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DOUBLE OUTLET RIGHT VENTRICLE

Surgical Specialty:	Cardiac
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Background:

- General Considerations
  - Double outlet right ventricle (DORV) is a complex cardiac malformation in which both great arteries arise from the right ventricle (RV)[1]
    - Almost always associated with ventricular septal defect (VSD), allowing mixing of oxygenated and deoxygenated blood
  - Incidence: approximately 1 in 10,000 births; 1% of all congenital heart defects[1-5]
  - DORV is a heterogenous lesion and differing classifications have been described. The Society of Thoracic Surgeons (STS) database divides DORV into five types as follows:[1,4,5]
    - VSD-type (no pulmonary stenosis)
    - Tetralogy of Fallot (TOF)-type (pulmonary stenosis present)
    - Transposition of Great Vessels (TGA)-type
    - Noncommitted VSD-type (VSD remote from semilunar valves)
    - DORV and atrioventricular septal defect (intact ventricular septum, rare)
  - The surgical approach may be palliation or correction[1]
  - Surgical repair in a neonate or infant helps correct cyanosis and prevent the development of pulmonary hypertension and Eisenmenger Syndrome[2]
- Patient Considerations[4,5]
  - Determining the type of DORV is important for understanding the patient's physiology and for developing surgical and anesthetic plans
  - Those with TOF-type and severe cyanosis from severe pulmonary stenosis or atresia require prostaglandins prior to surgical intervention to maintain ductal patency for pulmonary blood flow.
  - Those with TGA-type may have aortic arch disruption requiring prostaglandins for ductal patency to provide distal aortic blood flow.
  - Associated Comorbidities/Syndromes
    - Trisomy 13, 18, and 21
    - 22q11.2 deletion

Anesthetic Planning:

- Pre-Anesthetic Evaluation

- Detailed history for symptoms of congestive heart failure, episodes of cyanosis, or poor weight gain
- Physical examination should focus on:[2,4]
  - General Appearance
    - Cyanosis, activity level, work of breathing
  - Airway evaluation to identify barriers to mask ventilation/intubation and TEE probe placement, especially with associated syndrome/genetic defects.
  - Cardiac evaluation for the presence of tachycardia, holosystolic murmur, and mid-diastolic rumble
  - Pulmonary auscultation to evaluate for rales or tachypnea
- Review of available imaging such as echocardiogram, cardiac MRI, or cardiac catheterization to understand the patient's anatomy and physiology[5]
- Additional labs/tests indicated during work up[2-5]
  - Type and Screen
  - 12 lead EKG
  - Chest X-ray
- Discussion to have with surgeon/family
  - Code Status
  - ECMO candidacy

- Specific or Unique Room Set-up Requirements
  - Airway
    - Oral vs nasal intubation, depending on institutional protocol
  - Drugs/Infusions
    - Consider preoperative sedation to facilitate separation in older infants and children[5]
    - Antifibrinolytic infusion – aminocaproic acid or tranexamic acid[2]
    - Inotropes available for post-bypass period[5]
  - Monitors
    - Standard ASA monitors
    - 2 oxygen saturation probes (upper and lower extremity)
    - Central venous access
    - Arterial line, anatomic placement dependent on lesion and institutional guidelines
    - Near-infrared spectroscopy (NIRS) monitoring – cerebral and somatic[4,5]
  - Blood Availability
    - Packed red blood cells
    - Platelets, cryoprecipitate,fresh frozen plasma and/or factor concentrates (PCC or fibrinogen) to manage post-cardiopulmonary bypass coagulopathy, per institutional protocols[5]
  - ICU Bed Availability
    - Following surgery, directly admit to the intensive care unit[5]
  - Other indicated Resources[2,5]
    - Cardiopulmonary Bypass (CPB)
    - Transesophageal echocardiogram (TEE) to evaluate surgical repair, ventricular function, presence of residual intracardiac shunts, patency of ventricular outflow tracts, and adequacy of the AV valves

Intraoperative Considerations:

- General
  - Anesthetic management will vary depending on the type of DORV

  - Repairs can be accomplished as single or two ventricles, depending on multiple factors[3-5]
    - Neonatal palliative procedures: systemic to pulmonary artery shunt, coarctation repair, or pulmonary artery (PA) banding
    - Intraventricular repair with baffle from LV to aorta
    - Intraventricular baffle from LV to PA with arterial switch
    - Single ventricle pathway with staged repairs to Fontan completion
  - Bubble precautions due to the risk of systemic air embolism
- Induction
  - Intravenous or inhalational induction depending on provider preference and cardiac lesion
  - For TOF-type, ensure adequate anesthetic depth prior to airway manipulation to reduce the chance of hyper-cyanotic spell[5]
- Positioning
  - Supine
- Maintenance
  - Balanced anesthetic technique with inhalational agent, narcotics, and IV hypnotics
- Hemodynamic/Physiologic goals
  - Optimize systemic cardiac output and oxygen delivery with manipulation of PVR and SVR.
  - Avoid pulmonary over-circulation by maintaining pulmonary vascular resistance[5]
- Surgical Considerations
  - Extensive preoperative imaging studies can help guide the surgical plan, but intraoperative decision-making remains necessary due to unique patient presentations.
  - Due to the complexity of repairs, CPB and aortic cross-clamp times may be lengthy.
- Emergence/Disposition
  - Early extubation in the OR if the patient meets institutional criteria
  - Cardiac ICU for postoperative care
- Postoperative Care
  - Monitor for postoperative bleeding and hemodynamic abnormalities
  - Low cardiac output syndrome 6 to 12 hours after surgery is common[4,5]

Case-Specific Complications/Pitfalls

- Rhythm disturbances such as junctional ectopic tachycardia are common[4,5]
  - Reduced atrial contribution to ventricular filling can significantly affect cardiac output
  - Treat with amiodarone, pacing, cooling, or dexmedetomidine
- Left ventricular dysfunction[4]
  - Inadequate oxygen delivery is signaled by metabolic acidosis, oliguria, and peripheral vasoconstriction.
  - Manage by balancing preload, afterload reduction, and inotropy
  - If progressing with rising atrial pressure, decreasing SvO_2, and worsening metabolic acidosis, mechanical support may become necessary
- Right ventricular dysfunction[4]
  - This may be due to LV dysfunction or isolated RV failure
  - Presents with worsening systemic venous congestion, ascites, and edema
  - Manage with a combination of careful preload, inotropy, pacing, and pulmonary vasodilation
- Pulmonary hypertension[5]
  - More likely to occur in patients with preoperative pulmonary over-circulation

  - Increased vasoreactivity can occur due to CPB
  - Episodes can be induced by tracheal suctioning, hypercarbia, or hypoxia
  - Treatment includes chemical paralysis, inhaled nitric oxide, and hyperventilation
- Heart block[3,4]
  - Surgical manipulation near the AV node and bundle of His come with the risk of complete heart block
  - May be transient and only require temporary pacing in the postoperative period
    - If CHB remains after ten days, it is unlikely to recover and a permanent pacemaker is indicated

References

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