Total Anomalous Pulmonary Venous Return Guideline
What the Nurse Caring for a Patient with CHD Needs to Know

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Embryology
- Rare congenital heart defect occurring in 0.6 to 1.2 per 10,000 live births
- Incidence between 0.7 and 1.5 % of all CHD
- Normal development
  - Lung buds and systemic venous plexus formed at the same time
    - Both drain into the common cardinal and umbilicovitelline venous
    - Lung drainage system becomes the two right and two left pulmonary veins
    - All four join into pulmonary vein confluence (Common pulmonary vein)
  - Portion of the common pulmonary vein incorporated into the wall of the left atrium (LA)
- Total Anomalous Pulmonary Venous Return (TAPVR)
  - Persistent patency of primitive systemic veins
    - Causes failure of pulmonary venous development
    - May lead to persistent connections of the pulmonary venous system to the systemic veins
    - Can occur at almost any point in the central cardinal or umbilicovitelline venous systems
  - Disruption of both cardiac and abdominal vicera early in embryology results in the characteristic congenital anomalies
    - Associated with heterotaxy, particularly with asplenia
Thoracic lymphangiectasia and pulmonary congestion

Anatomy
- Supracardiac (See illustration below)
  - Pulmonary veins connect to right superior vena cava (SVC), azygous vein, left SVC or innominate vein
  - Accounts for 50% of cases

Supracardiac Total Anomalous Pulmonary Venous Return

- Cardiac (See illustration below)
  - Pulmonary veins connect directly to the right atrium (RA) or the coronary sinus (CS)

Intracardiac Anomalous Pulmonary Venous Return

- Infracardiac (See illustration below)
  - Pulmonary venous connect below the diaphragm to intra-abdominal veins
    - Includes portal venous system, ductus venosus or IVC

Infracardiac Total Anomalous Pulmonary Venous Return

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- Mixed
  - One of the main lobar pulmonary veins connects to a systemic vein
  - Remaining three veins connect normally
- Obstructed (See above illustration)
  - Most common with infracardiac TAPVR
  - Can occur at any point in the anomalous pathway
  - Causes changes in the pulmonary arterioles
- Increase in arterial muscularity
- Extension of muscle into smaller and more peripheral arteries
- Veins likely to be thick walled with intimal fibrous hyperplasia

- Associated anomalies
  - Tetralogy of Fallot
  - Double Outlet Right Ventricle
  - Hypoplastic Left Heart Syndrome
  - Endocardial fibroelastosis of the LV

**Physiology**

- Unobstructed
  - Increased pulmonary blood flow
    - Right heart volume load from pulmonary venous return to the right heart
    - Creates left to right shunt physiology
      - Pulmonary vascular resistance (PVR) decreases in the first few week of life
      - Causes increased Qp:Qs leading to heart failure symptoms
      - Untreated, may lead to pulmonary vascular changes and elevated PVR
  - At risk for pulmonary hypertensive crises postoperatively (See Peds/Neo Problem Guidelines on Pediatric Pulmonary Hypertension)

- Obstructed
  - Severe pulmonary venous congestion and small cardiac silhouette
    - Cyanosis and respiratory distress usually presents within minutes to hours after birth
    - Cyanosis profound
    - Followed by:
      - Cardiogenic shock
      - Severe metabolic acidosis
    - Causes right ventricular hypertrophy (RVH)
  - Immediate surgical repair required
  - Pre-operative management:
    - Optimize increased PVR with intubation
    - Correct metabolic and respiratory acidosis
    - Provide sedation and paralysis as needed
    - Consider ECMO prior to surgical repair

**Procedures and Interventions**

- Diagnostic Procedures
  - Chest radiograph:
    - Prominent right heart with a “snowman” appearance on a frontal view
    - ‘Ground glass’ appearance similar to neonatal respiratory distress syndrome
  - Echocardiography (ECHO)
    - Normal pulmonary venous connections to the left atrium (LA) visualized
Supracardiac lesions can present with:
- Common ascending collecting vein
- Dilated superior vena cava (SVC)
Infracardiac lesions can present with:
- Connection of the common descending collecting vein with the hepatic or portal vein
- Or dilated inferior vena cava (IVC)
Visualization of a connection between the pulmonary venous system and the RA or coronary sinus
- Dilated RA and RV
  - Right-to-left atrial level shunting
  - Angiography
    - May be used when individual pulmonary veins and vertical veins not visualized on ECHO
    - May be indicated if more hemodynamic information is required
  - Computed Axial Tomography (CT) or Magnetic Resonance (MR) angiography
    - CT may be needed to visualize the lung parenchyma and airways
    - MR requires less ionizing radiation, but requires more time and sedation may be needed
    - Both require IV contrast for optimal visualization of the vasculature
- Surgical correction
  - Required
  - Recommended regardless of the degree of obstruction
    - Timing depends on degree of obstruction and condition of the patient
    - Urgent surgical intervention needed if veins are completely obstructed
  - Surgical approach
    - Via median sternotomy
    - Performed under cardiopulmonary bypass with circulatory arrest
    - Surgical procedure varies depending on the anatomy of the defect
      - Supracardiac and infracardiac TAPVR with a common vertical vein
        - Anastomosis formed between the pulmonary venous confluence and the LA
        - Vertical vein ligated and divided
      - Pulmonary veins drain directly into the SVC
        - Intracardiac baffle formed to channel the blood from the RA, across the atrial septum to LA
      - Intracardiac TAPVR to the CS
        - CS and partition between the sinus and RA are incised, and connected to LA
      - Intracardiac TAPV to RA
        - Interatrial septum reconstructed to close the atrial septal opening and direct blood flow from the pulmonary veins directly to LA
  - Rate of reoperation
• Between 10 and 15%
• Due to stenosis of individual pulmonary vein and surgical anastomosis site
• Rarely required after a year following surgical repair

Specific Considerations
• Factors that determine severity of the symptoms are:
  o Presence of other anomalies
  o Presence and severity of obstruction to pulmonary venous drainage
  o Degree of obstruction at the atrial septal level
• Unobstructed pulmonary venous drainage and unrestricted atrial septal communication
  o Signs/symptoms
    ▪ Congestive heart failure
    ▪ Progressive right heart dilation
    ▪ Pulmonary hypertension (See Peds/Neo Problem Guidelines on Pediatric Pulmonary Hypertension)
  o If not managed will lead to irreversible pulmonary vascular obstructive disease (PVOD)
    ▪ Symptoms for PVOD include:
      • Progressive tachypnea
      • Cyanosis
      • Right–sided heart failure
• Obstruction of pulmonary venous drainage
  o Neonates with infracardiac TAPVR
  o High pulmonary pressure with large right-to-left shunt
  o Rapid progressive hypoxemia and hemodynamic collapse
  o Diagnosis
    ▪ Careful attention to history and physical examination findings, chest radiograph and echocardiogram
    ▪ Physical exam: Evaluate for signs that indicate pulmonary blood flow
      • Unobstructed = increased flow
        o Parasternal lift
        o Widely split second heart sound
        o Pulmonary flow murmur and diastolic murmur
        o Mild tachypnea and cyanosis
      • Obstructed pulmonary blood flow
        o Signs of pulmonary edema with or without evidence of hypoperfusion
    ▪ Chest radiograph:
      • Pulmonary arteries appear engorged with or without pulmonary edema
      • Snowman or figure-of-eight cardiac shadow
        o Seen later on in infancy
          ▪ Pulmonary venous connections enlarge
          ▪ Thymus diminishes in size
- Echocardiography:
  - Defines anatomy of TAPVR
    - RV dilatation
    - Absence of pulmonary veins draining into LA
    - Presence of anomalous venous channels or turbulent flow in RA
    - Other abnormal systemic venous structures
- Cardiac catheterization:
  - Useful in patients with multiple cardiac lesions
  - If significant pressure gradient found across atrial septum, a balloon atrial septostomy may allow a delay in surgical repair until patient is adequately resuscitated
- Preoperative Care
  - Pulmonary venous obstruction
    - Surgical emergency requires immediate surgical intervention due to:
      - Progressive hypoxemia
      - Systemic hypo-perfusion
      - Hemodynamic instability
      - Emergency repair optimal
    - Medical management
      - Intubation for hyperventilation and 100% fraction of inspired oxygen concentration (FiO2)
        - Decrease pulmonary vascular resistance
        - Maximize oxygen delivery
      - Inotropic support to assist the dilated and dysfunctional right ventricle
      - Correct metabolic acidosis to improve catecholamine responsiveness
      - Pulmonary vasodilators
        - Controversial in preoperative period
        - Include iNO, magnesium sulfate, prostaglandins
        - Requires close monitoring for untoward effects of worsening cyanosis and systemic hypotension
  - Extracorporeal membrane oxygenation (ECMO) [See Peds/Neo Problem Guidelines on Extracorporeal membrane oxygenation (ECMO)]
    - Patients with severe metabolic derangement and pulmonary hypertension
    - Used to stabilize and correct end-organ dysfunction
    - Improves outcome
- Postoperative Care (See Peds/Neo Problem Guidelines on Postoperative Care, Nutrition, Development Care, Pediatric Pulmonary Hypertension)
  - Goals:
    - Improve cardiac output
    - Manage pulmonary hypertension
Prevent pulmonary hypertensive crisis
Maximize respiratory efficiency

Prevention and management of pulmonary hypertensive crisis (See Peds/Neo Problem Guidelines on Pediatric Pulmonary Hypertension)
- Monitor closely
  - Direct pulmonary arterial pressure (PAP) measurements with intra cardiac PA line (normal PAP is <1/3 systemic pressure)
  - If direct monitoring unavailable, monitor for:
    - Unexplained desaturation
    - Tachycardia
    - High central venous pressure
    - Hypotension
- Manage analgesia
  - Provide optimal level of analgesia and sedation
  - Provide neuromuscular blockage as needed
- Manage ventilation
  - Use mechanical ventilation to maintain functional residual capacity
    - Use enriched inspired oxygen
    - Appropriate amount of PEEP
  - Avoid unnecessary endotracheal suctioning which can cause an acute increase of PVR
  - Prevent respiratory insufficiency
- Manage pulmonary pressures
  - Use intravenous vasodilators as indicated
    - Like nitroprusside (Nipride)
    - Watch for systemic hypotension
  - Use inhaled vasodilators
    - Relax constricted pulmonary vascular smooth muscle
    - Like nitric oxide (iNO)
    - Some patients may not respond
- Prevent acidosis
- Mechanical circulatory support/ ECMO (See Peds/Neo Problem Guidelines for Mechanical Circulatory Support (ECMO)
  - Severe cardiac dysfunction
  - Unresponsive pulmonary hypertension
- Avoid overaggressive volume replacement that may lead to excessive pressure elevation
- Maintain optimal heart rate
  - Maximize cardiac output
  - Low cardiac output occurs as a result of a noncompliant left ventricle with less effective stroke volume per heart beat
  - Chronotropic support with temporary pacing as needed
    - Temporary pacing wires placed postoperatively
- Provide myocardial support
  - Inotropes
- Calcium, especially in neonates
- Continuous infusion medications
  - Such as dopamine, epinephrine
  - Afterload reduction
  - Continuous infusion medications
    - Such as milrinone or nitroprusside (Nipride)
    - Continue family support utilizing principles of family centered care

Long Term Problems/Management

- Surgical mortality rate
  - Infants with unobstructed type - between 5% -10%
  - Infants with obstructed infracardiac type - as high as 20%
  - Most common cause of death
    - Cardiac failure due to pulmonary hypertensive crisis (See Peds/Neo Problem Guidelines on Pediatric Pulmonary Hypertension for postoperative management of pulmonary hypertensive crisis)
  - Risk factors for early postoperative mortality
    - Infracardiac drainage
    - Pulmonary venous obstruction
    - Poor preoperative state including persistent acidosis
- Postoperative pulmonary venous stenosis
  - Infracardiac and mixed type - between 6-11%
  - Management
    - Use of absorbable suture
    - Catheter intervention - Balloon angioplasty and endovascular stent placement
    - Surgical - reoperation
      - Usually occurs within the first 6-12 months after repair
      - Due to pulmonary vein stenosis and obstruction at the anastomosis site
    - Progressive pulmonary vein fibrosis
      - Remains an unpredictable rare cause of death
      - Starts within the first year after surgery
- Arrhythmias (See Adult and Peds/Neo Problem Guidelines for Arrhythmia Management)
  - Rare
  - Supraventricular tachycardia (SVT)
  - Junctional tachycardia
  - Reported in long term survivors
- Long term follow-up
  - Goal to identify for timely intervention
  - Monitor for:
    - Adequate growth,
    - Regression of right ventricular dilatation
    - Reversal of pulmonary vascular abnormalities
  - Identify long term psychological and cognitive abnormalities
• Reported following cardiopulmonary bypass
• Varied incidence
• Better outcomes with early intervention

• Long term survival
  o Excellent results related to:
    ▪ Early diagnosis
    ▪ Increasing surgical expertise in small neonates
    ▪ Improvement in postoperative care
    ▪ Treatment of pulmonary hypertensive crisis

Routine Cardiology Care
• Lifelong clinical evaluation with cardiologist trained in congenital heart disease
• Goals of long term follow-up include:
  o Adequate growth
  o Regression of right ventricular dilatation
  o Reversal of pulmonary vascular abnormalities
• Diagnostic tests
  o Electrocardiogram (ECG)
    ▪ Supraventricular arrhythmias most common
  o Exercise stress test
    ▪ Decrease in aerobic exercise capacity, lung volume and chronotropic response may be noted over time in postoperative patients
  o Echocardiogram (ECHO)
    ▪ Identify residual defects or complications
    ▪ Pulmonary vein obstruction / stenosis
      • Most frequent long term complication
      • Generally confirmed with angiography
    ▪ Ventricular function
    ▪ Pulmonary hypertension as evidenced by septal flattening or displacement
    ▪ Residual atrial septal defect
• Lifestyle Monitoring
  o Cholesterol panel
  o Obesity / weight control
  o Tobacco use / exposure
• Education
  o Assess knowledge
  o Review condition and potential complications
  o Discuss lifelong needs
• Pregnancy (See Adult Problem Guidelines for Management of Pregnancy in ACHD)
  o Requires cardiology evaluation prior to pregnancy to review risks
  o Multidisciplinary coordination necessary
References:


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