

Mitral Valve

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

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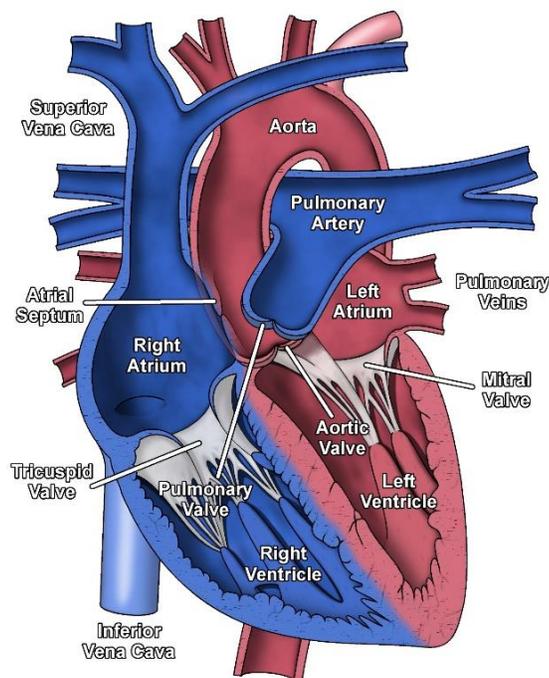
Embryology

- 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
- 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

- Positioned between left atrium (LA) and left ventricle (LV) (See illustration below for normal anatomy of heart valves and chambers)



Normal Heart

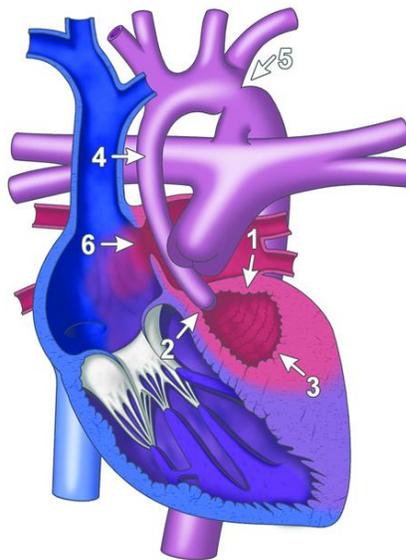
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- Elements of mitral valve (MV) (See above illustration of normal heart for MV components)
 - The LA wall
 - LA myocardium extends over the proximal portion of the posterior leaflet
 - Anterior leaflet attached to root of aorta

- LA enlargement may result in mitral regurgitation (MR) by stretching the posterior leaflet
 - Annulus
 - Fibrous ring that connects with the leaflets
 - Not continuous circular ring around mitral orifice
 - D-shaped, saddle-shaped
 - Functions as a sphincter
 - Contracts
 - Reduces surface area during systole to ensure closure of leaflets
 - Reduces orifice by 20-30%
 - Annular dilation
 - Causes incompetent leaflet closure
 - Results in MR
 - Anterior and posterior leaflets, chordae, papillary muscles
 - Consists of two leaflets
 - Thin, pliable, translucent, soft
 - Anterior leaflet
 - Longer
 - Positioned posterior to aortic root
 - Posterior leaflet
 - Shorter
 - Encompasses a wider portion of annulus
 - Chordae tendineae
 - Small, fibrous strings
 - Originate either from apical portion of papillary muscles or directly into ventricular wall
 - Insert directly into anterior or posterior valve leaflets
 - Papillary muscles
 - Located in the LV
 - Attach to the valve leaflets via the chordae tendineae
 - Contract and shorten during systole
 - Prevent inversion prolapse of valve
 - Cause closure of MV
 - Extreme fusion of papillary muscle results in mitral stenosis (MS)
 - Rupture of a papillary muscle results in acute mitral regurgitation (MR)
 - Congenital MV disease
 - Rarely an isolated abnormality
 - Associated with more complex diseases
 - Hypoplastic left heart syndrome
 - Shone's complex
 - Atrioventricular septal defect
- Mitral Valve Obstructive (Stenotic) Lesions
 - Supravalvular Mitral Ring

- Caused by the presence of extra left atrial connective tissue
 - Obscures the MV
 - Obstructs transvalvular flow.
- Constrains the leaflet motion
 - Provides only a small orifice for cardiac output
 - Results in left-sided heart obstruction
 - May result in PAH
- Usually associated with other defects
 - Parachute MV
 - Subaortic stenosis
 - Aortic coarctation
- MV Atresia (See illustration below for most common MV Atretic lesions)



Hypoplastic Left Heart Syndrome

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- All components of the MV apparatus are small (Number 1 in above illustration shows an Atretic MV)
- Majority (80%) associated with:
 - Severe left ventricular hypoplasia (Number 3 in above illustration)
 - Left ventricular outflow tract obstruction (Number 2 in above illustration)
- Additional associated defects
 - Complex single ventricle anatomy
 - Double-outlet right ventricle

- Single left ventricle with transposition of the great vessels
 - Large VSD or straddling tricuspid valve in association with MV atresia may have a normal LV size
- Congenital Mitral Stenosis
 - Most common form of congenital MV obstruction
 - All valve components can be involved
 - Symmetric congenital mitral stenosis
 - Normal distribution of chordae from each leaflet
 - Leaflets and annular components hypoplastic
 - This narrowed orifice creates obstruction
 - Asymmetric mitral stenosis
 - Cords attach predominantly or exclusively to one of the papillary muscles.
 - Anterior papillary muscle absent
 - Leaflet attached to a posterior papillary muscle --“true parachute” mitral valve
 - Associated with:
 - Aortic coarctation or aortic valve stenosis
 - Ventricle usually normal-sized
- Fusion of Mitral Valve Commissures
 - Poorly formed valve commissures
 - Poorly developed chordae
- Excessive Mitral Valve Chordal Tissue
 - Redundant chordal tissue or extra leaflet tissue
 - Obstructs flow at the subvalvular level and left ventricular outflow tract
- Double-Orifice Mitral Valve
 - A bridge of extraneous tissue or fusion of leaflets divides MV inlet into two separate orifices
 - Approximately 50% of the time associated with valvar dysfunction
 - Includes both valvar stenosis and insufficiency
 - Repair commonly leads to valve incompetence
- Mitral Arcade
 - Severely dysmorphic subvalvular and leaflet components
 - Leaflets thickened with dysplastic chordae
 - Leaflets may directly attach to papillary muscles
 - Leads to valvular tethering and excursion
 - Results in obstruction to flow into LV
- Mitral Valve Regurgitant Lesions
 - Mitral valve prolapse
 - Most common form of valve abnormality, 2-6% of CHD
 - Enlargement of mitral valve leaflets such that during systole the large leaflets project into left atrium.
 - Usually does not impair valve function
 - Can lead to mitral insufficiency
 - Cleft Mitral Valve Leaflet
 - Rarely occurs as an isolated defect

- Usually located in the anterior leaflet
- Posterior leaflet involvement rare
- Subvalvular apparatus (chordae and papillary muscle) can cross the left ventricular outflow tract and cause varying degrees of left ventricular outflow tract obstruction
- Other lesions
 - Lesions where the chordae are thickened and shortened: EFE changes, infarction

Physiology

- The mitral valve opens during diastole to allow blood to flow from the LA to the LV and closes during systole to prevent backflow of blood into the LA from the LV
- Closure of the mitral valve results from the complex interaction of several factors:
 - Rise of pressure in the left ventricle during contraction
 - Contraction of the papillary muscle which support the valve
 - Passive opening of the mitral valve during early diastole
 - Recoil in the early phase of filling contributing to ventricular filling
 - Opening of the valve during atrial contraction
- Mitral Valve Obstruction
 - Mitral stenosis
 - Most common cause – rheumatic heart disease
 - Characterized by a narrowing of the left ventricular inflow tract at the level of the mitral valve
 - Impairs left ventricular filling during diastole
 - Causes rise in left atrial pressure
 - Results in pulmonary hypertension
 - CHF results from increased left atrial pressure and decreased left ventricular preload
 - Effect on pulmonary pressure
 - Increases left atrial, pulmonary venous, and pulmonary capillary wedge pressures
 - Results in:
 - Increased interstitial and alveolar fluid and congestion
 - Increased airway resistance occurs from passive pulmonary edema
 - Affects pulmonary mechanics and gas exchange
 - Symptoms include hypoxemia, hypercapnia, increased work of breathing
 - Permanent damage to pulmonary parenchyma and vascular architecture
 - Increased pulmonary vascular resistance results in pulmonary artery hypertension and right ventricular hypertrophy
 - Combination of decreased left ventricular volume and dysfunction results in diminished cardiac output
 - Atrial dilation with arrhythmias may occur if the MV obstruction is long standing
- Mitral Valve Insufficiency
 - Presentation dependent on etiology, severity, and age of onset

- Characterized by the reversal of blood flow from the LV to LA
 - Result of failure of valve to close completely during systole
 - Reduces cardiac output and causes LV dysfunction.
 - Decreases ventricular stroke volume and increases LA pressure
- Compensation for the diminished cardiac output includes increased stroke volume, ejection fraction, and heart rate
- The excess LA volume leads to gradual dilation of atrium
- The end result is congestive heart failure with pulmonary congestion, pulmonary hypertension, and atrial dilation with the onset of atrial arrhythmias
- Common causes of MR are chordal rupture, rheumatic heart disease, infective endocarditis, coronary artery disease, cardiomyopathy, surgical complication

Procedures/Interventions

- Intricate knowledge of the aortic valve, atrioventricular node, and circumflex artery required for successful mitral valve surgery
- Inspect valve before repair:
 - Begin at supra-valvar region
 - Assess leaflets for size, mobility, prolapse
 - Examine chordae and papillary muscles
- Surgical Indications
 - Symptomatic patients- intractable CHF, pulmonary HTN, progressive cardiomegaly, failure to thrive, intractable endocarditis, prosthetic valve dysfunction
 - Recurrent atrial fibrillation
 - Valve repair preferred over replacement in children regardless of stenosis or regurgitation
- Percutaneous mitral commissurotomy
 - Procedure of choice for mitral stenosis
 - Avoid surgical incision
 - Progressively enlarged balloon technique with balloon size determined by annular size
 - Diameter of balloon equals size of mitral annulus
 - Risk residual mitral stenosis or regurgitation
- Closed Commissurotomy
 - Left thoracotomy
 - Progressively sized dilators inserted across MV leaflets
 - Risk of residual mitral stenosis or creation increased regurgitation
- Open mitral commissurotomy
 - Access to papillary muscles and chordae necessary through the orifice of the valve
 - If valve orifice too small, excision of posterior leaflet required
 - Depending on anatomy of papillary muscle perform thinning, mobilization from the posterior wall, spitting and fenestration of papillary muscles
 - Following this reconstruction of posterior leaflet with enlargement of valvar tissue if needed
 - Can enlarge either anterior or posterior leaflet

- Risk residual stenosis or increased regurgitation
- Valve replacement
 - Indications
 - Calcified valves
 - Moderate to severe MR
 - Malfunction or small size of previously placed prosthetic MV
 - Type of valve
 - Limitations on available sizes
 - Prosthetic valves
 - Longer durability
 - Require long-term anticoagulation therapy
 - Bioprosthetic valves
 - Do not require anticoagulation
 - Deteriorate quicker
 - Mitral valvuloplasty
 - Annuloplasty required for all valve repairs, limited to just posterior leaflet if annulus less than adult size
 - Chordal shortening if chordae thin and flexible
 - Chordal transfer for localized prolapse
 - Wedge resection and sliding plasty for prolapses of multiple chordae
- Catheterization Indications
 - Balloon dilatation valvuloplasty
 - Effective for rheumatic MS but less effective for congenital MS.
 - Procedure:
 - Passage of balloon catheter across the atrial septum, into mitral valve orifice
 - Inflation of balloon to tear valve leaflet, dilate annulus
 - Complications
 - Perforation of the LV
 - Transient complete heart block
 - Tearing/perforation of the anterior leaflet of the MV and severe MR
 - MitraClip for severe MR
 - Effective in inoperable, adult patients with severe MR
 - Procedure
 - Percutaneous placement of a clip on free edges of both anterior and posterior leaflet
 - Central placement of clip to decrease valve orifice
 - May require more than one clip
 - Complications
 - Damage to leaflets
 - Movement of device

Specific Considerations

- Preoperative
 - Medical management preferred in infants and small children
 - Goal is alleviate symptoms and promote growth and development
 - Digoxin for function may be helpful
 - Diuretics and afterload reduction for MR
 - Supportive feeding for weight gain
 - Oxygen if pulmonary pressures elevated may be necessary
 - Sildenafil for severe pulmonary hypertension
- Post-Operative
 - Post-Operative TEE
 - Evaluate repair, coronary compromise, perivalvular leak and leaflet motion with placement of prosthetic MV
 - Need for surgical revision of valve seating
 - Need for support of ventricular function with inotropes/vasoactives
 - Direct left atrial/left ventricle pressure measurement for residual mitral stenosis
 - Pacing or medication for heart block or arrhythmia
 - Control atrial arrhythmias
 - Preserve cardiac output
 - Mechanical ventilation
 - Afterload reduction on LV
 - Support of preload/pulmonary edema
 - Manipulation of elevations in PVR
 - Afterload reduction
 - Residual mitral regurgitation
 - Myocardial ischemia
 - Use with caution in mitral stenosis
 - Anticoagulation (See Neonatal/Pediatric and Adult Anticoagulation Guideline)
 - Heparin when postoperative bleeding subsided, usually day one
 - Coumadin when able to take oral medications
 - Complications
 - Bleeding
 - Low cardiac output
 - Support of LV diastolic dysfunction with volume and afterload reduction
 - Support of LV systolic dysfunction with inotropic agents
 - Support of PAH with oxygen, inhaled nitric oxide, sedations, mechanical ventilation at functional residual capacity
 - Support of heart rate and rhythm with medication and/or pacing
 - Complete heart block
 - Most common in infants and children with MV replacement
 - If annulus is hypoplastic may require excision of leaflet tissue along the crest of the ventricular septum and valve replacement in the region of the AV node and bundle of His
 - Hemolysis
 - May occur from prosthetic valve replacement

- Associated with perivalvular leak
- Hemoglobinuria, anemia, elevated LDH

Long-term problems/complications and routine care

- Endocarditis prophylaxis is necessary (See 2015 AHA Guidelines for both pediatric and adult endocarditis recommendations)
- Long-term monitoring for potential problems
 - Mitral valve function and gradient
 - Acute prosthetic valve dysfunction resulting in low cardiac output syndrome (LCOS)
 - Valve obstruction
 - Residual mitral stenosis following surgical repair
 - Progressive mitral stenosis due to somatic growth
 - Prosthetic valve obstruction
 - Clots
 - Tissue ingrowth (Pannus)
 - Stenosis due to annular seating, patient anemia, arrhythmia or somatic growth
 - Malfunction of leaflets
 - Left atrial dilation with compression bronchus and resultant respiratory compromise
 - Development of LV dysfunction
 - Development of PAH and PVOD from long standing mitral stenosis
 - Short and long term treatment of elevated PVR/PAH
 - Anticoagulation (See Neonatal/Pediatric and Adult Guidelines on Anticoagulation)
 - For mechanical prosthetic valve
 - For atrial arrhythmias
 - Arrhythmias (See Ped/Neo and Adult Guidelines on Arrhythmias)
 - Post-operative development of heart block
 - Atrial arrhythmias
 - Pacemaker follow-up if indicated
- Growth failure and developmental delay related to CHF, hospitalizations

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.

Baylen, B. G., & Atkinson, D. E. (2008). Mitral inflow obstruction. In Moss and Adams' Heart Disease in Infants, Children, and Adolescents (7th ed). Philadelphia, PA: Lippincott Williams & Wilkins.

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.

Bouleti, C., Lung, B., Himbert, D., Brochet, E., Messika-Zeitoun, D., Detaint, D., Garbarz, E., Cormier, B., Vahanian, A. (2013). Long-term efficacy percutaneous mitral commissurotomy for restenosis after previous mitral commissurotomy. *Heart*:99:1336-1341. Doi:10.1136/heartjnl-2013-303944.

Brizard, C. (2007). Mitral valve repair in children. *Mastery of Cardiothoracic Surgery* (2nd ed) Philadelphia, PA:Lippin cott Williams and Wilkins.

Hansen, M. (1998). Pathophysiology: Foundations of Disease and Clinical Intervention. Grand Forks, ND: W.B. Saunders Company.

Kanjanauthai, S., & Lange, R.A. (2013). Mitral Valve Anatomy. Retrieved from Emedicine.medscape.com/article/1878301

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

Nigro, J. J., Bart, R. D., & Starnes, V. (2006). Mitral valve diseases. In *Critical Heart Disease in Infants and Children* (2nd ed). Philadelphia, PA: Mosby.

Park, M. K. (2008). Valvular heart disease. In *Pediatric Cardiology for Practitioners* (5th ed). Philadelphia, PA: Mosby.

Rifaie, O., Abdel-Dayem, M. K., Ramzy, A., Ez-El-din, H., El-Ziady, G. et al. 2008. Percutaneous mitral valvotomy versus closed surgical commissurotomy. Up to 15 years of follow-up of a prospective randomized study. *Journal of Cardiology*, 53, 28-35. doi:10.1016/j.jjcc.2008.08.003.

Speicher, R., & Jennings, A., (2015). Mitral valve stenosis. In *Pediatric Cardiac Intensive Care Handbook*. Washington, D. C.: Pediatric Cardiac Intensive Care Books.

Seguela, P. E., Houyel, L., & Acar, P. (2011). Congenital Malformations of the Mitral Valve. *Archives of Cardiovascular Disease*, 104, 465-479.

Slota, M. C. (Ed.). (2006). Core Curriculum for Pediatric Critical Care Nursing (2nd ed.). St. Louis, MI: Elsevier.

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