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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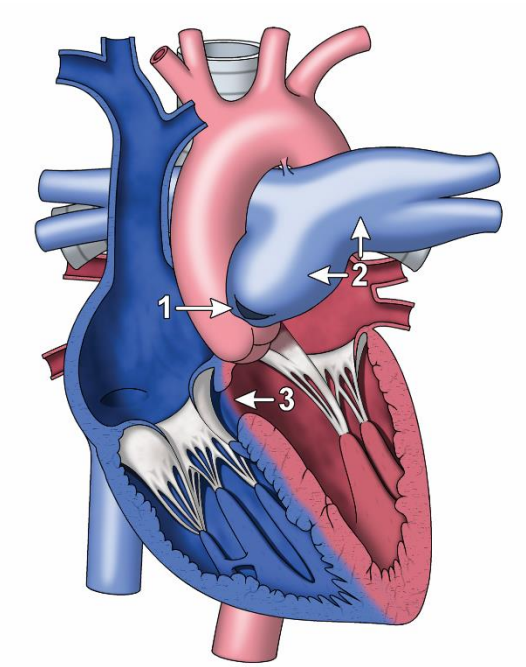
Embryology

- 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
    - May result in an absent pulmonary valve due to the increased blood flow in the right side of heart
    - Increased blood flow and pressure to the right side of the heart results in dilation of the pulmonary artery (PA) branches

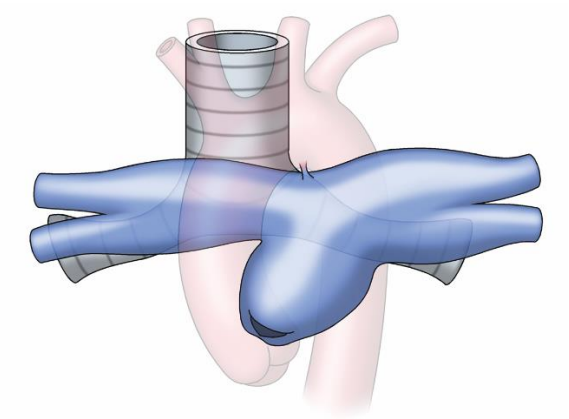
Anatomy (See illustration below)



Tetralogy of Fallot with Absent Pulmonary Valve

- Aorta overrides ventricular septum
    - Enlarged Aorta
    - Straddling or overriding VSD (Number 3 in above illustration)
        - Large malaligned septal defect
        - Non-restrictive
- Right ventricle outflow tract
    - Obstructed
    - Infundibular trabeculae malalignment
- Right ventricular hypertrophy
    - 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)
    - Dilation of pulmonary valve annulus
    - Functionally absent valve

- Dilated pulmonary artery branches (Number 2 in above illustration)
    - Unrestricted blood flow through the pulmonary arteries
    - Absent ductus arteriosus
    - Results in dilation of the pulmonary artery branches
    - 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

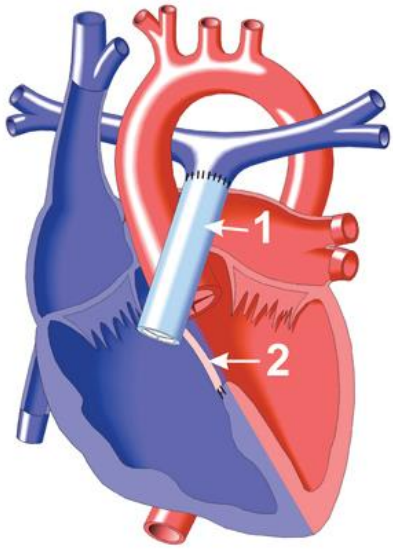
- Severity of symptoms associated with TOF/APV pulmonary valve (TOF/APV) vary
    - Depend on the degree of pulmonary artery dilation
    - Dilation of the pulmonary arteries results from the absence of a pulmonary valve
    - Mild dilation
        - Mild symptoms
        - Very little involvement of the bronchial tree and small airways
    - 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
    - Require more intervention and airway management than larger infants, children and adults
    - Airway compression
        - Leads to significant respiratory distress
        - May cause significant air-trapping

            - Leads to hypercarbia, hypoxemia
            - Increasing respiratory symptoms
        - Preoperative intubation/ventilation associated with longer postoperative ventilator requirements and mortality
- Ventilation-perfusion mismatch
    - From intrapulmonary and intracardiac shunting
    - Right-to-left shunting at the ventricular level
        - Secondary to severe right ventricular outflow obstruction
        - Causes hypoxemia
        - Less common
    - Most patients with well-balanced pulmonary blood flow

Procedures

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

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