Chapter 3

Double-Outlet Right Ventricle

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Case Scenario

A 2-year-old child weighing 12.3 kg with a history of double-outlet right ventricle, large ventricular septal defect, and pulmonary stenosis presents as an outpatient for elective surgical repair of bilateral strabismus. She was palliated with a right-sided modified Blalock–Taussig shunt shortly after birth. Her parents report that she is active but has been taking more "breathing breaks" lately. Her current medications include aspirin and multivitamins. She sees her cardiologist regularly and complete repair of her cardiac lesion is planned in the forthcoming year. Her parents would prefer to take her home after surgery and would like to know if that is safe.

Vital signs include blood pressure 86/39 mm Hg, respiratory rate 22 breaths/minute, heart rate 127 beats/minute, and SpO_2 86% on room air.

Recent echocardiography findings include:

- *Dynamic subpulmonary obstruction, peak gradient 65 mm Hg through the outflow tract and pulmonary valve*
- *No branch pulmonary artery stenosis*
- *Normal biventricular function*
- *Subaortic ventricular septal defect*

Key Objectives

- Identify anatomic subtypes of double-outlet right ventricle.
- Review the pathophysiology and clinical presentation of double-outlet right ventricle.
- Understand the physiology of a ventricular septal defect combined with severe pulmonary stenosis.
- Formulate a safe anesthetic plan for children with unrepaired double-outlet right ventricle.

Pathophysiology

What is double-outlet right ventricle?

Double-outlet right ventricle (DORV) includes a variety of anatomic arrangements wherein both great arteries, the aorta and pulmonary artery, associate primarily with the right ventricle (RV). Children with DORV nearly always have a ventricular septal defect (VSD). The incidence of DORV is estimated at 1 in 10,000 live births and comprises approximately 1.0% of congenital heart disease (CHD) defects.

Double-outlet right ventricle has many anatomic subtypes depending on the relationship of the VSD to the great arteries and the presence or absence of other coexisting cardiac anomalies. Morphologically DORV lies between VSD with overriding aorta and transposition of the great arteries (TGA). In most cases the great arteries are normally related to each other (aorta posterior and to the right of the pulmonary trunk), but they can be positioned in any configuration in relationship to one another. The VSD is usually unrestrictive and can be variably positioned beneath the great arteries or, less commonly, be remote from the great arteries. Understanding the child's specific cardiac anatomy aids in determining the physiologic implications and guides planning for either surgical palliation or correction.

Clinical Pearl

The clinical presentation of patients with DORV depends on the relationship of the VSD to the great arteries, the variable relationship of the great arteries to each other, and the presence or absence of other coexisting cardiac anomalies.

What additional cardiac defects can be associated with DORV?

The following cardiac defects may be seen in association with DORV:

- Multiple VSDs
- Atrioventricular canal defects
- Atrioventricular valve abnormalities, stenosis, or atresia
- Right ventricular outflow tract obstruction: infundibular, valvular, and/or hypoplasia of pulmonary arteries

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- Subaortic stenosis, aortic arch anomalies, or coarctation of the aorta
- Ventricular hypoplasia
- Transposition of the great arteries
- Coronary artery anomalies

Double-outlet right ventricle can also be associated with trisomy 13, 18, and 21 as well as 22q11 deletion.

Clinical Pearl

Patients with newly diagnosed DORV should undergo thorough evaluation for the existence of associated cardiac and noncardiac anomalies.

What anatomic features determine the pathophysiology displayed by an individual patient?

The pathophysiology displayed in DORV is a result of the intracardiac shunting that occurs. This varies according to the following anatomic features:

- The size of the VSD and its relationship to the great arteries
- The relationship of the great arteries to one another
- The presence of outflow tract obstruction, either right or left
- The presence of other associated coexisting cardiac anomalies

How may the relationship of the VSD to the great arteries be described anatomically?

The anatomic relationship of the VSD to the great arteries provides the primary basis for the four different subtypes of DORV. Importantly, these descriptions do not imply that the VSD moves within the intraventricular septum, but rather they emphasize the highly variable relationship of the great arteries to each other. (See Figure 3.1.)

Ventricular septal defect–great artery relationships may be described as

- Subaortic VSD (most common)
- Subpulmonic VSD
- Noncommitted VSD: apical or muscular, remote from the great arteries
- Doubly committed VSD (least common)

Double-outlet right ventricle with subaortic VSD (with or without PS) accounts for approximately 50% of DORV presentations, while DORV with subpulmonic VSD accounts for 30%.

Clinical Pearl

Descriptions of the VSD and its relationship to the great arteries do not imply that the VSD moves within the intraventricular septum, but rather the descriptions emphasize the highly variable relationship of the great arteries to each other.

What are the physiologic subtypes of DORV?

Double-outlet right ventricle subtypes are a result of the variations that can exist in ventriculoarterial alignment, ranging from concordant or normally related great vessels to discordant relationships or transposition-type relationships. The "VSD-type" and "tetralogy of Fallot (TOF)-type" subtypes are seen with concordant great vessel relationships while the "Taussig–Bing" or "transposition of the great arteries" (TGA) subtype is seen with discordant great vessel relationships (aorta anterior and leftward of pulmonary artery).

Based on the physiologic subtypes of DORV, what clinical presentations are seen?

Double-outlet right ventricle physiology can result in clinical presentations ranging from pulmonary overcirculation and congestive heart failure to cyanosis and pulmonary hypoperfusion.

Double-outlet right ventricle with subaortic VSD: The great vessels are normally related. Two thirds of patients with DORV and a subaortic VSD also have PS in varying degrees. (See Figure 3.1a.)

- ***Double-outlet right ventricle, subaortic VSD without PS*** presents with ***VSD-type physiology***. These patients will have excessive pulmonary blood flow (PBF) and signs of congestive heart failure as PVR falls after birth.
- ***Double-outlet right ventricle, subaortic VSD with PS*** presents with ***TOF–type physiology***, with signs of diminished PBF due to the PS. The degree of PS determines the severity of clinical symptoms and signs. The infant will present with a systolic murmur and varying degrees of cyanosis depending on the degree of PS. If pulmonary stenosis is severe or near-critical, additional flow from a patent ductus arteriosus (PDA) may be necessary to ensure adequate PBF. Patients with severe PS may require urgent institution of a prostaglandin (PGE_1) infusion to maintain ductal patency. However, if PS is less severe, with adequate antegrade flow via the right ventricular outflow tract and a balanced systemic and pulmonary circulation, the child could present later in infancy or childhood.

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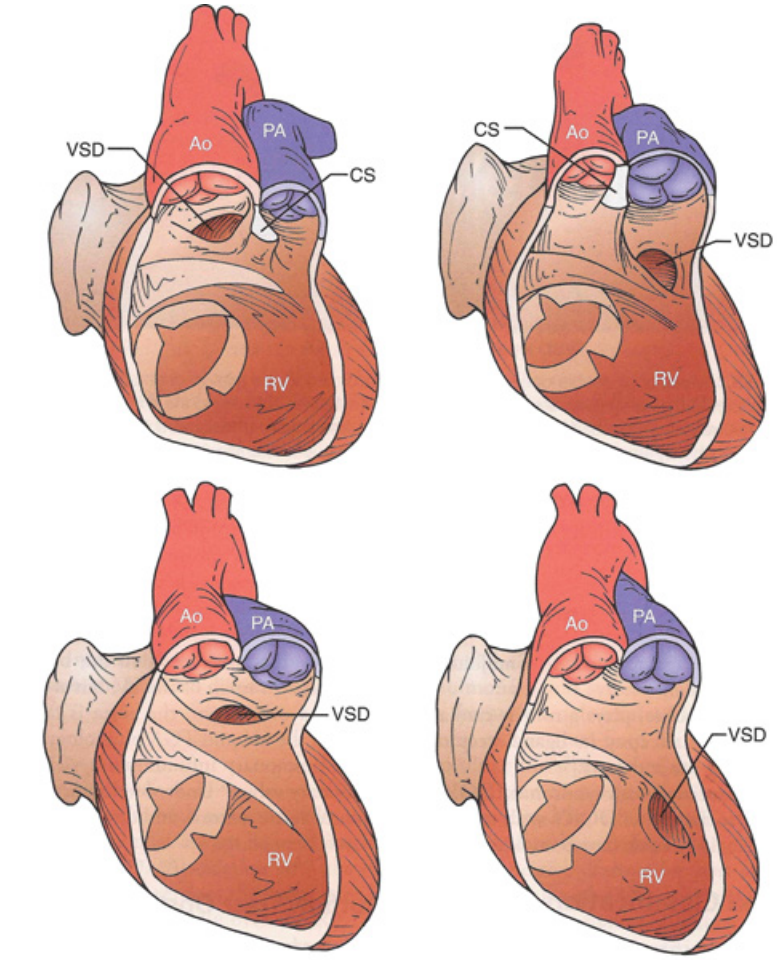

Figure 3.1 Double-outlet right ventricle subtypes according to ventricular septal defect location. Upper left, subaortic VSD. Upper right, subpulmonary VSD. Lower left, doubly committed VSD. Lower right, noncommitted VSD. From Bichell D. Double-outlet right ventricle. In Ungerleider R. M., Meliones J. N., McMillan K. N., et al., eds. *Critical Heart Disease in Infants and Children*, 3rd ed. Elsevier; 2019: 694–704. With permission.

Double-outlet right ventricle with subpulmonic VSD: Because the pulmonary artery (PA) is more closely associated with the LV in this variant, physiology more closely resembles transposition of the great arteries and is known as ***TGA-type***. (See Figure 3.1b.) In the Taussig–Bing anomaly, the great arteries are levo-malposed with no PS, and coronary artery and aortic arch anomalies are common. A parallel circulation exists because the LV primarily ejects oxygenated blood through the VSD into the PA while RV-to-aorta flow streams deoxygenated blood into the aorta. Infants with this subtype of DORV present in the neonatal period with cyanosis due to inadequate mixing of oxygenated and deoxygenated blood and may require a balloon atrial septostomy to improve intracardiac mixing and inotropic support to augment cardiac output.

Double-outlet right ventricle with doubly committed VSD can present as ***VSD-type*** physiology ***or TOF-type physiology*** depending on the presence or absence of PS or right ventricular outflow tract obstruction (RVOTO). A large component of the LV output is ejected through the VSD into the closely situated aortic valve. The amount of RV output (deoxygenated blood) directed to the aorta determines the oxygen saturation. Due to the high pulmonary vascular resistance (PVR) at birth these infants present with cyanosis of varying degrees. However, as PVR decreases pulmonary overcirculation may ensue. The infant then develops symptoms of congestive heart failure with tachypnea and failure to thrive. Medical management and optimizing caloric intake are mainstays of therapy until surgical intervention. (See Figure 3.1c.)

Double-outlet right ventricle with noncommitted VSD: Muscular or apical VSDs are more remote from the great arteries and therefore described as noncommitted. These patients have physiology that resembles a complete atrioventricular canal defect. (See Figure 3.1d.)

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Clinical Pearl

Double-outlet right ventricle physiology can result in clinical presentations ranging from pulmonary overcirculation and congestive heart failure to cyanosis and pulmonary hypoperfusion. In a patient with DORV, a subaortic VSD, and PS, the degree of PS determines the severity of clinical symptoms and signs.

What are the cardiac surgical treatment options for patients with DORV?

The goal of surgical treatment for DORV is complete anatomic repair, with closure of the VSD and connection of the RV to the pulmonary artery and the LV to the aorta. Timing of surgical repair is dependent on the patient's symptomatology and associated cardiac anomalies.

- ***VSD-type DORV***: An intraventricular baffle is placed to close the VSD and direct LV flow through the VSD into the aorta.
- ***TOF-type DORV***: Pulmonary stenosis can range from relatively mild to severe and may be valvular, subvalvular, or supravalvular. If PS is severe, the child may be surgically palliated via placement of a modified Blalock–Taussig (mBT) shunt (right subclavian or innominate artery to pulmonary artery) to improve PBF and allow growth while awaiting definitive repair. Corrective surgery will utilize an intraventricular baffle to close the VSD and either a patch to enlarge the RVOT or a right ventricle-to-pulmonary artery (RV–PA) conduit.
- ***TGA-type DORV***: An arterial switch procedure is performed along with a baffle closure of the VSD to the PA.

If patients have associated anomalies such as unbalanced ventricular size or straddling atrioventricular valve chordae that render the size of the ventricles or atrioventricular valves inadequate for a two-ventricle repair, they may need to undergo a staged single ventricle palliative approach.

Anesthetic Implications

What is expected clinically for this patient?

The clinical picture resembles that of TOF, with decreased PBF and resultant right-to-left (R-to-L) shunting due to PS. Even though this patient underwent an mBT shunt placement early in life, she may still be at risk for hypercyanotic spells. Most mBT shunts utilize a GOR-TEX® graft, which is not able to grow with the child, thus leading to progressively worsening cyanosis. At 2 years of age with an unrepaired DORV, this child is likely to have symptoms consistent with reduced PBF evidenced by her inability to keep up with her siblings, frequent stopping at play and squatting, and baseline oxygen saturation of 86% on room air. On physical examination central and peripheral cyanosis with clubbing of digits may be evident.

What information is relevant in the preoperative anesthesia assessment?

A detailed clinical history including previous anesthetic issues, current physical status, allergies, and current medications should be obtained. Her last report from her cardiologist should be reviewed as well, and he or she should be made aware of the impending surgery. The pediatric cardiology evaluation should outline the congenital cardiac anomaly, latest echocardiogram report and important current hemodynamic issues and management plans. It is important to understand the cardiac anatomy and pathophysiology by reviewing the echocardiogram and any other available imaging data. A review of the latest electrocardiogram to screen for arrhythmias is important. A discussion between cardiology and ophthalmology should occur regarding the patient's use of aspirin in the perioperative period.

What are some of the perioperative concerns and risks associated with anesthesia?

The perioperative anesthetic management of the patient with DORV, VSD, and PS is similar to that of patients with classic TOF. Physiologic goals include maintaining adequate PBF and baseline oxygen saturations. Increases in heart rate and contractility should be avoided if hyperdynamic components of RVOTO are present. Systemic hypotension should be avoided and systemic vascular resistance (SVR) maintained. Adequate preload should be assured by minimizing fasting time and judicious use of intravenous (IV) fluids. (See Chapters 7, 8, and 47.)

In view of the patient's cardiac physiology particular care should be taken to avoid additional risk factors that might require surgical postponement, such as recent upper or lower respiratory infections.

Clinical Pearl

The perioperative anesthetic management of the patient with DORV, VSD, and PS is similar to that of patients with classic TOF.

Is premedication appropriate for this patient?

Children with DORV, VSD, and PS are predisposed to cyanotic spells. Preoperative events such as prolonged

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fasting duration, acquisition of IV access, and agitation due to separation anxiety may affect the incidence of or provoke cyanotic spells. A preoperative premedication (either midazolam, or a combination of midazolam and ketamine) to decrease the patient's anxiety and enhance compliance with acquisition of IV access is generally beneficial. Alternatively, if the patient already has IV access, dexmedetomidine is an excellent choice as well.

Considering that this child has had an mBT shunt, what precautions should be observed?

In children with a palliative shunt, it is important to maintain adequate hydration, minimize fasting time, avoid stimuli that can provoke preoperative cyanotic spells, and reduce hypotension during all phases of anesthesia. Dehydration can reduce systemic blood pressure and blood flow through the shunt and increase viscosity, which may promote shunt thrombosis.

Most patients with an mBT shunt are maintained on oral anticoagulation or antiplatelet therapy to prevent shunt thrombosis. The pros and cons of withholding anticoagulants in these patients should be discussed with the surgeon and primary cardiology team. In all cases, a plan to restart or bridge anticoagulation for the perioperative period should be in place. The cardiology team should be consulted in cases where the patient is admitted postoperatively.

Clinical Pearl

In patients with mBT shunts, cessation of anticoagulation should be carefully considered and a plan for perioperative management discussed with the cardiologist and surgeon. If anticoagulation is stopped for a procedure, it should be stopped for a minimal amount of time and resumed as soon as possible.

What perioperative monitoring is appropriate for this patient?

Monitoring during strabismus surgery should follow the American Society of Anesthesiologists standard guidelines. Standard noninvasive monitoring is ideal for surgical procedures with minimal blood loss and minimal hemodynamic aberrations. The surgical procedure to be performed along with expected blood loss and fluid shifts often dictates the need for invasive access, particularly arterial blood pressure monitoring. In this case strabismus surgery is unlikely to result in dramatic hemodynamic changes and invasive monitoring is not warranted. Preoperative baseline hemoglobin-oxygen saturations should be noted and maintained throughout surgery.

What anesthetic considerations are specific to this case?

The occurrence of significant bradycardia and even asystole may occur during ocular manipulation via the oculocardiac reflex. Although anticholinergic premedication (atropine or glycopyrrolate) will help delay and decrease the occurrence of bradycardia, it will not abolish the reflex bradycardia during ocular surgery. In addition, anticholinergic premedication may cause significant tachycardia, which can reduce PBF in the patient with TOF-type physiology. If bradycardia secondary to the oculocardiac reflex does occur, it is best to withhold further surgical stimulation until the heart rate recovers spontaneously. If significant bradycardia persists with the absence of surgical eye manipulation, then treatment with anticholinergics may be warranted.

For this procedure, a laryngeal mask airway may be appropriate, avoiding the depth of anesthesia required for endotracheal intubation. Additionally, analgesia, either topical or by formal ocular block, may be requested from the surgeon and can reduce hemodynamic changes associated with strabismus surgery. Topical medications instilled into the eyes should be discussed with the anesthesiologist and announced prior to administration as they often have systemic absorption.

Clinical Pearl

All ophthalmic medications should be discussed with the anesthesiologist as their systemic absorption can affect cardiac physiology.

What are important intraoperative considerations?

Intraoperative systemic hypotension should be avoided as it will exaggerate the amount of R-to-L shunting and thereby the degree of cyanosis. Patients with a palliative shunt are at risk of shunt occlusion during hypotensive episodes and prolonged fasting periods. A good clinical practice is to auscultate the chest for a shunt murmur over the right parasternal area, assessing mBT shunt flow. This should be done periodically intraoperatively and postoperatively.

The approach to avoiding intraoperative cyanotic spells includes avoiding dehydration, optimizing IV hydration once access is obtained, and optimizing analgesia during surgery. During anesthetic induction it is important to

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recognize that significant reductions in SVR will increase R-to-L shunting. Should significant cyanosis occur, treatment includes IV fluid boluses, administration of 100% oxygen, and prompt treatment with vasopressors such as phenylephrine.

Clinical Pearl

Significant reductions in SVR should be avoided as they will increase R-to-L shunting and cyanosis.

Should sudden changes in oxygenation and hemodynamics occur, what are most likely causes?

Changes in the quality of the shunt murmur along with abrupt changes in blood pressure, oxygen saturation and/or end-tidal CO_2 should be immediately evaluated and aggressively treated to exclude shunt occlusion as an etiology for decreased PBF. Increasing blood pressure with a systemic vasoconstrictor such as phenylephrine should increase blood pressure and improve oxygen saturations. If the decrease in oxygen saturations persists despite the administration of fluids and phenylephrine, a high probability of shunt occlusion should be assumed and the patient should be given intravenous heparin as an anticoagulant to attempt to reestablish shunt flow and an echocardiogram should immediately be performed to assess the flow characteristics of the mBT shunt.

Clinical Pearl

A sudden change in hemodynamics, with a decrease in oxygen saturation that is nonresponsive to fluid administration or phenylephrine, should prompt the anesthesia provider to consider the possibility of a shunt occlusion.

How should extubation of this patient be managed?

Aside from the standard extubation criteria, avoidance of excessive coughing and bucking is crucial to avoid significant intrathoracic pressure elevation and cyanotic episodes. Titration of dexmedetomidine to achieve a smooth and stable emergence while preserving airway patency is often a useful approach.

What are some concerns for postoperative recovery in this patient?

This patient should be observed postoperatively in a monitored setting with providers able to recognize and treat cyanotic spells. The patient should be monitored for the maintenance of an appropriate heart rate and rhythm, blood pressure, and oxygen saturations. In the patient with a palliative mBT shunt it is important that the cardiology team see the child postoperatively and that anticoagulant therapies be resumed as soon as possible unless an alternate plan is already in place.

If the patient presented as an outpatient, what are the criteria for discharge home?

In many cardiac centers a patient with an mBT shunt is monitored in the hospital overnight even after an uneventful anesthetic and surgery. These patients are still at risk for postoperative pain and poor oral intake, which could increase the risk for shunt thrombosis or a hypercyanotic spell. In the patient with an mBT shunt it is important that anticoagulation medications are resumed prior to discharge home.

Suggested Reading

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