Pulmonary Atresia Intact Ventricular Septum (PA-IVS) Guideline

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

Ashleigh Harlow, BSN, RN, CCRN
Clinical Educator, Cardiac Intensive Care Unit
Children's National Health System, Washington, DC

Angela Blankenship, MS, RN, CPNP-AC/PC
Nurse Practitioner, Cardiac Intensive Care Unit,
Nationwide Children's Hospital, Columbus, OH

Justine Mize, MSN, RN, CCRN, CPN,
Professional Practice Specialist, Cardiac Intensive Care Unit,
Children's National Health System, Washington, DC

Megan Matiasek, MS, RN, CPNP-PC/AC
Nurse Practitioner, Cardiac Intensive Care Unit,
Boston Children's Hospital, Boston, MA

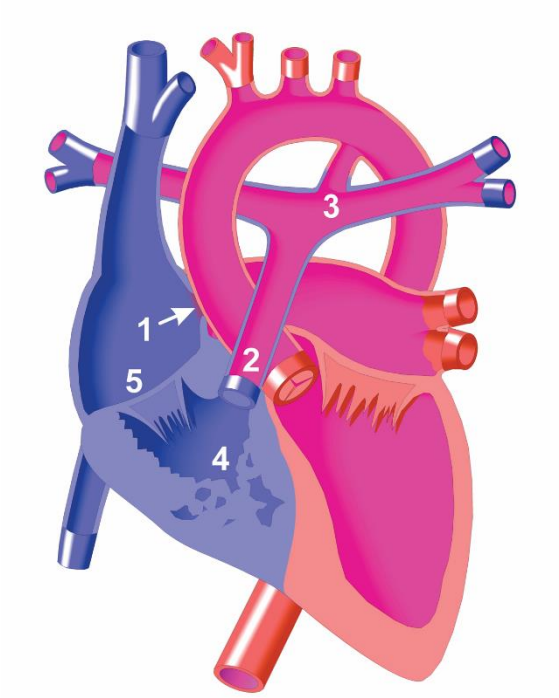
Christine Riley, MSN, APRN, CPNP-AC
Nurse Practitioner, Cardiac Intensive Care Unit,
Children's National Health System, Washington, DC

Embryology

- Accounts for < 1% of all congenital heart defects and 2.5% of critically ill infants with congenital heart disease (CHD)
- Development of PA-IVS varies
    - Thought to occur between 6-10 weeks gestation
    - Specific mechanisms of pulmonary valve (PV) malformation unknown
        - PV leaflet malformations
        - Failure in separation of valve leaflets
    - Inflammatory or infectious processes may contribute
        - Right ventricular outflow tract (RVOT) develops late in embryonic development
        - Leads to obstruction of pulmonary artery (PA) (See number 2 in illustration below)
    - Right ventricle (RV) hypoplasia and tricuspid valve (TV) hypoplasia
        - "Upstream" of the atretic valve
        - Presumably related to aberrant flow patterns through the right side of the heart
            - Blood shunts right to left across the foramen ovalae (FO) (See number 1 in illustration below)
            - Limits growth and development of the bypassed RV
- Coronary artery anomalies common
    - Normal right coronary artery (RCA) circulation flows directly into the coronary sinus

- With PA-IVS most RCA flows through primitive vessels known as Thebesian veins
  - Increased RV pressure
    - Creates retrograde flow through the Thebesian veins
    - Flows back into the coronary circulation
  - Some Thebesian veins communicate directly with the coronary arterial circulation ("coronary fistulas")
  - Others have no obvious connection with the coronary arterial circulation ("coronary sinusoids")

Anatomy (See illustration below for PA-IVS)



Pulmonary Atresia – Intact Ventricular Septum

- Right Ventricle (Number 4 in above illustration)
  - Size and shape of the RV varies along a spectrum
    - In 90% of cases
      - Small cavity size
      - Hypertrophic wall
    - In 50% of cases
      - Severely hypoplastic
  - Three components describe RV

        - Infundibulum (outflow tract), trabeculae (body), and inlet (under tricuspid valve)
        - Description based on the presence and/or absence of structures
    - Volume
        - Estimated using the diameter of TV
        - Smaller RV more likely to display signs of chronic hypertension and ischemia
        - "Ebstein-like" variants
            - 5-10% of cases
            - Regurgitant TV leads to a large, dilated RV
            - Coronary fistulas not associated with this variant
            - Supports theory that coronary fistulas persist under the conditions of a competent TV and hypertensive RV
        - Uhl's anomaly: rare, dilated, parchment-like RV
- Tricuspid Valve (Number 5 in above illustration)
    - Used for surgical plan
        - Consideration of single-ventricle versus biventricular repairs
        - Successful surgical outcomes
            - Predicted most closely by TV z-scores
            - Relative size and volume of the RV
            - Presence or absence of coronary fistulae and RV-dependent coronary circulation
    - Ebstein-like variant
        - TV enlarged and regurgitant.
- Pulmonary Arteries (Number 3 in above illustration)
    - Main pulmonary artery (MPA)
        - Generally normally formed
        - May be somewhat hypoplastic
    - Branch PAs
        - Generally normal
        - Size depends on blood flow

Physiology

- Ductal dependent pulmonary blood flow
    - Requires consistent source of pulmonary blood flow
        - Ductus arteriosus (DA)
            - Prostaglandin infusion
            - Stent in DA
        - Multiple aorto-pulmonary collaterals (MAPCAs)
            - Rare

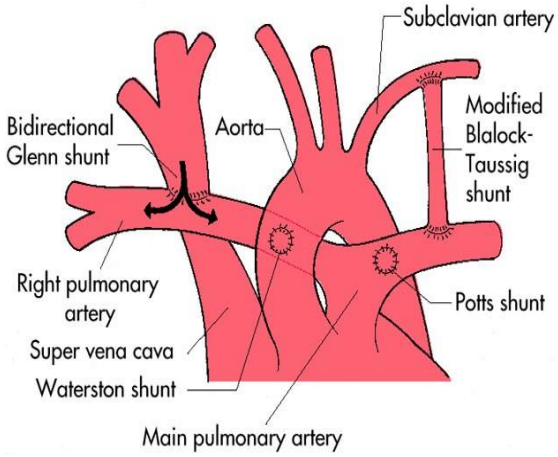
- Atrial level shunting
    - Required for systemic venous return to mix with pulmonary venous return in left atrium (LA)
    - Required for adequate cardiac output (CO)
        - Frequent monitoring of CO
        - Poor CO and perfusion despite a patent PDA
            - Suggests a restrictive atrial septum
            - May require atrial septostomy
- Coronary sinusoids
    - Must determine presence
        - Dependent on the RV for circulation
        - Occurs in patients with a hypertensive RV; not those with tricuspid regurgitation (TR)
    - Determine surgical management vs transplant
- Clinical manifestations
    - Cyanosis at birth
        - May be severe
        - Associated with tachypnea
    - Heart sounds
        - Single S2
        - Soft TR murmur
        - Continuous PDA murmur, may not hear with increased flow
    - EKG: normal QRS axis and LVH
    - Chest X-ray
        - Heart: normal or enlarged from right atrial enlargement
        - Pulmonary vascular markings: decreased
    - Echo
        - Thickened, immobile, atretic PV
        - Hypertrophied RV
        - Patent but small TV
        - Right to left shunt through ASD
        - Pulmonary blood flow - Ductus arteriosus vs aorto-pulmonary shunt collaterals
        - Right and left PA branches normally developed
        - Color doppler flow may show coronary artery fistulas

Procedures and Interventions

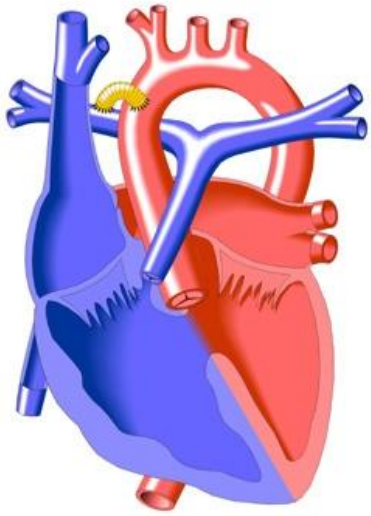
- Management plan (intervention) based on:
    - TV z-score
    - Coronary artery anatomy and perfusion

- RV size
- Catheter based
  - Goal: RV decompression
  - Balloon valvotomy with radiofrequency perforation
    - Membranous PV atresia and a well-developed tripartite RV
    - Contraindicated with the presence of either right or left coronary artery systems with stenosis or occlusion
    - Large coronary sinusoids and a right dominant coronary circulation
      - Precludes RV decompression
      - May lead to coronary flow reversal and myocardial infarction
    - 80% reported success rate
    - 50% will require an additional procedure to provide adequate pulmonary blood flow
    - Procedure
      - Angiography of RV and RVOT performed to determine RV size and presence of coronary sinusoids
      - A radiofrequency wire used to perforate the atretic PV
      - A balloon catheter then used to dilate the valve to up to 120%
  - Ductal stent placement
    - Patient criteria
      - Membranous PV atresia
      - Infundibulum well developed
      - Moderately hypoplastic bipartite RV
    - Procedure
      - Generally performed in conjunction with PV balloon dilation
      - DA accessed via the aorta
      - Stent deployed in DA
- Surgical
  - Goal: RV decompression and growth
  - Surgical pulmonary valvotomy
    - Membranous PV atresia and a well-developed tripartite RV
    - Approximately 1/3 of PA-IVS patients require an isolated surgical pulmonary valvotomy
    - Two-thirds require additional interventions
    - Procedure
      - Direct visualization and surgical division of pulmonary valve leaflets
      - Surgical division of fused papillary muscles and/or anomalous muscle bands
  - Systemic to right PA shunt (See illustration below for types of shunts)

- Membranous PV atresia, an infundibulum that is well developed, and a moderately hypoplastic bipartite RV
- Additional pulmonary blood flow promotes RV growth and compliance
- Procedure
    - Modified Blalock-Taussig shunt: Gortex® between subclavian or innominate artery to the right or left pulmonary artery
    - Central shunt: Gortex® between aortic arch and bifurcation of branch PAs



Types of Shunts



Tetralogy of Fallot with Modified Blalock Taussig Shunt

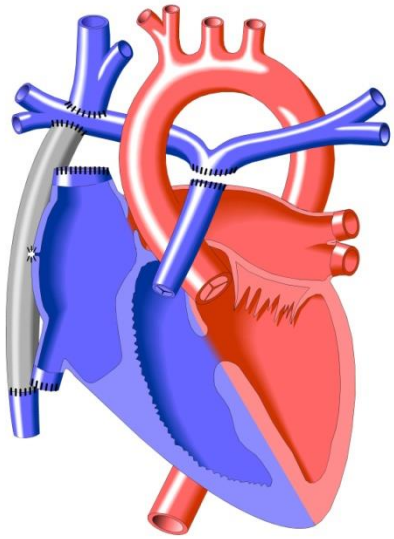
- RVOT repair / atrial septal fenestration

        - Severe RVOT obstruction and moderate RV hypoplasia
        - Slightly less than half of patients who receive a pulmonary valvotomy also require RVOT augmentation
        - Procedure
            - Transannular outflow tract patch
                - Encourages flow across TV
                - Stimulates RV growth and adequate CO
            - Creation of an ASD or fenestrated ASD patch
                - Provides or increases right to left shunt
                - Stimulates RV growth and adequate CO
    - Biventricular pathway
        - Ultimate goal when feasible
        - RV volume and outflow track must be adequate
        - Success rates
            - Variable
            - Based on RV volume and tricuspid valve size (z-score)
        - Procedure
            - RVOT augmentation via right myomectomy
            - ASD closure
            - Ligation (division) of the systemic to pulmonary shunt (if present)
            - May include RV to PA conduit (when main PA absent)
    - One and one-half ventricle pathway
        - Moderately hypoplastic RV or RV dependent coronary circulation
        - Procedure
            - Goals
                - Low RA pressure
                - Pulsatile pulmonary blood flow
                - Improved oxygen saturation
            - RVOT augmentation
            - Partial restriction of ASD
            - Anastomosis of the superior vena cava (SVC) to the PA for passive pulmonary blood flow (bidirectional Glenn)
            - Diversion of a portion of the systemic venous return unloads RV
    - Single ventricle pathway (See illustration of single ventricle surgery with LV as primary pumping chamber below)
        - Indications
            - Severe RV hypoplasia
            - Absent infundibulum
            - Failure of attempted TV growth
            - Adequate sized PAs

- Low pulmonary vascular resistance
- Satisfactory left sided hemodynamics and cardiac output

▪ Benefits

- Morphological LV as the primary pumping chamber (verses other variations of single ventricle physiology that rely on the RV as the systemic ventricle)



Extra cardiac Fontan (Original defect = Tricuspid Atresia)

o Heart transplantation

▪ Neonates with a complete RV dependent coronary circulation

Specific Considerations

- Neonatal
  - o May have ductal dependent pulmonary blood flow
    - ▪ Presentation
      - Systemic to suprasystemic RV pressure
      - Right to left atrial shunting
      - Hypoxia that results in acidosis
        - o Inadequate pulmonary blood flow
        - o Decreased cardiac output
    - ▪ Careful monitoring, including echocardiogram, of pulmonary blood flow and RV pressure as PDA closes
      - May need to start prostaglandin (PGE_1) infusion
        - o Maintain patency of ductus arteriosus (DA)

        - Anticipate side effects of PGE_1 /prepare for immediate intervention
            - Apnea/intubation and ventilation
            - Vasodilation/fluid administration
            - Bradycardia/inotropes
            - Fever/antipyretics
            - Seizures/evaluation
    - Prevent complications
        - Air emboli – ensure that NO air enters IV
        - Dehydration
        - Infection
- Surgical intervention (See Peds/Neo Guidelines for Postoperative Management)
    - Based on RV anatomy and coronary sinusoids
        - Single ventricular palliation vs biventricular repair
            - May be candidate for neonatal transplant
    - Postoperative complications
        - RVOT augmentation
            - Diminutive RV
                - Needs careful attention to RV volume and function
                - Monitor
                    - Oxygen saturation
                    - CO
            - Surgical intervention of RVOT augmentation alone
                - May require additional pulmonary blood flow
                - Up to 50% require addition of a systemic to pulmonary shunt within the first month
        - Systemic to pulmonary shunt
            - Augments pulmonary blood flow at the expense of volume load to the LV
            - Pulmonary overcirculation
                - May be caused by antegrade pulmonary blood flow in addition to shunt flow
                - Systemic hypoperfusion
                - Acidosis
            - Shunt thrombosis (See Peds/Neo Guidelines on Anticoagulant Management)
                - Significant hypoxia
                - Hypotension
                - No response to changes in inotrope administration and/or ventilation maneuvers
                - Management
                    - Anticoagulants
                    - Antithrombotics
                    - ECMO
            - Bidirectional cavopulmonary anastomosis
                - Pulmonary blood flow from Glenn shunt (SVC to PA)

- Positive pressure ventilation can impede pulmonary blood flow
  - Use low (physiologic) positive end expiratory pressure (PEEP)
  - Goal for early extubation
- Optimize pulmonary blood flow
  - Phosphodiasterase inhibitors (Milrinone/sildenafil)
  - Inhaled Nitric Oxide (iNO)

Long Term Considerations

- Anatomic considerations
  - Septal distortion and hypertrophy in the setting of RVH
  - Leads to left ventricular outflow tract obstruction
  - Leads to impaired left ventricular function
- Coronary artery stenosis
  - Increased by prolonged supra-systemic RV pressure; and RV decompression
  - May lead to myocardial ischemia
- Potential catheter and surgery based long-term considerations
  - Balloon Valvotomy
    - Non-compliant RV
    - May reduce pulmonary blood flow
    - Need to surgically provide additional pulmonary blood flow while RV remodels
  - Systemic to pulmonary shunt
    - LV volume overload
    - Pulmonary over-circulation may cause systemic hypoperfusion and acidosis
- Biventricular Repair
  - RV diastolic dysfunction may lead to hypertrophy
  - PV regurgitation may need surgical repair or replacement
  - Conduit stenosis may need surgical replacement
  - TV regurgitation may need surgical repair
  - Systemic venous hypertension
  - Arrhythmias (See Adult and Peds/Neo guidelines for Arrhythmia Management)
    - Related to:
      - Tricuspid regurgitation
      - RA dilation
      - Ventriculotomy
  - Ventricular hypertrophy may lead to elevated coronary vascular resistance
  - Residual pulmonary stenosis
- One and one-half ventricular repair
  - Increased incidence of chylothorax or pleural effusions related to competing sources of pulmonary blood flow.
- Single Ventricle pathway [See Adult Guidelines for Arrhythmia Management, Long Term Effects of Cyanosis (Eisenmenger's Syndrome)
  - Formation of veno-venous collaterals

- Development of pulmonary arteriovenous malformations
- Atrial arrhythmias
- Cyanosis
- Right atrial thrombus
- Cardiac ischemia

Routine Care

- Requires life-long follow-up
  - Pediatric cardiologist
  - Adult cardiologist specialized in ACHD
  - Timing of assessment and diagnostic evaluation
    - Depends on age
    - Depends of function/symptoms
    - At least annual follow-up
  - Follow up
    - Periodic assessment of RV size and function: echocardiogram, transesophageal echocardiogram, nuclear stress testing
    - Evaluation of adequate coronary perfusion: cardiac catheterization, computerized tomography (CT), magnetic resonance imaging (MRI)
    - Evaluation of rate and rhythm: ECG, Holter monitoring, stress testing
- Single ventricle monitoring
  - Inter-stage monitoring (See Peds/Neo Problem Guidelines on Interstage Monitoring)
  - Periodic invasive and non-invasive monitoring
    - Prior to interventions
    - To assess management strategies
    - Includes cardiac catheterizations to evaluate ventricular function
- Anti-coagulation management (if applicable) (See Adult and Peds/Neo Problem Guidelines on Arrhythmia Management)
- Prophylactic antibiotics for dental procedure (See American Heart Association Bacterial Endocarditis Prophylaxis Guidelines for Adults and Children, 2015)

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