Perioperative Management of DORV Case

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Objectives:

1. Discuss considerations regarding anesthetic care in the patient with double-outlet right ventricle
2. Review national data about DORV from CCAS/STS database

The diagnosis of “double-outlet right ventricle” characterizes a heterogeneous group of congenital cardiac malformations in which both great arteries are related to the right ventricle and a ventricular septal defect is almost always present. Characteristics of the anatomy which must be elucidated include the position of the VSD in relation to the great arteries, the relationship of the great arteries to one other, the morphology of the ventricles and their outflow tracts, and the presence of associated anomalies. Other associated anomalies seen with a DORV include A-V canal defects, aortic arch obstruction, outflow tract obstruction, ventricular hypoplasia, and heterotaxy syndromes. Multiple classification schemes have been developed to describe this condition. A clear understanding of a patient’s anatomy is critical to understanding the physiologic consequences of the specific lesion. The Society of Thoracic Surgeons (STS) database currently divides patients with a DORV into 5 basic types:

1) VSD type: DORV with subaortic or doubly committed VSD (no PS)
2) TOF type: DORV with subaortic or double committed VSD and PS
3) TGA type: DORV with subpulmonary VSD (with or without PS)
4) Remote VSD type: DORV with a remote VSD (with or without PS)
5) DORV and AVSD

A clear understanding of the specific type of DORV and other associated anomalies is critical to understanding the physiologic consequences and developing an anesthetic plan. Depending on the specific anatomy, the child with
a DORV is deemed to have a one or two-ventricle heart, may require palliative procedures during the first few months of life, or undergo a complete surgical repair.

**DORV-VSD Type**

This type of DORV is characterized by either a subaortic or doubly committed VSD and no pulmonary stenosis (PS). The location of the VSD in close proximity to the aortic valve leads to fully oxygenated blood from the left ventricle being directed towards the aorta. These children generally present in the first few months of life with pulmonary overcirculation and congestive heart failure, and in many centers will undergo a complete repair in the first 6 months of life. This diagnosis was present in 0.9% of infants undergoing cardiac surgery between 2008 and 2011 (STS database). Anesthetic management is similar to the child with a large VSD, and ventilation strategies to minimize pulmonary overcirculation are utilized. Low inspired oxygen concentrations and mild hypoventilation are used to increase pulmonary vascular resistance and improve systemic cardiac output.

**DORV-TOF Type**

This type is characterized by either a subaortic or doubly committed VSD and PS. This diagnosis was present in 0.5% of neonates and 1% of infants undergoing surgery between 2008 and 2011 (STS database). The clinical presentation is similar to the child with Tetralogy of Fallot (TOF) and the timing of surgical intervention depends on the degree of obstruction to pulmonary blood flow. Compared to the child with TOF, the VSD is usually located farther away from the aorta. Some surgeons choose to perform palliative systemic arterial to pulmonary artery shunts and delay a complete repair until later in infancy. Anesthetic management is similar to the child with TOF. Supplemental oxygen is administered to maintain oxygen saturations >80%, and desaturation episodes are managed by administration of volume or medications to increase systemic vascular resistance such as phenylephrine.

**DORV-TGA Type**

This type of DORV is characterized by a subpulmonary VSD with or without PS and occurs in approximately 25% of patients with DORV. This diagnosis is present in 1.1% of neonates undergoing surgery between 2008 and 2011 (STS database). The Taussig-Bing anomaly is a subgroup of this type in which there is L-malposition of the great arteries and no pulmonary stenosis. A significant
number of these patients also exhibit subaortic stenosis and/or aortic arch obstruction. These children usually present in the neonatal period with cyanosis and physiology similar to the patient with TGA/VSD; this is due to the posterior and rightward deviation of the conal septum which leads to streaming of LV blood through the VSD and into the pulmonary artery.\(^6\) Management is based on interventions to improve intracardiac mixing of blood and optimizing systemic cardiac output. Although supplemental oxygen may lead to an increase arterial oxygen saturations, it is important to recognize that there is significant pulmonary overcirculation with transposition physiology and systemic oxygen delivery must be closely monitored (NIRS, lactate). In many centers a complete repair is performed during the neonatal period consisting of an arterial switch operation and VSD to pulmonary artery baffle, and will require enlargement of the aortic arch when obstruction is present. A recent publication from the Melbourne group found an early mortality of 5.3% from 1983 to 2009 in patients with the Taussig-Bing anomaly that underwent the arterial switch operation.\(^7\)

**Remote VSD Type**

This type of DORV is characterized by a remote VSD with or without PS, and is frequently associated with AV-canal defects. The VSD is usually located in the muscular septum and is distant from both the pulmonary artery and aorta.\(^8\) Early surgical interventions are usually palliative. When feasible, definitive two-ventricle repairs are usually delayed until later in infancy, and require the creation of an intraventricular tunnel (VSD to aorta or VSD to PA/ASO). Patients with this type of DORV usually present with pulmonary overcirculation and anesthetic management is similar to the patient with a large VSD.

**DORV and AVSD**

This type of DORV is characterized by an AVSD, and may be accompanied by heterotaxy syndrome, right ventricular outflow tract obstruction, and total anomalous venous return (TAPVR).\(^4\) The need for early surgical intervention is dependent on the balance between pulmonary and systemic blood flow and whether there is TAPVR.

**Anesthetic Management**

The preoperative evaluation is critical to developing an anesthetic plan for the child with a DORV. A complete understanding of the patient’s anatomy and physiology must be obtained by reviewing the echocardiogram, cardiac
catheterization data, and other imaging studies (MRI, CT). It is vital to recognize the presence of ventricular dysfunction since this may impact the anesthetic technique utilized. The EKG and any previous history of rhythm abnormalities should be reviewed. Patient allergies, medications, and a history of previous anesthetic issues are identified. A discussion with the surgeon about the planned procedure and any specific concerns prior to starting the case is beneficial.

Anesthetic induction can be performed using an inhalational anesthetic such as sevoflurane or with intravenous agents. In neonates that have intravenous access we prefer to use an opioid-based induction technique, as compared to most infants that receive an inhalational mask induction with sevoflurane. After placing intravenous catheters a non-depolarizing muscle relaxant is administered and a nasal endotracheal tube placed. Arterial and central lines are placed using ultrasound guidance. Because these patients may be undergoing a redo sternotomy and/or complex repair with multiple suture lines, it is important to have venous access which will allow for rapid transfusion of blood products. Maintenance of anesthesia is with a balanced technique consisting of fentanyl and inhalational anesthetic agents, and the dose of opioid administered is dependent on the age of the patient, ventricular function, and the postoperative ventilation plan. The ventilation strategy is based on the patient’s unique physiology with the goal being to balance systemic and pulmonary blood flow. The inspired oxygen concentration, use of hyper/hypoventilation, and pH can used to modulate pulmonary vascular resistance. In patients with ductal-dependent systemic blood flow, inspired CO₂ can be used to improve systemic cardiac output.

Monitoring at CCHMC includes a 5-lead ECG, non-invasive blood pressure, 2 oxygen saturation probes, ETCO₂, temperature monitoring (NP and bladder/rectal), arterial line, and central line. Three-site NIRS monitoring (R and L cerebral, somatic) is utilized in most neonates and infants. Transesophageal echocardiography is critical in the patient with a DORV and is performed prior to starting surgery and after separation from cardiopulmonary bypass (CPB).

Management of CPB is dependent on the planned surgical procedure. Moderate hypothermia (25-28 degrees C) is generally used for intra-cardiac repair, and deep hypothermia (18-21 degrees C) and antegrade cerebral perfusion when aortic arch reconstruction is necessary. Depending on the center and planned surgery, the CPB time and period of arrest for the heart may be lengthy. During cardiopulmonary bypass the hematocrit is maintained between 28-30%, and
lactate levels and NIRS are closely monitored to assess the adequacy of oxygen delivery.

Following surgical repair and prior to separation from bypass, inotropic agents are started and the heart rhythm evaluated. At our institution we use milrinone and epinephrine in most infants following complex cardiac repairs, and in neonates also initiate a calcium infusion. Following intraventricular baffling and VSD enlargement both atrial and ventricular arrhythmias are common, and temporary pacing may be necessary prior to separation from bypass. TEE is used to evaluate the surgical repair after separation from CPB and focuses on ventricular function, the presence of residual intra-cardiac shunts, the patency of the ventricular outflow tracts, and adequacy of the AV-valves. Patients that exhibit pulmonary overcirculation preoperatively are at risk for pulmonary hypertension and may benefit from treatment with nitric oxide. Once the surgical repair and patient hemodynamics are felt to be optimal, protamine is administered. For children undergoing a complex repair with multiple suture lines (i.e. Taussig-bing heart), platelets and cryoprecipitate are usually administered.

The patient with a double-outlet right ventricle can provide significant challenges for the anesthesiologist. This diagnosis represents a heterogeneous group of malformations which present in very different physiologic states. The anesthesiologist must be knowledgeable about the type of DORV and develop an anesthetic plan based on the individual physiology.

References:

3. Classifications of DORV from STS database
