

## **Pulmonary Valve**

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

Catherine Baxter, MSN, RN, CPNP-AC  
Nurse Practitioner, Pediatric Cardiac Surgery,  
Levine Children's Hospital, Charlotte, NC

Misty Ellis, MSN, CPNP-PC/AC  
Pediatric Cardiac Intensive Care Nurse Practitioner  
University of Louisville, Kosair Children's Hospital

Victoria Winter RN, MSN, CNS, CCRN  
Clinical Nurse IV, Adjunct Professor,  
Children's Hospital Los Angeles and  
Azusa Pacific University School of Nursing

Louise Callow, MSN, RN, CPNP  
Pediatric Cardiac Surgery Nurse Practitioner,  
University of Michigan, CS Mott Children's Hospital

Mary Rummell, MN, RN, CPNP, CNS, FAHA  
Clinical Nurse Specialist, Pediatric Cardiology/Cardiac Services,  
Oregon Health & Science University (Retired)

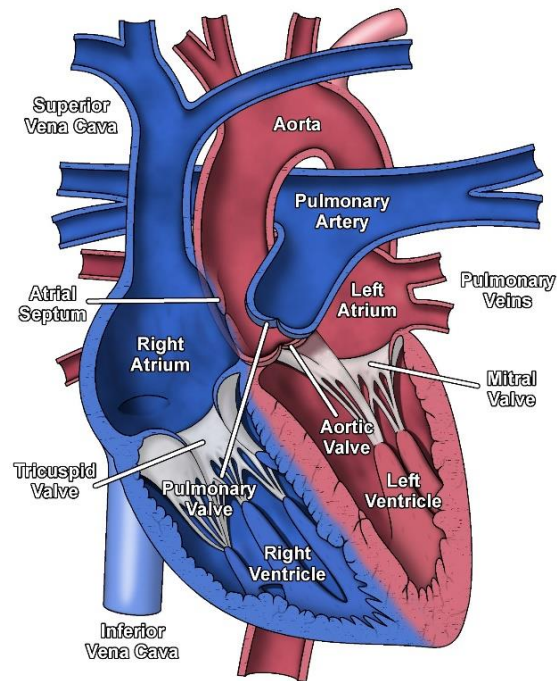
### **Embryology**

- Occurrence:
  - Defects of cardiac valves are the most common subtype of cardiac malformations
  - Account for 25% to 30% of all congenital heart defects
  - Most costly and relevant CHD
  - Wide spectrum of congenital defects in pulmonary valve
- Development of the heart valves occurs during the fourth to eighth weeks of gestation-after tubular heart looping
  - Walls of the tubular heart consist of an outer lining of myocardium and an inner lining of endocardial cells
  - Cardiac jelly, extensive extracellular matrix (ECM), separates the two layers
  - Cardiac jelly expands to form cardiac cushions at the sites of future valves
    - Outflow track (OT) valves = aortic and pulmonic valves
      - Final valves derived from endothelial-mesenchymal cells with neural crest cells from the brachial arches
      - Valves (Semilunar) have 3 equal cusp-shaped leaflets
      - Aortic valve incorporates coronary arteries
    - Atrioventricular (AV) valves = mitral and tricuspid
      - Final valves derived entirely from endocardial cushion tissue
      - Leaflet formed without a cusp

- Two leaflets associated with left ventricle (mitral)
  - Three leaflets associated with right ventricle (tricuspid)
- Coordinated by complex interplay of:
  - Genetics
  - Signaling pathways that regulate cell apoptosis and proliferation
  - Environmental factors
    - Maternal hyperglycemia
    - Acidosis
    - Blood flow through developing heart

### Anatomy

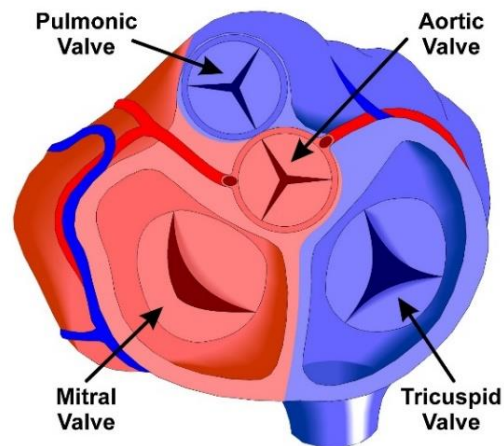
- Located between the right ventricular outflow track (RVOT) and pulmonary artery (PA)  
See illustration below for anatomic location.



Normal Heart

Illustrations reprinted from [PedHeart Resource](http://PedHeartResource.com). [www.HeartPassport.com](http://www.HeartPassport.com).  
© Scientific Software Solutions, 2016. All rights reserved

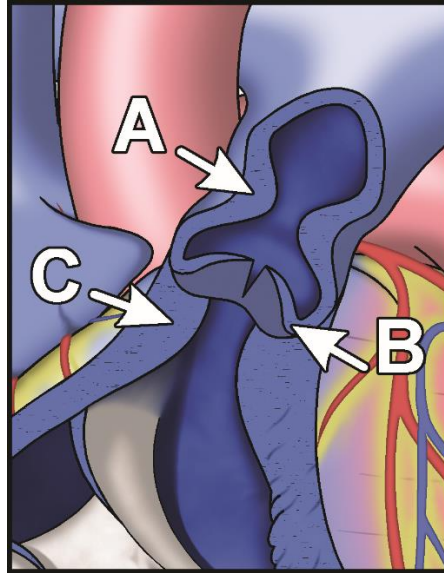
- Cross sectional view at valvar level illustrated below



Cross Section View of Heart Valves

Illustrations reprinted from [PedHeart Resource](http://PedHeartResource.com). [www.HeartPassport.com](http://www.HeartPassport.com).  
© Scientific Software Solutions, 2016. All rights reserved

- Pulmonary Valve (PV) Disorders
  - Most often congenital
    - Stenotic
    - Atretic
    - Absent leaflets
  - Acquired disorders
    - Cancer
    - Rheumatic fever affect the pulmonary valve.
- Pulmonary Stenosis (PS): Valvar, subvalvar, supra-valvar or branch stenosis (See illustration below for different levels of stenosis)



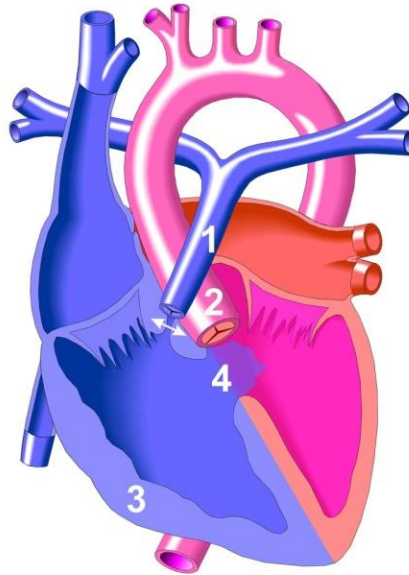
Pulmonary Stenosis

Illustrations reprinted from PedHeart Resource. [www.HeartPassport.com](http://www.HeartPassport.com).  
 © Scientific Software Solutions, 2016. All rights reserved

- Valvar PS (See 'B' in illustration above)
  - Classic valvular stenosis
    - Eight to 12% of all CHD
    - Varying degrees of severity
    - Eighty to 90% of right ventricular outflow tract obstructive (RVOT) lesions
    - Valve characteristics
      - Dome shaped pulmonary valve
      - Fused leaflets protrude from their attachment into the pulmonary artery (PA) as a conical, windsock-like structure
    - Valve orifice
      - Size varies from a pinhole to several millimeters
      - Most usually central but can be eccentric
    - Pulmonary valve ring
      - Hypoplasia
      - Characterized by:
        - Thickened, nodular, and redundant valvular leaflets with minimal or no commissural fusion
        - Lack of post-stenotic dilation of PA
- Supralvalvar pulmonary stenosis (See 'A' in illustration above)
  - Rare
  - Narrowing of PA lumen above PV

- Obstruction at main and/or branch pulmonary arteries
- Usually occurs with syndrome
  - Noonan's
  - William's
- Subvalvular pulmonary stenosis (See 'C' in illustration above)
  - Rare as isolated defect
  - Commonly associated with other lesions, mostly variants of tetralogy of Fallot
  - RVOT below PV dynamically obstructed by muscular tissue
  - Two types
    - Thickened fibromuscular thickening in wall of right ventricle (RV) infundibulum
    - Obstructive muscle band at junction of RV cavity and proximal infundibulum
- Common characteristics
  - RV hypertrophy, particularly prominent in the infundibular region
  - Dilated main pulmonary artery
    - Wide range in complexity, severity of pathology

- Associated defects
  - **Tetralogy of Fallot** (See illustration below) (Also see Defect Document on Tetralogy of Fallot)
    - Most common cyanotic CHD beyond infancy

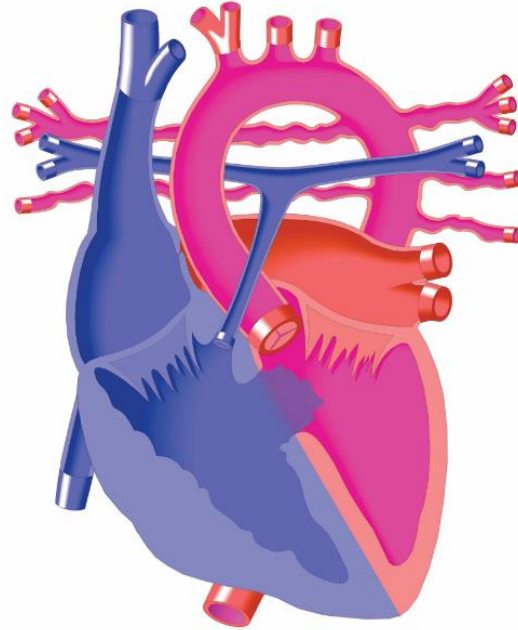


Tetralogy of Fallot

Illustrations reprinted from PedHeart Resource. [www.HeartPassport.com](http://www.HeartPassport.com).  
© Scientific Software Solutions, 2016. All rights reserved

- Four components
  - Large malaligned ventricular septal defect (VSD) (Number 4 in above illustration)
  - Stenosis of RVOT, including PV stenosis ( Arrow marks RVOT in above illustration, number 1 is small pulmonary main artery))
  - Overriding aorta (Number 2 in above illustration)
  - RV hypertrophy (Number 3 in above illustration)

- **Pulmonary Atresia with Ventricular Septal Defect** (See illustration below) [Also see Defect Document on Tetralogy of Fallot/Pulmonary Atresia (TOF/PA)]

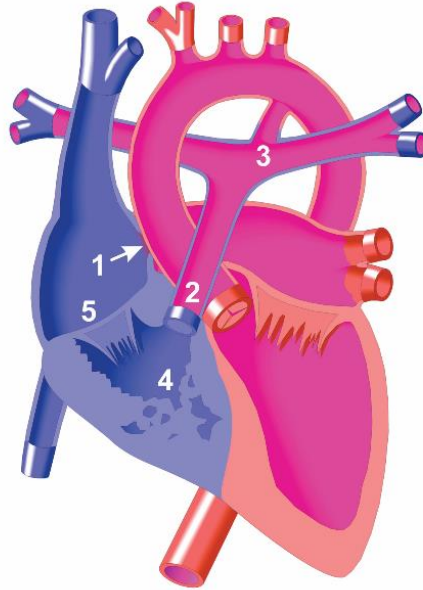


Pulmonary Atresia with Ventricular Septal Defect

Illustrations reprinted from PedHeart Resource. [www.HeartPassport.com](http://www.HeartPassport.com).  
© Scientific Software Solutions, 2016. All rights reserved

- Cyanotic congenital defect
- Components: underdevelopment of RVOT (subpulmonary infundibulum)

- **Pulmonary Atresia with Intact Ventricular Septum** (See illustration below) (Also see Defect Document on Pulmonary Atresia with Intact Ventricular Septum (PA/IVS))



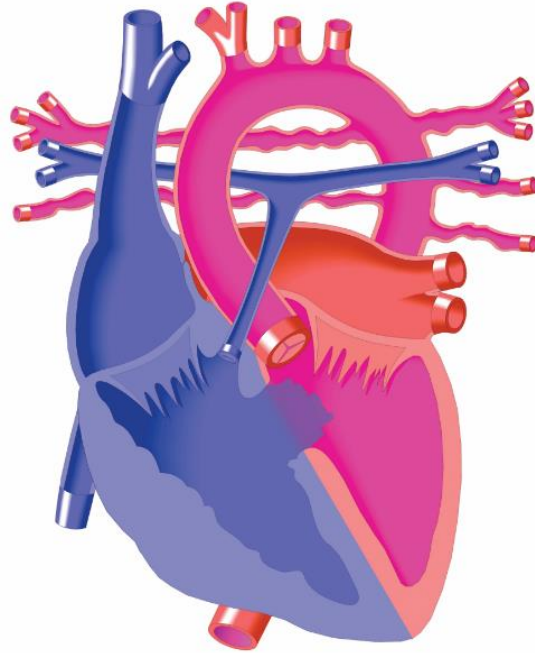
### Pulmonary Atresia with Intact Ventricular Septum

Illustrations reprinted from PedHeart Resource. [www.HeartPassport.com](http://www.HeartPassport.com).  
 © Scientific Software Solutions, 2016. All rights reserved

- Rare cyanotic congenital defect
- Components: heterogeneous right ventricular development, an imperforate pulmonary valve, and possible extensive ventricular-coronary connections (Atretic PV number 2 in illustration above)
- Ventricular septum functionally intact
- Hypertrophic RV with normal to hypoplastic cavity (Number 4 in illustration above)
- Dilated RA with PFO or ASD (Number 1 in illustration above)
- May have ventricular-coronary sinusoids, coronary stenosis, or right ventricular-dependent coronary circulation



- **Tetralogy of Fallot with Pulmonary Atresia** (Pulmonary Atresia and Ventricular Septal Defect illustrated below, TOF/PA has overriding aorta) [Also see Defect Document on Tetralogy of Fallot with Pulmonary Atresia (TOF/PA)]



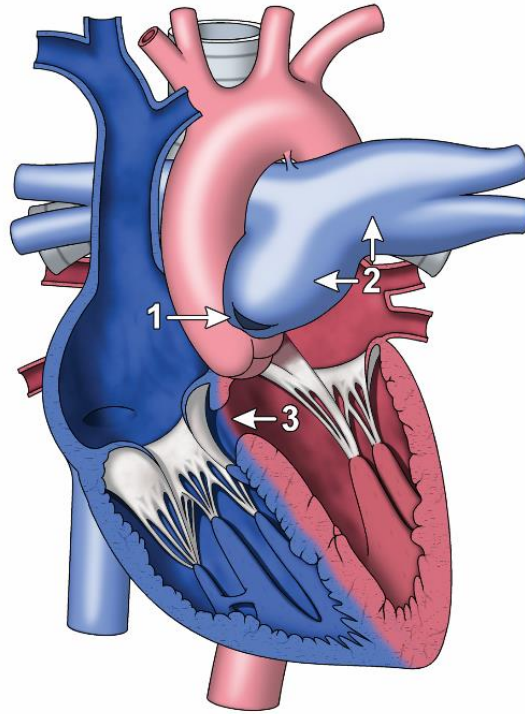
Pulmonary Atresia with Ventricular Septal Defect

Illustrations reprinted from PedHeart Resource. [www.HeartPassport.com](http://www.HeartPassport.com).

© Scientific Software Solutions, 2016. All rights reserved

- Complex cyanotic congenital defect
- Components: atresia of the PV, anterior malaligned VSD (may also be membranous or infundibular) wide range of origin, size, and distribution of pulmonary blood flow

- **Tetralogy of Fallot with Absent Pulmonary Valve** (See illustration below)  
(Also see Defect Document on Tetralogy of Fallot with Absent Pulmonary Valve)



Tetralogy of Fallot with Absent Pulmonary Valve

Illustrations reprinted from PedHeart Resource. [www.HeartPassport.com](http://www.HeartPassport.com).  
© Scientific Software Solutions, 2016. All rights reserved.

- Rare, complex cyanotic congenital defect
- Components:
  - Undeveloped PV leaflets (Number 1 in illustration above)
  - Stenotic and regurgitant, aneurysmal/massive dilation of PAs secondary to in-utero pulmonary regurgitation (Number 2 in illustration above)
  - Intracardiac features of TOF (Number 3 in illustration above)
- Should be considered a syndrome with associated findings in lungs and airways
  - Associated tracheobronchial anomalies, including significant malacia
  - Abnormal pulmonary vascular branching and wall structure

## Physiology

- Pulmonary Stenosis
  - Stenosis of PV, subvalvar, supra-valvar, and main/branch pulmonary arteries
  - Hypertrophy of RV
    - Develops due to obstruction in an effort to maintain forward flow
    - Degree of obstruction proportional to the increase in RV pressure and pressure gradient across the valve
    - RV hypertrophy decreases RV compliance and increases RA pressure causing waves in right atrium
    - Increasing RA pressures a right-to-left shunt can occur [patent foramen ovalae (PFO) / or ASD]
    - Right sided heart failure occurs in severe obstruction
- Associated defects
  - (See Defect Documents for Tetralogy of Fallot, Tetralogy of Fallot with Pulmonary Atresia, Tetralogy of Fallot with Absent Pulmonary Valve, Pulmonary Atresia with Intact Ventricular Septum, Pulmonary Atresia with Ventricular Septal Defect)

## Procedures/Interventions

- Pulmonary valve insufficiency
  - Monitor RV dilation and patient symptoms
  - Monitor for arrhythmia
  - Monitor for compromise cardiac output, compression left ventricle
  - Surgical intervention: PV replacement with conduit or bioprosthetic valve
  - Catheter intervention: PV replacement with Melody valve
- Pulmonary Valve Stenosis
  - Symptomatic patients
    - RV pressure > 50% systemic
    - RV dysfunction
  - Interventions:
    - Balloon pulmonary valvuloplasty
    - Surgical valvuloplasty via median sternotomy
    - Transannular patch for subvalvar and supra-valvar stenosis
- Pulmonary Valve Atresia (See Defect Documents for Pulmonary Atresia, PA/IVS, PA/VSD)
  - Intervention dependent upon PA anatomy
  - Ductal-dependent pulmonary blood flow and confluent PA's
    - Repair with RV-PA conduit and VSD closure
    - Staged with Blalock-Taussig Shunt (BTS) or patent ductus arteriosus (PDA) stent

- Variations of tetralogy of Fallot (See Defect Documents for Tetralogy of Fallot, Tetralogy of Fallot with Pulmonary Atresia, Tetralogy of Fallot with Absent Pulmonary Valve)

### **Specific considerations and routine care**

- Preprocedure considerations
  - Neonates with critical PS
    - Ductal dependent pulmonary blood flow
    - Present with systemic to suprasystemic RV pressure, right to left atrial shunting, and hypoxia
    - As the PDA closes
      - Develop profound hypoxemia
      - Results in acidosis
        - Inadequate pulmonary blood flow
        - Decreased cardiac output
  - Progression of moderate PS
    - RV dilation
    - Decreased RV function
    - Tricuspid regurgitation
  - Isolated mild pulmonary stenosis
    - May present with murmur
    - May develop dyspnea with exertion over time
    - Further progression of obstruction
      - Increased symptoms
      - Manifestations of right heart failure: tachycardia, peripheral edema, hepatomegaly, dyspnea, syncope, exercise intolerance, arrhythmias and even sudden death
  - Management of associated defects (See Defect Documents for Tetralogy of Fallot, Tetralogy of Fallot with Pulmonary Atresia, Tetralogy of Fallot with Absent Pulmonary Valve, Pulmonary Atresia with Intact Ventricular Septum, Pulmonary Atresia with Ventricular Septal Defect)
- Preprocedure Management
  - Hypercyanotic spells
    - Knee chest positioning, oxygen, sedation, beta blockers and volume expansion
    - Extreme spell management: General anesthesiology, alpha agonist and emergent surgery
  - Neonates with Critical PS (PS/RVOTO and PA/VSD)
    - Initially stabilize the patient with PGE infusion for critical PS
      - Provide adequate pulmonary blood flow
      - Provide systemic oxygenation
    - With profound cardiogenic shock

- Mechanical ventilation
    - Inotropic/vasoactive support of systemic ventricle and cardiac output
    - Assessment of end organ complications
  - Neonates with excessive pulmonary blood flow
    - May require ventilation maneuvers to control pulmonary-systemic flow ratios
- Procedures
  - Catheter interventions
    - Balloon valvuloplasty
      - Neonatal period
        - Critical PS
        - Pulmonary atresia with membranous pulmonary valve
      - Post neonatal period
        - Increasing symptoms
        - Repeat procedure
    - RF ablation of membranous pulmonary valve/balloon valvuloplasty
  - Surgical interventions
    - Valvotomy
    - Valvotomy with placement of systemic-pulmonary shunt ( Modified Blalock-Taussig, Central Shunt)
    - Unifocalization of pulmonary arteries, unilateral or bilateral with systemic to pulmonary artery shunt
    - Unifocalization of pulmonary arteries with placement of RV-PA conduit
    - Isolated placement of RA-PA conduit
    - Complete anatomic repair with closure of VSD and establishment of continuity between RV and PA with valved conduit
    - Consideration of maintenance of ASD or PFO for acute decompression of non-compliant RV
- Post procedure Management
  - Bleeding
    - Important assessment at site of catheter insertion
    - Common postoperative complications specifically with unifocalization procedures
  - Hypoxemia
    - May result from inadequate relief PS
    - Branch PA stenosis from surgical intervention may require re-intervention in catheterization lab or operating room
  - Injury to TV apparatus or residual VSD may require re-intervention due to intractable low cardiac output and hypoxemia in the postoperative period
  - Small right ventricle (Neonatal period)

- May need to maintain prostaglandin infusion for adequate pulmonary blood flow
- Monitor volume status/central venous pressure
- Patency of ASD/PFO
- Right Ventricular noncompliance
  - RV dysfunction with decreased compliance
  - Monitor volume status closely
  - Need to maintain adequate preload for non-compliant RV, avoid hypovolemia
  - Avoid tachycardia to improve preload to RV, time to empty
  - May require inotropic/vasoactive support for short period of time following procedure (Both balloon and surgical intervention)
  - Persistent cyanosis due to RV noncompliance with right to left atrial shunting if ASD not completely closed
  - “Suicide RV”
    - Occurs if RV pressures are systemic or suprasystemic prior to balloon valvuloplasty
    - Causes RVOT to collapse on itself
    - Milrinone drip and preload may improve RV pressure and function
    - Prevent dehydration – careful use of diuretics
- Arrhythmias
  - Associated with VSD closure, RV dysfunction, and/or RV muscle resection
  - Potential arrhythmias include: junctional ectopic tachycardia, atrial tachycardia, ventricular tachycardia. Heart block uncommon but possible due to VSD closure
  - Right ventricular dependent coronary circulation (RVDCC) requires high RV pressure for adequate coronary circulation. Ischemic changes on EKG suggest coronary perfusion deficit
- Residual valvar stenosis or regurgitation
  - Significant residual stenosis
    - Prolongs RV dysfunction
    - Worsens TR
    - Increases right to left atrial shunt
    - Increases hypoxemia
    - May result in ascites, LCOS, and effusions
  - Pulmonary regurgitation
    - Well tolerated in early post procedure period
    - Progresses over time
      - RV dilation
      - Decreased function
      - TR

- Arrhythmia
- Exercise intolerance, fatigue
- Necessitates re-intervention

### **Long term problems/complications**

- Residual and progressive pulmonary regurgitation in tetralogy of Fallot or critical pulmonary stenosis repairs
  - May progress to TR, RV dilation, symptoms of exercise intolerance, fatigue and arrhythmia
  - May necessitate pulmonary valve replacement
- Pulmonary artery collaterals
  - Become stenotic over time
  - Limiting pulmonary blood flow
  - Result in hypoxemia
  - May require unifocalization of one or both PA's at one or more operations
- Branch pulmonary artery stenosis
  - May require multiple re-interventions
    - Surgical patching
    - Catheter dilation
    - Surgical/catheter stenting
  - Full assessment of pulmonary artery anatomy will require catheterization or MRI/MRA
- Placement of RV to PA conduits for repairs at any age
  - Will require eventual replacement
    - Catheterization intervention (Melody Valve)
    - Surgical replacement
- Long term development of arrhythmias (See Adult Guidelines on Arrhythmias in ACHD)
  - Ventricular arrhythmias
    - Due to ventriculotomy for conduit placement
    - Intracardiac surgical scars
    - May require medications or ablation, ICD pacemaker
  - Atrial arrhythmias
    - Atrial fibrillation/flutter due to atrial dilatation
    - May require pacemaker placement, medication or ablation (operative or catheterization)
- Aneurysmal formation of the RVOT from placement of outflow tract patch or conduit
- Associated syndromes such as DiGeorge or William's syndrome
  - Requires follow up for syndrome specific complications

### **References:**

Armstrong, E.J., & Bischoff, J. (2004). Heart valve development. Endothelial cell signaling and differentiation. *Circulation Research*. Published online at <http://www.circresaha.org>. doi: 10.1161/01.RES.0001411.95728.da Accessed 8/2015.

Biechler, S.V., et al. (2014). The impact of flow-induced forces on the morphogenesis of the outflow tract, *Frontiers in Physiology*, 5(225). Published online at <http://www.frontiersin.org> doi:10.3389/fphys.2014.00225. Accessed 9/2015

Cheung, Y.F., Leung, M. P., Lee, J. W., Chau, A.K., Yung, T. C., Evolving management of critical pulmonary stenosis in neonates and young infants. *CardiolYoung*. 2000 May; 10(3):186-92.

Davis, S. (2006). Tetralogy of Fallot with and without Pulmonary Atresia. In *Critical Heart Disease in Infants and Children* (2<sup>nd</sup> ed). Philadelphia, PA: Mosby

Domina, Y., Munoz, R., Kreutzer, J., & Morell, V. (2010). Pulmonary Stenosis. In *Critical Care of Children with Heart Disease* (2<sup>nd</sup> ed). Philadelphia, PA: Mosby.

Everett, A. D., & Lim, D. S. (2010). *Illustrated Field Guide to Congenital Heart Disease and Repair*. (3<sup>rd</sup> ed.) Charlottesville, VA: Scientific Software Solutions, Inc.

Gupta, D., Fricker, J., & Udassi, J. (2015). Pulmonary Valve Stenosis. In *Pediatric Cardiac Intensive Care Handbook*. Washington, D.C.: Pediatric Cardiac Intensive Care Books.

Martin, L.D., Nyhan, d., Wetzel, R.C. (2006) Regulation of Pulmonary Vascular Resistance and Blood Flow. In *Critical Heart Disease in Infants and Children* (2<sup>nd</sup> ed). Philadelphia, PA: Mosby

Moore, K.L. & Persaud, T.V.N. (2008). The cardiovascular system: in *The Developing Human. Clinically Oriented Embryology* (8<sup>th</sup> ed). Philadelphia, PA: Saunders, an imprint of Elsevier Inc.

O'Laughlin, M.P., Ringel, R.E. (2006) Diagnostic and Therapeutic Cardiac Catheterization. In *Critical Heart Disease in Infants and Children* (2<sup>nd</sup> ed) Philadelphia, PA: Mosby.

O'Laughlin, M., & Ringel, R. (2006) Diagnostic and Therapeutic Cardiac Catheterization. In *Critical Heart Disease in Infants and Children*. (2<sup>nd</sup> ed). Philadelphia, PA: Mosby.

Park, M. K. (2014) *Park's Pediatric Cardiology for Practitioners* (6<sup>th</sup> ed.). Philadelphia, PA: Elsevier.

Plunkett, M. D., Laks, H. (2007) Pulmonary Stenosis and Pulmonary Atresia with Intact Septum. In *Mastery of Cardiothoracic Surgery*. (2<sup>nd</sup> ed). Philadelphia, PA: Lippincott Williams and Wilkins.

Rudd, N. (2013) Pulmonary Stenosis: Acyanotic defects: Obstructive lesions. Chapter 8 Cardiovascular Disorders. In M. F. Hazinski (Ed.). *Nursing Care of the Critically Ill Child* (3<sup>rd</sup> ed.). Philadelphia, PA: Elsevier

Thompson, W.R., Nichols, D.G., Spevak, P.J. and Ungerleider, R.M.(2006) Double-outlet Right Ventricle and Double-outlet Left Ventricle. In *Critical Heart Disease in Infants and Children* (2<sup>nd</sup> ed). Philadelphia, PA: Mosby



Vivda, V.K., Rito, M.L., Zucchetto, F., Biffanti, R., Padalino, M.A., Milanesi, O., Stellin, G.  
Pulmonary Artery Branch Stenosis in Patients with Congenital Heart Disease. DOI:  
10.1111/jocs.12121 (J Card Surg 2013; 28: 439-445.

Illustrations reprinted from PedHeart Resource. [www.HeartPassport.com](http://www.HeartPassport.com). © Scientific Software Solutions, 2016. All rights reserved.

12/2015