

## Aortic Valve

What the Nurse Caring for a Patient with Congenital Heart Disease Needs to Know

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### Embryology

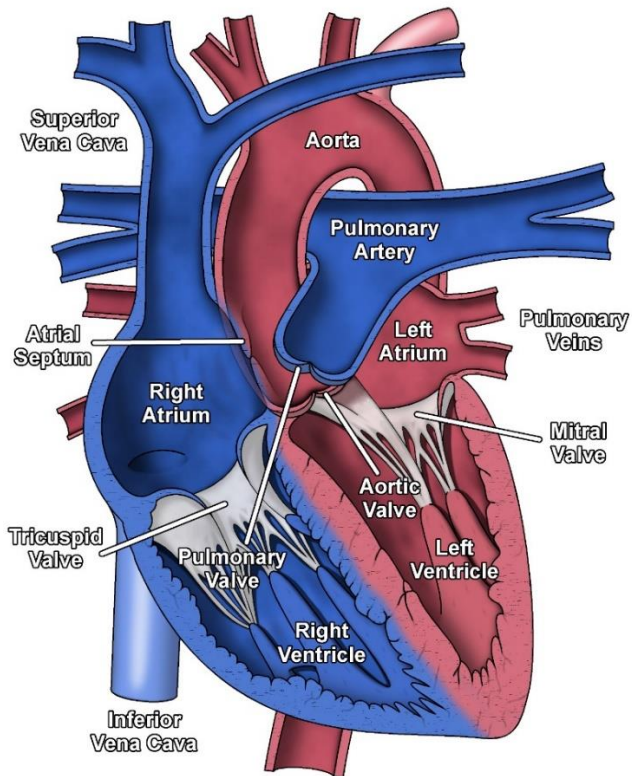
- Occurrence:
  - Defects of cardiac valves are the most common subtype of cardiac malformations
  - Account for 25% to 30% of all congenital heart defects
  - Most costly and relevant CHD
  - Wide spectrum of congenital defects in aortic valve
- Development of the heart valves occurs during the fourth 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**

- Clinical spectrum varies from presence of a malformed bicuspid aortic valve that functions normally to severe aortic stenosis (AS)
- Types/anatomic location of stenosis (See illustration below for intracardiac position of aortic valve and relation of other structures involved in anatomic locations of stenosis.)
  - Valvular
    - Seventy to 80% of all AS
    - Decrease in orifice size
      - Results from thickening and increased rigidity of valve leaflets.
    - Most common defect
      - Bicuspid aortic valve
      - Only two valve cusps present
      - Results from partial or complete fusion of two of the aortic valve cusps
      - Conjoined vs. nonconjoined cusps may be equal or asymmetric
      - Valve orifice may be central or non-central.
    - Other forms
      - Unicuspid valve
        - Fusion of more than one cusp
        - Results in a single slit like opening that extends to the annulus
      - Partial fusion of all three cusps with small central orifice
      - Hypoplasia of annulus
        - Rare
        - Aortic valve cusps relatively normal
  - Subvalvular
    - Ten to 20% of all AS
    - Common associated defects:
      - Ventricular septal defect
      - Coarctation of the aorta
      - Atrioventricular septal defect
      - Valvular aortic stenosis
      - Mitral valve anomalies.
    - Obstruction
      - Ridge of membranous and/or fibrous tissue
        - Encircles left ventricular outflow tract (LVOT)
        - Or diffuse and form a tunnel

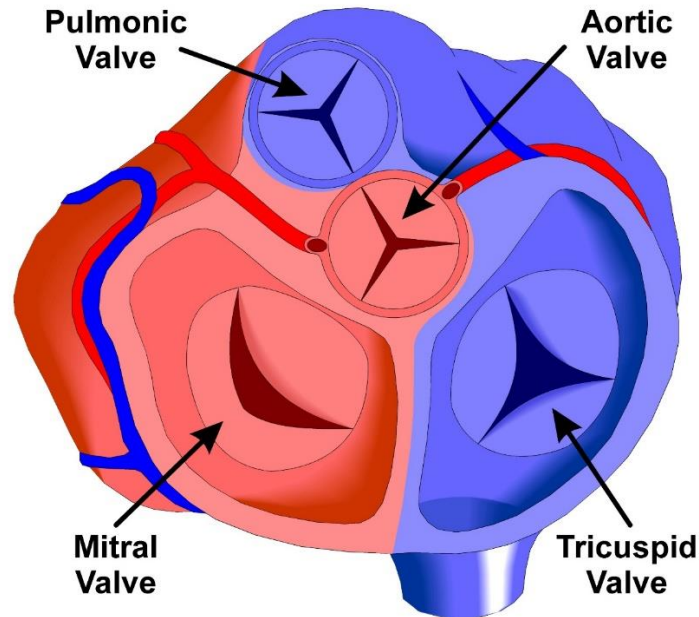
- Tissue may be tethered to:
  - Aortic valve
  - Or anterior mitral leaflet
- Aortic valve itself may become thickened due to subvalvular turbulent flow jet damaging the aortic valve cusps
- Supravalvular
  - Least common type of AS
  - Approximately 30% to 50% have Williams syndrome
  - Cause: reduced elastin in the arterial media causes decreased elasticity and thickened media with smooth muscle hypertrophy and increased collagen deposition.
    - Most commonly occurs at the sinotubular junction
    - Changes may also occur throughout the entire arterial system
      - Ascending aorta or aortic arch branches
      - Main or proximal branch pulmonary arteries
      - Renal and mesenteric arteries
  - Abnormal attachment of AV commissures
    - Peripherally at level of the sinotubular junction
    - May cause
      - Abnormalities of the aortic valve
      - Impaired coronary blood flow



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- Cross sectional illustration shows aortic valve cusps in relation to other heart valves and coronary arteries.



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- Other pathologic features that impact aortic valve dysmorphology and dysfunction include:
  - Calcification (rare in childhood and adolescence, but common in adults)
  - Fibrosis
  - Lipid accumulation
  - Inflammatory changes
  - Myxomatous degeneration
  - Annular dilation
  - Acquired fibrotic fusion of true commissures
- Congenitally abnormal aortic valves may result in weakening of ascending aorta
  - May result in:
    - Annular dilation, as well as
    - Dilation or aneurysm of ascending aorta
    - At risk for aortic dissection or rupture
- May develop left ventricular (LV) hypertrophy and myocardial fibrosis.

### Physiology

- In utero presence of moderate to severe aortic stenosis
  - Increases LV pressure

- May lead to:
  - LV hypertrophy
  - Decreased LV compliance
  - Decreased flow through the left heart
  - May result in hypoplasia of the LV, mitral valve, aortic valve annulus, and LV outflow tract.
- Valvar AS
  - Causes obstruction of LV outflow,
  - Increases LV afterload
  - LV pressure > aortic pressure during ejection due to decreased effective area of the valve orifice
  - With normal stroke volume, the pressure gradient reflects severity of the stenosis
  - Neonatal critical aortic stenosis:
    - Limited antegrade flow across the LV outflow tract
      - Requires a patent ductus arteriosus (PDA) to provide adequate systemic perfusion
    - Results from closure of the PDA
      - Cardiogenic shock
      - Severe hypoperfusion
      - Profound acidosis
  - Pediatric AS (Patients present after one year of age)
    - See compensatory LV hypertrophy
    - Maintains normal LV wall stress despite elevation in peak systolic pressure
    - Maintains cardiac output
    - Increases LV end-diastolic volume and pressure
- Valvular/subvalvular AS
  - LV subendocardial ischemia and infarction from:
    - Imbalance in coronary blood flow to the hypertrophied left ventricle
    - Increased myocardial oxygen demand
    - Ventricular pressure overload
  - Severe aortic stenosis
    - Little coronary reserve during stress
    - Exercise
      - Minimally increases stroke volume
      - Results in:
        - Increased heart rate
        - Shortened systole and diastole
        - Decreased time for ejection
        - Increased LV systolic pressure
        - Increased oxygen demand
        - Decreased coronary perfusion from shorter diastole
      - Increases systemic vasodilation
        - Further decreases diastolic blood pressure
        - May impair coronary perfusion
  - Supravalvular aortic stenosis

- Physiology similar to valvar and subvalvular
- Effect on coronary arteries
  - Proximal to the obstruction
  - Exposed to high pressure during systole
    - Leads to changes in coronary vasculature
    - Limited diastolic flow
      - Results in inadequate oxygen supply to meet demand
      - Leads to ischemia, infarction, and sudden death

### Procedures/Interventions

- Medical Treatment
  - Bacterial endocarditis prophylaxis
  - Exercise restrictions
  - Periodic follow-up evaluations to monitor progression of valve dysfunction
- Indications
  - Adults - intervention is recommended only when symptoms develop
  - Children and adolescents
    - Earlier intervention even in asymptomatic patients
    - Relief of obstruction
      - Reduces the risk of sudden cardiac death
      - Decreases the extent of subtle and progressive myocardial injury
- Catheter Intervention
  - Balloon Valvuloplasty
    - Children and adolescents
      - Initial treatment
      - Re-stenosis or regurgitation may occur
      - Valve replacement may become necessary for definitive treatment
      - Not indicated if aortic valve regurgitation
    - Adults
      - Not indicated if cusp calcification develops
      - Not indicated with significant aortic valve regurgitation
  - American College of Cardiology/American Heart Association guidelines:

| <b>Patient population</b>  | <b>Intervention</b>                                      |
|--|--|
| Asymptomatic children and young adults with peak Doppler gradients $\geq 70$ mmHg or peak-to-peak gradient $> 60$ mmHg.                  | Consider cardiac cath and possible balloon valvuloplasty |
| Patients who play competitive sports or may become pregnant, with peak Doppler gradients 50-70 mmHg or peak-to-peak gradient $> 50$ mmHg | Consider cardiac cath and possible balloon valvuloplasty |

|  |  |
|--|--|
| Patients with symptoms (angina, syncope, dyspnea on exertion) or ischemic changes at rest or on exercise ECG | Valvuloplasty if peak-to-peak gradient is > 50 mmHg, other symptoms should be sought if gradient does not meet criteria                |
| Asymptomatic patients with peak-to-peak gradients < 50 mmHg  | Valvuloplasty not recommended unless cardiac output is impaired (gradient underestimates true severity of obstruction in this setting) |

- Transcatheter aortic valve replacement (TAVR)
  - Minimally invasive catheter placement of a bioprosthetic, expandible aortic valve within a native, calcified, severely stenotic aortic valve
  - Inserted in a hybrid (surgical suite with bi-plane imaging) lab
  - Inserted by a multidisciplinary team
    - Interventional cardiologist
    - Cardiothoracic surgeon
  - Inserted by an atrial or transthoracic approach
    - Involves sheaths and catheters > 16Fr
    - Too large for pediatric arteries
  - Consider in severe symptomatic aortic valve stenosis
  - Available for patients with severe symptoms at increased surgical risk
    - Advanced age
    - Multiple comorbidities
    - Surgical options determined to be contraindicated or at extreme risk by two cardiovascular surgeons
    - Currently (2015) not available for congenital unicuspid or bicuspid aortic valves
  
- Surgical Treatment
  - Indications
    - Development of progressive aortic valve regurgitation
    - Recurrent stenosis refractory to balloon valvuloplasty
  - Mechanical prosthesis
    - Requires long-term anticoagulation
    - No potential for valve growth
    - Limited availability of small sizes
  - Bioprosthetic valves (homograft or heterograft)
    - Avoids the need for anticoagulation
    - No potential for valve growth
    - Longevity less than mechanical, especially in small children
  - Ross procedure
    - May be preferred in infants and small children
    - Translocation of semi-lunar valves (See cross sectional illustration for proximity of semi lunar valves and similar structure.)

- Native pulmonary valve translocated to the aortic position
- Pulmonary homograft implanted in the pulmonary valve position
- Native aortic valve may be surgically revised and inserted into pulmonary position (Double Switch)
- Benefits:
  - No need for anticoagulation
  - Potential autograft (neoaortic valve) growth
- Disadvantage:
  - May develop pulmonary homograft dysfunction
  - May require additional procedures
    - Pulmonary homograft replacement
    - Neoaortic valve dilation
    - Stenosis of both pulmonary and aortic suture lines
- Surgical resection of subaortic stenosis
  - Indications:
    - Progression of subaortic obstruction
    - Development of aortic regurgitation
  - Procedure
    - LV muscle resection
    - Membrane excision
    - Potential surgical correction of mitral valve abnormalities that contribute to the subaortic obstruction, such as anomalous papillary muscle insertion
  - Complications:
    - Recurrent subaortic stenosis in 20% of patients
    - Heart block
    - Worsening of aortic or mitral valve regurgitation
    - Inadvertent creation of a ventricular septal defect
- Severe subaortic stenosis with a small aortic annulus
  - May require more extensive tissue resection
  - Ross/Konno procedure
- Konno procedure
  - Indications
    - Treat all levels of LVOTO
    - Tunnel like subaortic stenosis
    - Diffuse obstructive hypertrophic cardiomyopathy
    - Congenital aortic valve stenosis
    - Proximal ascending aorta stenosis
  - May be combined with Ross procedure (Ross/Konno) specifically for tunnel subaortic stenosis and aortic annular hypoplasia
  - Procedure: Konno operation with Aortic valve replacement
    - Longitudinal incision on anterior aspect of ascending aorta distal to the aortic valve



- Continue incision to left of right coronary artery and into the right ventricle and down into the interventricular septum below any subvalvar stenosis
- Patch sutured to left ventricular side of the VSD and continued across the aortic annulus and onto the aorta enlarging the aortic annulus
- Prosthetic valve placed in the enlarged aortic root
- RVOT reconstructed with patch
- Procedure: Modified Konno
  - Enlargement of the LVOT without replacement of the aortic valve (normal size aortic annulus but presence of subaortic stenosis)
  - Right ventricular incision into the ventricular septum and into the LVOT up to the aortic valve
  - Patch placed on the right ventricular side of the surgically created VSD
- Complications
  - Complete heart block and requirement for pacemaker
  - Incomplete relief of LVOTO
  - Residual VSD
  - Damage to the mitral valve apparatus
- Surgical repair of supra-ventricular aortic stenosis
  - Patch enlargement of the sinotubular junction above the noncoronary sinus
  - Extended aortoplasty with a patch into the noncoronary and right coronary sinuses
  - Insertion of separate patches in all three sinuses after transecting the aorta (Brom's technique)
  - Surgical correction of any obstruction to coronary blood flow
  - Patch augmentation of the ascending or transverse aorta as necessary

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

- AS pre-procedure management depends on the degree of obstruction to forward flow from the left ventricle and the presence of systemic hypoperfusion
- Neonates with critical AS
  - Important to determine if the left heart and aortic structures are compatible with a two ventricle repair
    - Based on echocardiographic information
    - Considerations
      - Ventricular size, end diastolic volume
      - Left sided lesions
        - Mitral stenosis, adequacy of mitral valve annulus
        - Coarctation of the aorta
- Neonatal Critical AS pre-procedure Management
  - Prostaglandin infusion

- Establishes ductal-dependent systemic flow
    - Alleviates pulmonary hypertension seen with severe LV dysfunction
  - Vasoactive support
    - Resuscitate
    - Support LV
    - Increase contractility
  - Intubation and mechanical ventilation
    - Correct severe acidosis
    - Reduce metabolic demand
    - Control pulmonary hypertension
    - Afterload reduce left ventricle
  - May require emergent atrial septostomy or even management with ECMO.
  - Monitor for signs of end-organ compromise
- Procedures: (See Peds/Neo Guidelines for Post-operative Care)
  - Key post-procedure management points
  - All procedures
    - Continual monitoring of cardiac rhythm for arrhythmias and ST segment changes
    - Systolic and diastolic blood pressure, widening pulse pressure
    - Low cardiac output syndrome
  - Balloon valvuloplasty
    - Impacted by the degree of severity of critical AS.
    - Low cardiac output syndrome
      - Best managed by inotropes to improve cardiac function
      - Ventilation with high concentrations of inspired oxygen, normal pH to treat pulmonary vascular reactivity
    - Significant pulmonary hypertension
      - Manage ventilation, consider use of nitric oxide
      - Continue prostaglandin infusion to maintain a patent ductus
        - Maintain systemic perfusion
        - Decompress the pulmonary artery hypertension
    - Monitor for aortic insufficiency
    - Monitor for bleeding
      - Catheter insertion site
      - Retroperitoneal
  - Surgical valvotomy
    - Monitor for hypertension
      - LV function usually preserved
      - Often hyperdynamic due to long standing stenosis and high afterload results in hypertension
    - Low cardiac output syndrome (LCOS)
      - May be prolonged due to the hypertrophied LV
      - Positive pressure ventilation may reduce LV afterload and improve cardiac output
    - Monitor for arrhythmias and changes in coronary artery perfusion
      - Left bundle branch block or complete heart block

- ST segment changes
  - Ventricular arrhythmia
- Surgical aortic valve replacement
  - Valve selection depends on:
    - Patient age
    - Size of aortic valve annulus
  - Anticoagulation necessary if a prosthetic mechanical valve used (See both Ped/Neo and Adult Anticoagulation Guidelines.)
- Ross Procedure – (Pulmonary Autograft)
  - Increased bleeding risk
    - Re-exploration may occur in a small amount of patients
    - Dehiscence of aortic root anastomosis
  - Increased risk of arrhythmias
    - Complete heart block
      - May require a permanent pacemaker
    - Compromised coronary artery perfusion
- Surgical Repair of Subvalvular Stenosis
  - Increased sub aortic resection
    - Increased risk for third degree/complete heart block
    - Increased risk for significant ventricular dysfunction
  - Inadvertent creation of a ventricular septal defect
  - Surgical disruption of mitral valve apparatus and resultant mitral regurgitation
- Surgical Repair of Supravalvular Stenosis
  - Risk of pulmonary hypertension
  - Associated CHD in genetic syndromes such as William’s syndrome
    - Main pulmonary artery stenosis
    - Branch pulmonary artery stenosis
    - Risk of suicide RV if relief supravalvar AS results in suprasystemic RV pressure due to unrelieved PA stenosis

### **Long-term problems/complications and routine care**

- Endocarditis prophylaxis is necessary
- Patients with bicuspid aortic valve are at risk for progressive aortic root dilation and aortic dissection, even in the absence of stenosis or significant regurgitation
  - Most patients with aortic dissection have hypertension, so medical management of hypertension is crucial
  - Careful surveillance with serial echocardiograms is warranted to detect aortic root dilation
- Long-term monitoring
  - Aortic valve function and gradient
  - Development of LV hypertrophy or dysfunction
- Anticoagulation (See both Ped/Neo and Adult Guidelines for Anticoagulation)
  - Prosthetic mechanical valve replacement

- Bioprosthetic valve replacement may require anticoagulation with added risk factors
  - Atrial fibrillation
  - LV dysfunction
  - Hypercoagulable state
- Recommendations during pregnancy
  - See Adult Guidelines on Pregnancy in Adult Congenital Heart Disease for more specific considerations and anticoagulation management
- Following intervention, either surgical or by catheterization, patients must be monitored for re-stenosis or development of valve regurgitation

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