Tetralogy of Fallot with Absent Pulmonary Valve (TOF/APV) Guideline
What the Nurse Caring for a Patient with CHD Needs to Know

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Embryology

- Initial development of heart
  - Tube-like structure
  - Venous channels lead flow in
  - Arterial trunk provides flow out
- Development of tube
  - Distal portion becomes bulbus cordis (ventricle)
  - Proximal portion becomes the truncus arteriosus (great arteries)
- Septation of the truncus arteriosus
  - During week 5-6 of fetal development
    - Aortopulmonary septum of the truncus arteriosus usually completes a clockwise 180 degree rotation
    - Enables division for the aorta and pulmonary trunk
    - Creates great arteries and aortopulmonary septum
  - Malrotation of aortopulmonary septum
    - May cause tetralogy of Fallot
    - Septum pulls anteriorly and superiorly
    - Causes aorta
      - To be larger and rotated
      - To override the ventricular trabecular septum
      - Malalignment contributes to a right ventricle (RV) outflow tract obstruction
• Failure in development of ductus arteriosus
  o May result in an absent pulmonary valve due to the increased blood flow in the right side of heart
  o Increased blood flow and pressure to the right side of the heart results in dilation of the pulmonary artery (PA) branches

Anatomy (See illustration below)


• Aorta overrides ventricular septum
  o Enlarged Aorta
  o Straddling or overriding VSD (Number 3 in above illustration)
    ▪ Large malaligned septal defect
    ▪ Non-restrictive
• Right ventricle outflow tract
  o Obstructed
  o Infundibular trabeculae malalignment
• Right ventricular hypertrophy
  o Results from pressure load
    ▪ Generated from work of RV to overcome outflow tract obstruction
    ▪ Mass and physiology similar to LV
• Absent Pulmonary Valve (Number 1 in above illustration)
  o Dilation of pulmonary valve annulus
  o Functionally absent valve

Tetralogy of Fallot with Absent Pulmonary Valve
Dilated pulmonary artery branches (Number 2 in above illustration)
  o Unrestricted blood flow through the pulmonary arteries
  o Absent ductus arteriosus
  o Results in dilation of the pulmonary artery branches
  o Secondary compression of the airway may occur, results in bronchomalacia (See illustration below for relationship between pulmonary arteries and bronchi)

Dilated Pulmonary Arteries in TOF/APV


Physiology
  • Severity of symptoms associated with TOF/APV pulmonary valve (TOF/APV) vary
    o Depend on the degree of pulmonary artery dilation
    o Dilation of the pulmonary arteries results from the absence of a pulmonary valve
    o Mild dilation
      ▪ Mild symptoms
      ▪ Very little involvement of the bronchial tree and small airways
    o Severe dilation
      ▪ Compresses the bronchial tree and small airways
      ▪ Precludes normal growth of the airways
      ▪ Ultimately, compromises ventilation
  • Respiratory distress in small infants and neonates
    o Require more intervention and airway management than larger infants, children and adults
    o Airway compression
      ▪ Leads to significant respiratory distress
      ▪ May cause significant air-trapping
• Leads to hypercarbia, hypoxemia
• Increasing respiratory symptoms
  o Preoperative intubation/ventilation associated with longer postoperative ventilator requirements and mortality
• Ventilation-perfusion mismatch
  o From intrapulmonary and intracardiac shunting
  o Right-to-left shunting at the ventricular level
    ▪ Secondary to severe right ventricular outflow obstruction
    ▪ Causes hypoxemia
    ▪ Less common
  o Most patients with well-balanced pulmonary blood flow

**Procedures**

- Diagnostic evaluation of pulmonary pathology includes:
  o Chest x-ray to assess hyper expansion of the lung
  o Echocardiography to determine the location and extent of pulmonary artery dilation
  o Computerized tomography (CT) scan and Magnetic Resonance Imaging (MRI) are helpful to define sites of airway compression and arterial dilation
  o Bronchoscopy to visualize the degree of airway compression
  o Cardiac catheterization with angiography to delineate the degree of peripheral pulmonary artery dilation

• Medical management
  o Manage airway compression
    ▪ Maintain neonate in the prone position as tolerated to improve ventilation
    ▪ Gravitational force often allows the pulmonary arteries to fall forward and away from bronchi
    ▪ Decreases compression on the bronchi
  o Provide positive pressure ventilation

• Surgical management
  o Depends on severity of symptoms
    ▪ Asymptomatic patients
      • Scheduled for elective surgery
      • Scheduled shortly after diagnosis
    ▪ Severe respiratory compromise
      • Neonatal surgery indicated
      • Timing driven by preoperative presentation
  o Surgical repair varies
    ▪ Depends on severity of pulmonary artery dilation
    ▪ Manageable or very mild respiratory compromise
      • Native PA left in place
      • Reduction pulmonary arterioplasty performed
        o Reduces size of the main and branch pulmonary arteries
- A Le Compte maneuver may be indicated
  - Dilated pulmonary artery placed posterior to the aorta
  - Reduces compression on the airway
- Severe distress from airway compression (See illustration below for TOF repair with conduit)
  - Valved pulmonary homograft
    - Replaces dilated main pulmonary artery
    - Controls flow through the pulmonary valve annulus
  - Reduction arterioplasty on branch pulmonary arteries

Tetralogy of Fallot Repair with Right Ventricle to Pulmonary Artery Conduit


- Patch closure of VSD
  - All patients with TOF
  - Primary surgical goal

**Postoperative Risk Factors/Specific Considerations** (See Neonatal Guidelines and Peds/Neo Guidelines for Post-operative Care)
- Right ventricular dysfunction
  - May result from right ventriculotomy
    - Assess for signs of diastolic dysfunction
      - Elevated RA pressures
      - Tachycardia
      - Hypotension
- Management
  - Time
• RV afterload reduction
  - Milrinone
  - Inhaled Nitric Oxide (iNO)
• Inotropic support as indicated
• Arrhythmias (See Peds/Neo Problem Guidelines on Arrhythmia Management)
  - Temporary pacing wires post-operatively
  - Right bundle branch block most common
  - Complete heart block requiring permanent pacing
  - Junctional ectopic tachycardia (JET)
    - Degree of hemodynamic instability related to the degree of RV dysfunction prior to the arrhythmia
• Pulmonary complications
  - Common throughout fetal and neonatal development
  - Dilated PAs compress developing trachea and bronchi
    - Often leads to tracheomalacia and bronchomalacia
    - May require bronchoscopy and/or otolaryngology evaluation
    - Produces airway obstruction and respiratory distress (atelectasis and pneumonia)
  - Respiratory complications often the cause of death (not cardiac defect)
    - Initial presentation mild with medical management = surgical mortality of 20-40%
    - Initial presentation with severe pulmonary complications = increased surgical mortality as high as 75%

**Long Term Problems**
• Life-long cardiology follow-up required
• Pulmonology follow up indicated for pulmonary complications
• Airway compression at the tracheal and bronchial levels
  - May require tracheostomy and long term mechanical ventilation
  - Key role in postoperative morbidity and mortality
  - Persistent distal airway compression increases mortality risk
    - Endobronchial stents may be used, but difficult to place in distal airways
    - Potential for requiring home oxygenation
      - Often out-grown by age 4
      - May result in recurrent pneumonias
• Pulmonary regurgitation with pulmonary valve replacement
  - May lead to increased RV volume load and potential for arrhythmias
  - RV compression of LV and decreased cardiac output
  - Persistent PA dilation and airway distress
  - Exercise intolerance
• Pulmonary conduit replacement
• Arrhythmias (See both Adult and Peds/Neo Guidelines on Arrhythmia Management)
  - Possible pacemaker placement for heart block
  - Ventricular arrhythmias
Sudden cardiac death

Genetic/syndrome concerns
- DiGeorge Syndrome (22q11 deletion)
  - Increased incidence with conotruncal defects (TOF, Truncus arteriosus)

References:


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