# Total Anomalous Pulmonary Venous Return Guideline

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

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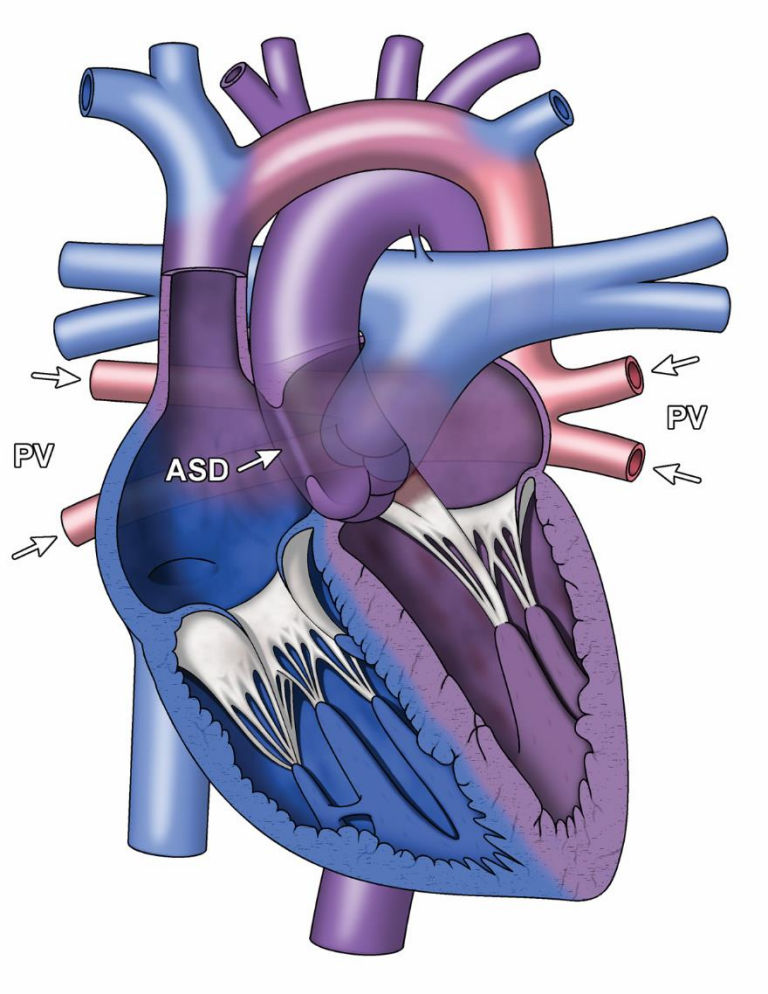
## Embryology

- 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 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

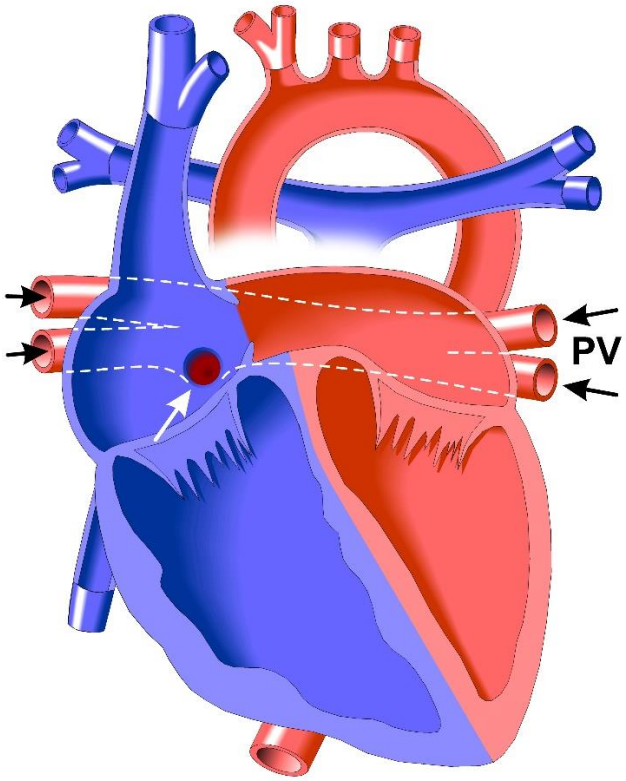
**Anatomy**

- 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



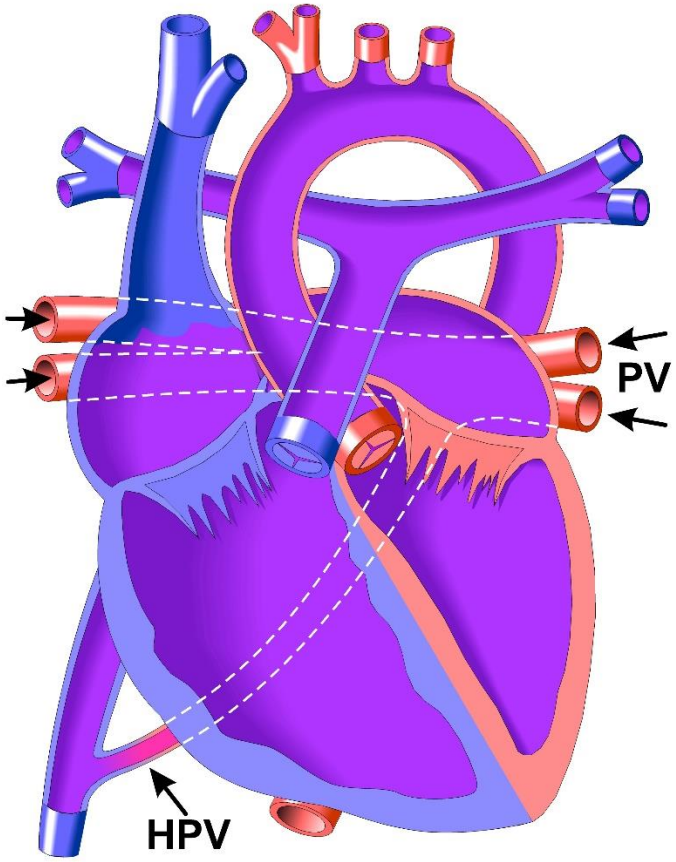

Supracardiac Total Anomalous Pulmonary Venous Return

- 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



- 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:
  - Presence of other anomalies
  - Presence and severity of obstruction to pulmonary venous drainage
  - Degree of obstruction at the atrial septal level
- Unobstructed pulmonary venous drainage and unrestricted atrial septal communication
  - Signs/symptoms
    - Congestive heart failure
    - Progressive right heart dilation
    - Pulmonary hypertension (See Peds/Neo Problem Guidelines on Pediatric Pulmonary Hypertension)
  - 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
  - Neonates with infracardiac TAPVR
  - High pulmonary pressure with large right-to-left shunt
  - Rapid progressive hypoxemia and hemodynamic collapse
  - 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
        - Parasternal lift
        - Widely split second heart sound
        - Pulmonary flow murmur and diastolic murmur
        - Mild tachypnea and cyanosis
      - Obstructed pulmonary blood flow
        - 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
        - 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
    - 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:
    - Adequate growth
    - Regression of right ventricular dilatation
    - Reversal of pulmonary vascular abnormalities
- Diagnostic tests
    - Electrocardiogram (ECG)
        - Supraventricular arrhythmias most common
    - Exercise stress test
        - Decrease in aerobic exercise capacity, lung volume and chronotropic response may be noted over time in postoperative patients
    - 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
    - Cholesterol panel
    - Obesity / weight control
    - Tobacco use / exposure
- Education
    - Assess knowledge
    - Review condition and potential complications
    - Discuss lifelong needs
- Pregnancy (See Adult Problem Guidelines for Management of Pregnancy in ACHD)
    - Requires cardiology evaluation prior to pregnancy to review risks
    - Multidisciplinary coordination necessary

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