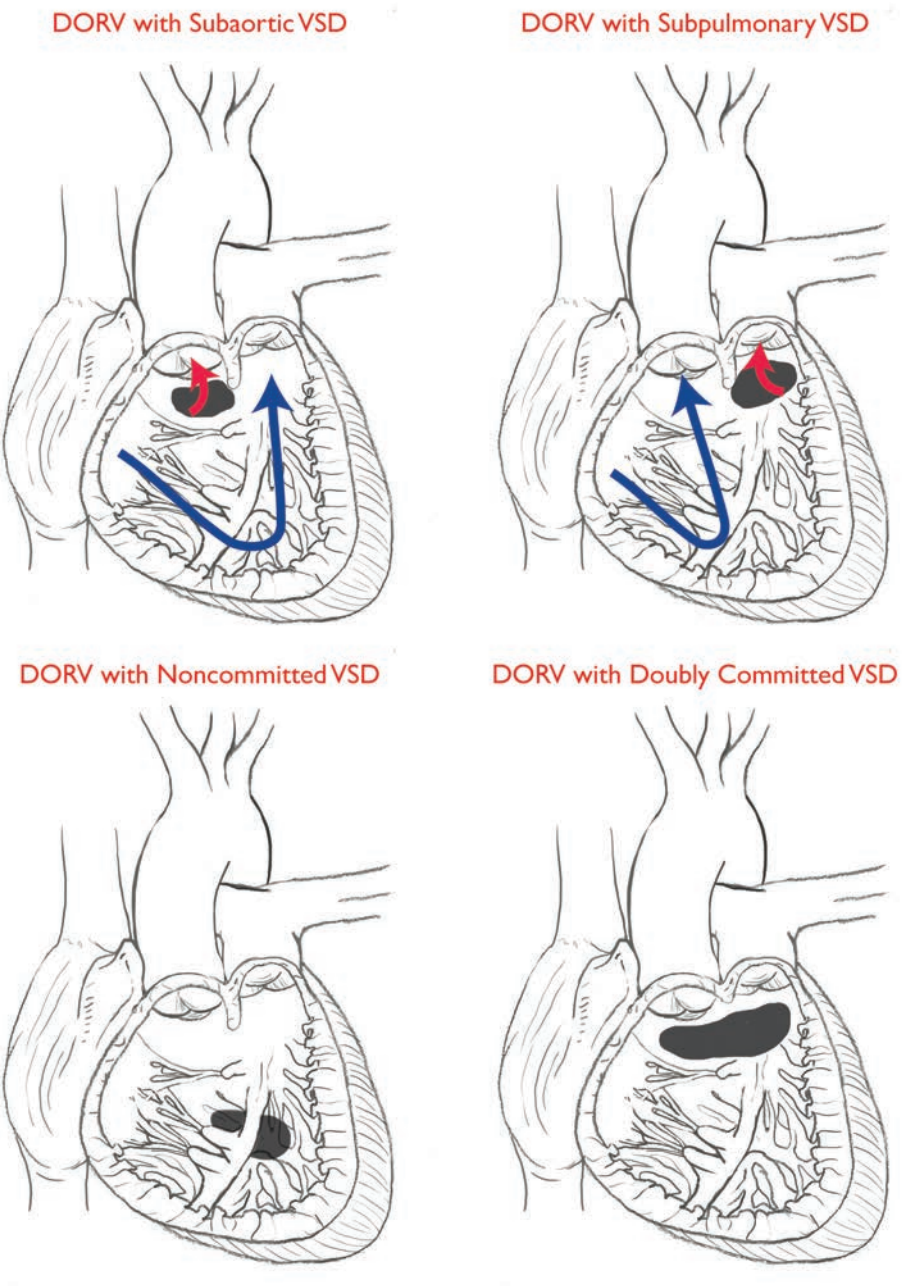

Figure 16-1. Anatomic classification of DORV based on the relationship of the VSD to the great vessels.

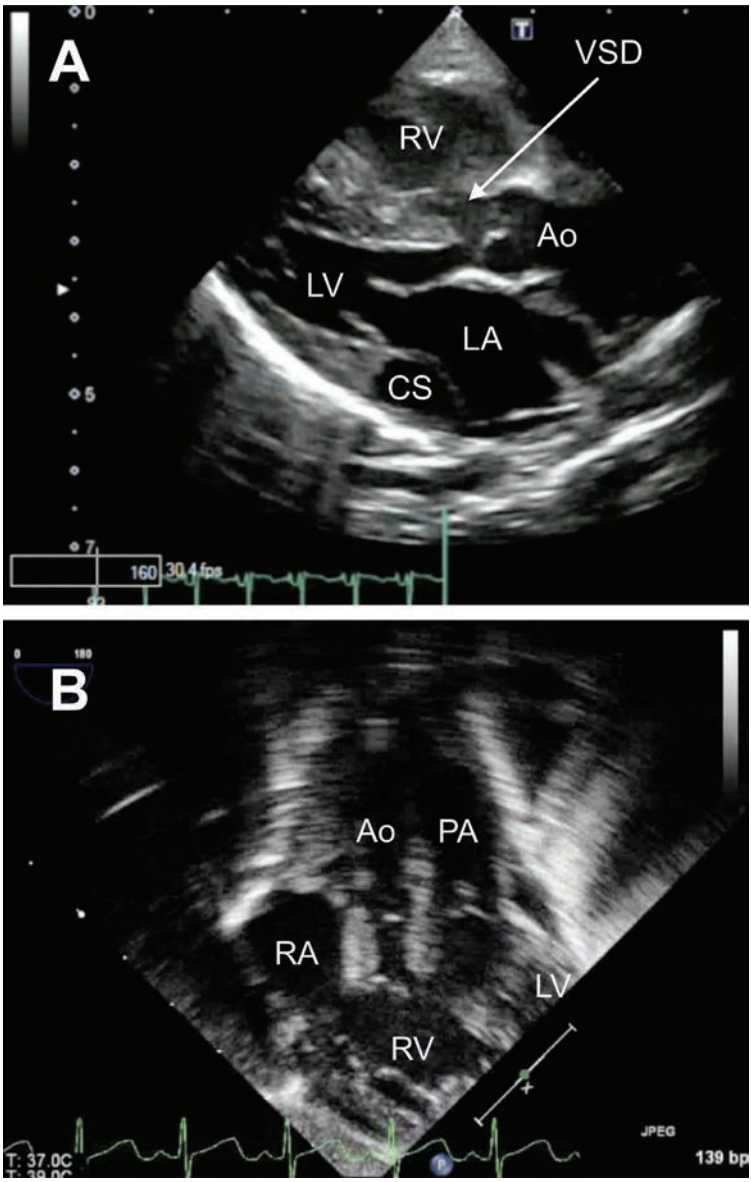

Figure 16-2. Echocardiographic diagnosis of DORV. A) TTE parasternal long-axis image of a patient with DORV and subaortic VSD showing the position of the aortic root over both ventricles, a VSD, and aortomitral discontinuity related to conal tissue. A dilated coronary sinus (CS) is seen related to a left SVC draining into the CS. The patient also had pulmonary stenosis (not shown). B) TEE deep transgastric image in a patient with DORV and subpulmonary VSD (Taussig-Bing) showing the origin of both great arteries from the RV, bilateral conal tissue, and narrowing of the subaortic region. There was associated aortic arch obstruction.

Table 16-1. Clinical and anatomic classifications of DORV.

Clinical classification	Anatomic classification	Pulmonary stenosis
VSD-type	Subaortic VSD Doubly committed VSD	-
TOF-type	Subaortic VSD Doubly committed VSD	+
TGA-type	Subpulmonary VSD	+ / -
Remote VSD	Non committed VSD	+ / -

stenosis (e.g., in the Taussig-Bing anomaly), aortic arch obstruction (may be associated with subaortic stenosis in the Taussig-Bing variant), right aortic arch, and coronary artery anomalies.

Diagnosis

- **ECG.** Frequently displays right-axis deviation, RA enlargement, and RVH.
- **CXR.** Findings are variable and not of significant diagnostic value in terms of the underlying pathology. Increased pulmonary vascular markings and cardiomegaly are consistent with an unrestrictive VSD. In contrast, the presence of oligemic lungs is suggestive of pulmonary outflow tract obstruction.
- **Echocardiogram (Figure 16-2).** Fetal echocardiography is diagnostic in most cases. Initial TTE focuses on the size and location of the VSD and its relationship to the arterial outlets, great artery relationships, morphology and patency of the outflow tracts, assessment of the AV valves, and characterization of associated anomalies. Noninvasive imaging guides balloon atrial septostomy (BAS) when indicated. TEE is used in most cases for intraoperative monitoring, to assist in surgical planning, and to evaluate the adequacy of the repair. This technology plays an important role in the exclusion of potential hemodynamically significant residual lesions such as intracardiac shunts, baffle obstruction, and valvar regurgitation. TTE remains the primary imaging modality for long-term surveillance.
- **Cardiac catheterization.** Not necessary in most cases. May be used to facilitate BAS in the Taussig-Bing variant. Angiography can be helpful in patients with complex anatomy. Hemodynamic measurements that include PA pressure and PVR are rarely needed, unless there is a late presentation or clinical concerns following a palliative procedure.
- **CTA/MRI.** May assist in the delineation of associated complex aortic arch anomalies and much less commonly, in the characterization of the anatomy that may impact surgical management (i.e., relationship of semilunar valves to the VSD). Postoperatively, it can be helpful during long-term surveillance.

Medical Management

The preoperative management of the patient with DORV is primarily influenced by the anatomic type and associated physiology.

Subaortic and Doubly Committed VSD

If a subaortic or doubly committed VSD is present and there is no PS, the clinical presentation is typically that of pulmonary overcirculation within the first few weeks of life. After the gradual expected decrease in PVR, PS may become apparent. If PS is of mild severity or nonexistent, tachypnea, intercostal retractions, hepatomegaly, and feeding difficulties are likely to ensue. Diuretic therapy is indicated in these cases (furosemide 1 mg/kg/dose every 6-12 hours). Overcirculation will follow the management of a large VSD (see Chapter 11).

TOF-Type

Children with TOF-type physiology require close monitoring depending on the degree of PS. Progressive cyanosis related to increasing subpulmonary obstruction or hypercyanotic episodes may develop (see Chapter 13). A hypercyanotic spell meets indication for hospital admission with consideration for surgical intervention (complete repair vs. palliation by means of systemic-to-pulmonary artery shunting).

TGA-Type

The neonate with TGA-type anatomy requires evaluation to ensure adequate intercirculatory mixing and aortic arch patency. PGE therapy may be necessary. Given that the VSD and PDA are usually not adequate sites for mixing, a BAS is recommended prior to surgical intervention in most cases unless shunting at the atrial level is unrestrictive.

Remote VSD

These patients usually present with signs and symptoms of pulmonary overcirculation. The VSD tends to be large enough to allow outflow from the LV and mixing of the systemic and pulmonary circulations. Overcirculation is managed medically until PA banding is performed.

Indications / Timing of Intervention

All patients with DORV require cardiac surgery. The majority of these procedures are undertaken during the first year of life. However, the specific type and timing of the surgical intervention depends on the pathophysiologic variant of DORV (see "Surgical Intervention" and Figure 16-3).

Anesthetic Considerations

The anesthetic management during the initial approach to this lesion hinges primarily on the particular physiologic variant and the planned surgical intervention.

Anesthetic goals may include balancing the pulmonary and systemic circulations and maintaining ductal patency by means of a PGE infusion as required to support pulmonary or systemic blood flow. Palliative procedures in the neonate and young infant such as PA banding or aortopulmonary shunting may be favored over initial corrective interventions. These usually require full invasive monitoring and adequate preparation for the respective procedure, however there is rarely a need for CPB. In contrast, corrective interventions imply the use of CPB. These procedures can be complex requiring a long aortic cross-clamp period. Main considerations during the

post-CPB phase include managing hemodynamics and the coagulation system. Efforts revolve around optimizing: (1) ventricular preload, assisted by TEE and guided by LAP monitoring, (2) myocardial performance and systemic vascular tone, with the use of inotropic/vasoactive agents, (3) pulmonary mechanics, by selecting suitable ventilation strategies, and (4) hemostasis, by the administration of blood products.

Surgical Intervention

VSD-Type

The repair entails creation of an intracardiac baffle between the LV and the aorta. For *subaortic* VSDs, since the defect is in close proximity to the aortic valve, the repair is very similar to closure of a perimembranous/outlet VSD. Ideally, the repair is performed between 6 and 8 months of age although it may take place earlier in life (including the neonatal period) if symptoms are not controlled with medical treatment. Repair of a subaortic VSD is performed through a right atriotomy using glutaraldehyde-fixed autologous pericardium for the baffle. The pericardium is secured to the rim of the defect and the tricuspid valve using either interrupted pledgeted sutures or a combination of running and interrupted pledgeted sutures. Analogous to perimembranous VSDs, the conduction system travels on the posteroinferior rim of the defect and is at risk of injury during the repair.

DORV with *doubly committed* VSDs are analogous to doubly committed juxta-arterial VSDs. Since the defect is underneath the semilunar valves, the repair is usually performed through a transverse pulmonary arteriotomy. A series of interrupted pericardial pledgeted sutures are placed through the pulmonary valve annulus and into the rim of the defect. A glutaraldehyde-fixed autologous pericardial patch is used to close the defect, therefore creating an intracardiac baffle between the LV and the aorta. The conduction system is usually away from the rim of these defects unless they extend into the perimembranous area. Due to the location of the semilunar valves mainly arising from the RV, it is possible for the baffle to impinge into the RV causing RVOT obstruction. The procedure is therefore ideally performed between 8 and 12 months of age. If patients require earlier intervention due to symptoms, placement of a PA band may be useful to delay surgical repair.

TOF-Type

Repair of TOF-type DORV is similar to repair of simple TOF, although it tends to be technically more difficult due to the more anterior location of the aorta. The procedure entails resection of RV muscle bundles and creation of an intracardiac autologous pericardial baffle through a right atriotomy followed by further resection of RV muscle bundles through a longitudinal pulmonary arteriotomy. If the pulmonary valve annulus is significantly hypoplastic, the incision is extended for a few millimeters through the annulus into the RV (transannular incision). The pulmonary arteriotomy is closed with a second autologous pericardial patch (see Chapter 13). The repair is ideally performed after 4-6 months of age. Younger patients with symptoms (significant cyanosis or hypercyanotic spells) may require placement of a modified Blalock-Taussig-Thomas shunt (mBTTS) prior to surgical repair.

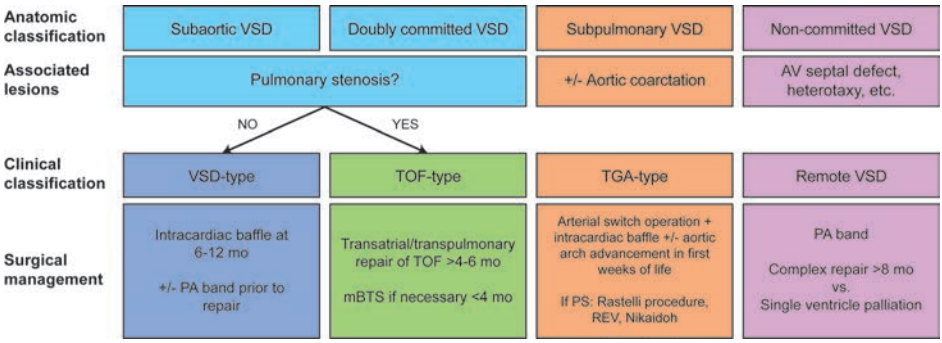

Figure 16-3. Algorithm for surgical intervention in patients with DORV.

TGA-Type

Repair of TGA-type DORV (Taussig-Bing anomaly) entails creation of an autologous pericardial baffle between the LV and the pulmonary valve (the semilunar valve closer to the VSD) and an arterial switch operation (see Chapter 14). Aortic arch hypoplasia or aortic coarctation are not unusual in this setting, requiring arch reconstruction in the form of an aortic arch advancement (see Chapter 25). Surgical intervention is usually undertaken in the first few weeks of life.

Some patients with subpulmonary stenosis may still be candidates for an arterial switch operation and subpulmonary resection. However, an arterial swtich operation may not be possible in some patients with severe PS, as it would translate into LVOT obstruction. The favored approach in these cases consists of a Rastelli procedure that includes creation of an intracardiac baffle between the VSD and the aortic valve with insertion of an RV-PA conduit. The native pulmonary valve is oversewn and the PA stump sutured closed. The conus between the aortic and pulmonary valves, which can be quite prominent, may be excised in order to allow a more direct connection between the VSD and the aorta ("reparation a l'etage ventriculaire" or REV procedure). In certain circumstances, a Nikaidoh procedure may be performed. This operation consists of translocating the aortic root posteriorly (to the previous position of the stenotic pulmonary valve) and reconstructing the RVOT with an RV-PA conduit or a patch. The coronary arteries are usually not translocated and therefore at risk of torsion during this procedure. Repair of TGA-type DORV with PS is usually performed later in life, ideally after infancy.

Remote VSD

Management of patients with a remote or noncommitted VSD is individualized depending on the anatomy. Since the outcomes of creating a very complex intracardiac baffle are suboptimal, a large proportion of these patients may be managed with single ventricle palliation. If so, placement of a PA band (see Chapter 39) is performed within the first few weeks of life to protect the pulmonary vascular bed, followed by subsequent creation of a bidirectional Glenn connection with the goal of eventual total cavopulmonary (Fontan) completion (see Chapter 39).

Postoperative Management

VSD-Type

Management after surgical treatment of VSD-type DORV is similar to that after closure of a large simple VSD (see Chapter 11).

TOF-type

Patients undergoing TOF-type repair can develop diastolic RV dysfunction postoperatively. Initiation of an esmolol infusion might be considered along with avoidance of catecholamine administration in an effort to optimize filling (preload) times. The rationale for esmolol therapy is to enhance diastolic filling times as RV diastolic dysfunction is transient, and to reduce the risk of dynamic RVOT obstruction from tachycardia (see Chapter 13). Diastolic dysfunction may require volume resuscitation. Intravascular volume is better assessed by LAP, as CVP may be elevated due to poor compliance of the RV. These patients are usually 200-300 mL positive after their first postoperative night.

TGA-Type

Following an arterial switch operation, concerning findings include increased LAP, arrhythmias, or ST-segment changes as these suggest myocardial dysfunction and/or coronary insufficiency (see Chapter 14). Perioperative management usually includes infusion of inotropes, agents that influence systemic/pulmonary vascular tone, and calcium. Ventricular dysfunction is in most cases systolic in nature, and therefore, is treated with inotropic support. Volume administration should be minimized. Nitroglycerin is routinely added to potentially limit coronary vascular reactivity given the necessary manipulations of the coronary arteries.

After the Rastelli or REV operation, RV systolic or diastolic dysfunction can be present, which tends to improve after a period of 24-48 hrs. Inotropic support with milrinone and low-dose catecholamine is customary in these patients.

If the operation was a Nikaidoh procedure, one should be vigilant about potential coronary torsion and/or AI. Inotropic support should be maintained until LCOS is resolved (usually the first 24 hours postoperatively). Careful attention to LAP, diastolic BP, and ST segments aid in the recognition of potential complications.

Remote VSD

If the operation was a PA band, the balance to achieve optimal arterial oxygen saturations will depend on minimizing pulmonary venous desaturation and maintaining moderate levels of oxygen provision (<60% FiO_2). Given that the increased afterload is seen by the two ventricles in parallel, it tends to be well tolerated. For details on PA band management and subsequent palliation, refer to Chapter 39.

Complications

- **Pleural effusions.** When there is important RV diastolic dysfunction, increases in CVP could prevent adequate clearance of pleural fluid and drainage of lymph/chyle into the innominate vein from the thoracic duct. Initial diuresis in conjunction with fluid restriction may stave off the accumulation of pleural effusions.
- **JET.** Although the incidence of JET at TCH is very low (atrial tachycardia is the most

common postoperative arrhythmia), it can affect some patients, particularly related to more extensive/complex intracardiac repairs. Addressing all potential triggers or exacerbating factors could "cool off" the automatic focus in JET. If these efforts are not successful (including cooling, lowering doses of catecholamine infusions, sedation, magnesium administration, overdrive pacing), antiarrhtymic treatment with amiodarone, esmolol, sotalol, or procainamide may be necessary (see Chapter 74).

TCH experience with biventricular repair of DORV (1995-2016)

Number of patients: 151

- VSD-type: 65
- TOF-type: 46
- TGA-type: 40

Perioperative mortality: 1.3%

5-year survival: 95%

5-year incidence of reintervention: 20-25%

- **Coronary translocation/torsion issues.** These tend to present early with regional segmental wall motion abnormalities on echocardiography, ECG changes (ST- and T-wave abnormalities), and increased LAP, accompanied by hemodynamic alterations and need for high inotropic support. In this setting, volume administration will only increase ventricular wall stress, further accentuating myocardial ischemia. Inotropic / vasoactive drugs are the agents of choice for hemodynamic optimization while diagnostic investigation is undertaken or interventions are considered to address coronary problems.
- **Complete heart block.** This represents an extremely rare complication. If AV dissociation is initially present in the patient receiving dexmedetomidine, the infusion should be discontinued, as it could cause or contribute to the conduction abnormalities. If high-grade block is still present 10 days after surgery (Mobitz II or third-degree AV block), implantation of a permanent pacemaker should be considered (see Chapter 75).
- **LCOS/LV dysfunction.** Usually transient and managed with epinephrine +/- milrinone infusions. Rarely, the epinephrine dose would exceed 0.05 mcg/kg/min. (see Chapter 71).

Long-Term Follow-Up

Close follow-up of patients with DORV undergoing either biventricular repair or single ventricle palliation is mandatory as long-term reintervention is not uncommon. The 5-year incidence of any reintervention (surgical or catheter-based) after biventricular repair is between 20-25%. Reinterventions most commonly include subaortic resection, pulmonary valvotomy/RVOT resection, conduit replacement, and branch PA intervention, depending on the type of DORV repair.