Tetralogy of Fallot-Pulmonary Atresia (TOF-PA) Guideline

What the Nurse Caring for a Patient with CHD Needs to Know

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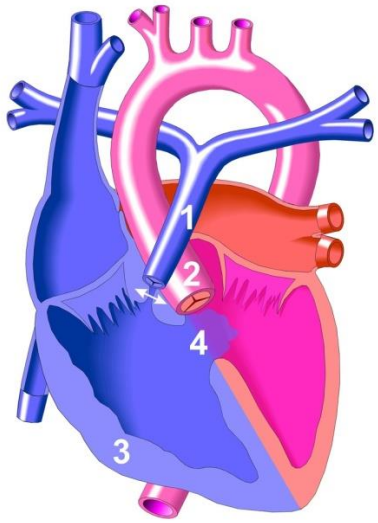
Embryology

- Tetralogy of Fallot (TOF)
  - Most common cause of cyanotic congenital heart disease (CHD)
  - Incidence of 7-10% of all CHD
  - TOF-PA
    - Accounts for about 2% of CHD; incidence of 0.07 per 1000 live births
    - Accounts for 20.3% of all forms of TOF
    - With multiple aortopulmonary collaterals (MAPCAs)
      - Is the most extreme form of TOF
      - Accounts for 20% of all cases of TOF
- Normal development
  - Lungs develop from foregut and carry nutrient supply from the paired dorsal aorta
  - Paired 6th aortic arches give rise to branches that fuse with the pulmonary vascular tree at 27 days gestation
  - Branches from the descending thoracic arch regress and 6th aortic arch enlarges
  - Aorta and pulmonary arteries (PAs) form from the distal bulbus cordis
  - Truncus arteriosus (TA) positioned above the right ventricle (RV)
  - Bulbotruncal ridges separate the great arteries

    - Aortic component rotates posteriorly
- Abnormal development of TOF-PA
    - Occurs between 5th-6th week gestation
    - Faulty rotation of bulbus-truncus results in incomplete transfer of aorta above the left ventricle (LV)
    - Posterior malalignment of infundibular septum results in ventricular septal defect (VSD)
    - Infundibular stenosis developmental theories
        - Anterior displacement of the bulbotruncal region, or
        - Underdevelopment of the subpulmonic infundibulum resulting in conal septum hypoplasia
- The RV outflow obstruction often multi-level
    - Anterior and cephalad deviation of the infundibular septum results in subvalvar obstruction
    - Hypertrophy of muscular bands can cause further subvalvar obstruction
    - Pulmonary valve (PV) annulus usually hypoplastic, but may be normal size
    - PV can be biscuspid and stenotic
    - May have supravalvar narrowing in the main pulmonary artery (MPA) at the sinotubular ridge
    - Further obstruction at the branch PAs due to hypoplasia or focal areas of stenosis
        - Commonly at the proximal branch pulmonary arteries
        - Especially ?Usually (better word?) the proximal left pulmonary artery (LPA) near the site of ductal insertion
- Associated genetic syndromes
    - Genetic testing
        - Recommended for all patients with TOF
        - Must be done prior to cardiopulmonary bypass (CPB)
    - Conotruncal defect
    - High incidence of 22q11 deletion (DiGeorge Syndrome)
        - Up to 50%

Anatomy (See illustration below for TOF)

- Characterized by the combination of four anatomic malformations:
    - Ventricular Septal Defect (VSD) (Number 4 in illustration below)
    - Overriding aorta, overriding the muscular ventricular septum (Number 2 in illustration below)
    - Obstruction of RV outflow tract (Number 1 in illustration below)
    - PAs usually confluent (Number 3 in illustration below) #3 looks like it refers to RV, not PAs
    - RV hypertrophy (Area of arrows in illustration below)



Tetralogy of Fallot

- TOF with PA
  - More severe anatomical variant
    - Solid tissue forms in place of the pulmonary valve
    - Prevents any valve opening
  - Pulmonary blood flow occurs through the PDA
    - About 70% of TOF with PA
  - PAs typically confluent
    - Branch PAs confluent in 85%
    - Non-confluent in 15%
- TOF with PA and MAPCAs
  - Pulmonary blood flow is multifocal
    - Via MAPCAs
    - PAs often non-confluent (70%)
  - MAPCAs
    - Typically arise from the descending aorta
    - May arise from any vessel including:
      - Ascending aorta
      - Head and neck vessels
      - Coronary arteries
  - MAPCAs
    - Create highly variable patterns:
      - PA size and arborization
      - Origin of collateral vessels
      - Number of vessels

        - Course of vessels
    - Connections between the PA and collaterals
        - Unpredictable
        - Change as the patient grows.
    - Can become irregularly shaped, thickened, kinked, or stenotic

Physiology

- In TOF
    - Four cardinal anatomic structures and degree of presentation
        - Determine physiology
        - Determine clinical presentation
    - Degree of RV outflow tract obstruction determines
        - Pulmonary blood flow
        - Degree of left to right shunting through the VSD
        - Degree of cyanosis
- In TOF with PA
    - Complete obstruction of the RV outflow tract obstruction
        - Must have alternate source of blood flow to the pulmonary arteries
        - May be either via the PDA or by MAPCAs
    - Pulmonary blood flow via the PDA
        - Prostaglandins (PGE) required to ensure pulmonary blood flow
    - Pulmonary blood flow via MAPCAs
        - May or may not require PGE; depends on:
            - Anatomy of the MAPCAs
            - Presence of PDA
- Clinical Manifestations
    - Cyanotic at birth
        - Degree of cyanosis depends on PDA and MAPCAs
    - Heart sounds
        - Murmur
            - Usually not heard
            - May be a faint continuous murmur of PDA/MAPCAs
        - Single, loud S2
    - Electrocardiogram (ECG)
        - RV hypertrophy with right axis deviation
        - Prominent R waves anteriorly and S waves posteriorly
        - Upright T wave in V1
        - May also see a qR (?QR) pattern in the right sided chest lead
    - Chest X-ray
        - Normal-sized, boot-shaped heart
        - Decreased pulmonary vascular markings
        - A concavity in the region of the main pulmonary artery
        - Right-sided aortic arch – 26-50%
    - Echocardiogram (ECHO)
        - Parasternal-long axis view
            - Large aortic valve (AV) that overrides a large malalignment VSD

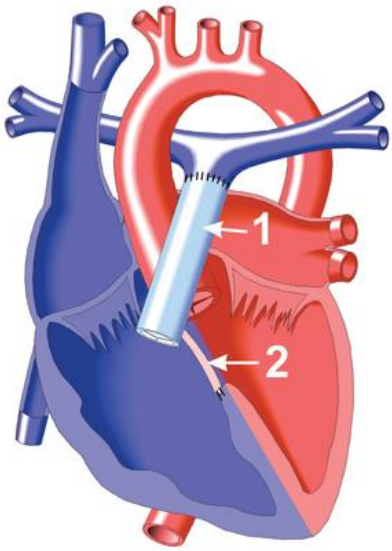
        - Color flow demonstrates lack of patency of RV outflow tract
        - Suprasternal and high parasternal views evaluate:
            - Pulmonary trunk
            - Right and left pulmonary artery size
            - Confluence of PAs

Procedures and Interventions

- Diagnostic Procedures
    - Required as anatomy of the PAs and the source of pulmonary blood supply may vary widely
    - 2-dimenasional (2-D) ECHOwith color flow and 2-D doppler
        - Main diagnostic tool
        - Identifies
            - Sources of PA blood flow-includes PDA and MAPCAs
            - Significant hypoplasia of the central pulmonary arteries
            - Presence of a small PDA
                - Highly predictive of the presence of MAPCAs
                - If present, further imaging by MRI or angiography likely
    - Magnetic Resonance Imaging (MRI)
        - Non-invasive tool
        - Visualize PAs and collateral supplies
    - Cardiac catheterization and angiography
        - Delineate all sources of pulmonary blood supply
        - Facilitates surgical planning
- Interventions
    - Cardiac catheterization
        - Diagnostic
            - Evaluation for surgical intervention
            - Identify sources of/obstruction to pulmonary blood
            - Evaluate ventricular size, structure
        - Intervention
            - Initial evaluation of RV/PA connection
                - Possible radiofrequency ablation (RFA) of membranous PV
                - Balloon dilation of pulmonary stenosis (PS)/pulmonary atresia
                - Stent placement
                    - PV annulus
                    - PDA
            - Repeat catheterizations
                - Balloon dilation/stent placements in stenotic pulmonary artery segments
                - Coil embolization dual source of pulmonary blood supply
                - Coil embolization of MAPCAs
    - Surgical repair
        - Options depend on PA anatomy and presence/extent of MAPCAs

- Single stage repair
  - Considered when PAs confluent and of good size
  - MAPCAs
    - Ligated at the aorta
    - Mobilized toward the posterior mediastinum to construct a pulmonary artery confluence
    - Conduit placed between confluence and RV
  - PAs reconstructed to relieve any surgically accessible stenotic areas
  - VSD closure
  - Mortality rate - 5-20%
- Staged repair
  - Depends on PA anatomy
    - May be required if PAs
      - Hypoplastic
      - Non-confluent
      - Supplied by extensive MAPCAs.
  - Stage 1 – Palliative shunting
    - Induces enlargement and growth of the native PAs
    - Shunt types - Blalock-Taussig shunt, central shunt, or RV to PA conduit
  - Stage 2 Early unifocalization
    - Direct MAPCAs into a central pulmonary artery confluence
    - Improve long-term outcomes
      - Maximize the recruitment of lung segments
      - Increase likelihood of definitive repair
      - Eliminate dual blood supply to a lung segment
        - Coil occlude MAPCA in cardiac catheterization laboratory (cath lab)
        - Ligate at time of unifocalization
    - Objectives
      - To recruit as many of the perfused lung segments as possible
      - Maximize the cross-sectional area of the pulmonary vascular bed
      - Manage unprotected lung segments with a large blood supply
        - At risk for developing pulmonary vascular disease by four to six months of age if untreated
  - Stage 3- Final stage (See illustration below)
    - Complete intracardiac repair with VSD closure (Number 2 in illustration below)
    - Placement/replacement of a RV to pulmonary artery conduit (Number 1 in illustration below)

- PA reconstruction as needed to meet following requirements:
  - Central pulmonary arterial area should be greater than 50% of normal
  - Presence of predominantly left-to-right intracardiac shunting
  - Equivalent of an entire lung must be supplied by the central pulmonary artery confluence
  - Stenotic lesions in the pulmonary artery outflow must be addressed



Repair of Tetralogy of Fallot with RV to PA Conduit

- Surgical placement of a pulmonic valve may be necessary for patients with RV to PA conduits if substantial RV dilation and dysfunction occurs

Specific Considerations

- Preoperative
  - Pulmonary blood flow supplied by:
    - PDA, MAPCAs, or both
    - Alternative sources of pulmonary blood flow accounts for variable clinical presentation
  - Neonates
    - With insufficient pulmonary blood flow, usually present with:
      - Cyanosis - PGE necessary to maintain ductal patency to improve/maintain pulmonary blood flow
      - Hypoxemia

        - Metabolic acidosis
    - With adequate pulmonary blood flow
        - Large, unobstructed MAPCAs
            - With unrestricted blood flow
            - May lead to congestive heart failure
        - As pulmonary vascular resistance (PVR) decreases
            - Pulmonary blood flow may become excessive
            - Results in congestive heart failure
        - MAPCAs may provide pulmonary blood flow, but are prone to stenosis
    - Require screening for chromosomal anomalies
        - Common with conotruncal defects
        - Frequently see 22q11 deletion (DiGeorge syndrome)
            - Abnormal function of parathyroid glands leads to hypocalcemia
            - Immunodeficiency from abnormal T-cell-mediated response predisposes to increased infection risk
            - Physical defects include:
                - Palatal defects causing feeding difficulties
                - Kidney abnormalities
                - Gastrointestinal issues including abnormal motility which may lead to constipation
                - Dysmorphic facies (microstomia, micrognathia, unusually shaped ears, long nose)
            - Learning and psychiatric disorders
- Intraoperative
    - Operative goals:
        - Tailored to specific anatomy
        - Provide adequate, separate pulmonary and systemic circulations
    - Irradiated blood only if DiGeorge or absent thymus
    - Anticipate coagulopathies with severe cyanosis and polycythemia
- Postoperative (See Peds/Neo Problem Guidelines for Postoperative Care)
    - Concerns differ depending on repair
        - Palliation versus correction
        - Multi-staged repair versus one-stage repair
    - Residual defects
        - VSD or VSD patch leak
        - RV outflow tract obstruction
    - RV dysfunction may result in low cardiac output; may be caused by:
        - Increased RV volume loading
        - Ventriculotomy if performed
        - Lower compliance of neonatal myocardium
    - Arrhythmias (See Peds/Neo Problem Guidelines for Arrhythmia Management)
        - Complete Heart Block (CHB)
            - Requires temporary pacing
            - Possible permanent pacemaker, incidence is rare

        - Junctional ectopic tachycardia (JET)
            - Potential for significant hemodynamic compromise
            - Reduce degree of hemodynamic impairment
                - Early recognition
                - Prompt treatment
                    - Cooling to core temperature less than 36 degrees
                    - Antiarrhythmic mediations
    - Elevated RV pressure
        - May result from residual defects
            - Stenosis in pulmonary arteries
            - Stenosis at anastomosis sites
            - Residual MAPCAs
        - If prolonged, cardiac catheterization may be required
            - Dilation of stenotic pulmonary arteries
            - Embolization of residual MAPCAs
    - Respiratory complications
        - Increased occurrence with unifocalization of MAPCAs
        - Bronchospasm related to dissection around bronchopulmonary tree
        - Reperfusion injury in patients with preoperative stenosis of MAPCAs
        - Pulmonary complications such as pneumonia, pulmonary hemorrhage, large airway compression
        - Prolonged respiratory failure requiring prolonged ventilation
    - Genetic syndrome
        - 22q11 deletion (DiGeorge)
            - Anticipate hypocalcemia
                - May require frequent calcium replacement or infusion
            - Immune deficiencies
                - Require use of irradiated blood
                - Increased incidence of infections (See Peds/Neo Problem Guidelines for Infection Prevention)
        - Provide parental education and support

Routine Care

- Lifelong disease requires careful follow-up through adulthood
    - Follow-up at least annually with adult CHD (ACHD) trained cardiologist/NP
    - Potential for additional surgical and interventional procedures
- Infants require frequent follow-up by pediatric cardiologist/NP trained in CHD
    - Prior to and evaluation of surgical intervention(s)
    - ECHO
        - Monitor RV pressure and function
        - Monitor pulmonary circulation
            - Conduit function
            - Evaluation for increasing stenosis/flow in MAPCAs
            - Evaluation for development of aortopulmonary collaterals
    - Repair with RV to PA conduits
        - Require conduit replacement

        - Risk for conduit stenosis and/or conduit valve degradation
    - Cardiac catheterization
        - Hemodynamic evaluation of RV function, PA stenosis
            - Intervention balloon dilation of PAs/ stent placement
            - Coil MAPCAs.
- Subacute Bacterial Endocarditis prophylaxis (See American Heart Association recommendations for Adult and Pediatric SBE Prophylaxis, 2015)

Long-term Complications/Problems (See Adult Problem Guidelines on Arrhythmia Management, Ventricular Dysfunction)

- Abnormal RV physiology secondary to chronic pulmonary regurgitation
    - Ventricular arrhythmias
    - Decreased RV compliance
    - Need for PV/conduit replacements
    - Exercise Intolerance
- Related to DiGeorge
    - Learning disabilities
    - Behavioral & mental health problems
    - Immune disorders
    - Poor vision and hearing
    - Velopharyngeal insufficiency
    - Mypoathic facies
    - Short stature

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