Tricuspid 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
  - Wise spectrum of congenital defects in tricuspid 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**
- Sits between right atrium (RA) and right ventricle (RV) (See illustration below for location of tricuspid valve in relation to other cardiac structures)

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- Composed of three leaflets
  - Unequal in size
  - Attach to papillary muscles in RV by chordae tendineae
- Acquired disease of the tricuspid valve is uncommon
  - May result from endocarditis
    - Rheumatic fever
    - Illicit intravenous drug use
    - Long-term indwelling intravenous catheters, including dialysis catheters
    - Additional septal defects
- Right ventricular dysfunction, dilation
- Tricuspid Atresia (See illustration below, also see Defect Document on Tricuspid Atresia)
  - Definition: absence of the tricuspid valve with no direct communication between the RA and the RV
  - All variations of tricuspid atresia share the following characteristics
    - Lack of communication between the atrium (most commonly the right atrium) and the RV (Number 1 on illustration)
    - Presence of an interatrial communication (Number 4 on illustration)
    - Enlarged mitral valve
    - Usually associated with some degree of right ventricular hypoplasia
  - Three classifications of tricuspid atresia
    - Based on ventriculoarterial relationship
    - Subdivided based on presence/size of a VSD (Number 3 on illustration) or restriction to pulmonary blood flow (Number 5 on illustration)
    - Type I: normally related great arteries, occurrence 70% to 80%
      - Type Ia: VSD and associate pulmonary atresia
      - Type Ib: small VSD and some restriction to pulmonary blood flow
      - Type Ic: large VSD and no pulmonary stenosis
    - Type II: D-transposition of the great arteries, occurrence 12% to 25%
      - Type IIa: pulmonary atresia
      - Type IIb: pulmonary stenosis
      - Type IIc: no obstruction into the transposed pulmonary artery
    - Type III: used to describe patients with more complex lesions, such as truncus arteriosus or atrioventricular septal defect, and malposed great arteries, occurrence 3-6%

Tricuspid Atresia
• **Tricuspid Stenosis**
  - Valvar abnormalities
    - Large annulus
      - Thickened leaflets are thickened with
      - Commisures fused
      - Short chorea tendineae
    - Hypoplastic annulus
      - Small leaflets and chordae
      - Structurally normal
  - Usually associated with other anomalies
    - Right ventricular (RV) outflow tract obstruction
    - Atresia with hypoplasia of the RV

• **Tricuspid Regurgitation**
  - Rare unless associated with Ebstein’s anomaly
  - Anatomic abnormalities
    - Nodular thickening of the valve leaflets with shortened chordae tendineae and hypoplastic or absent papillary muscles
    - Isolated cleft of a valve leaflet
    - Complete absence of valve tissue
    - Associated with Ebstein’s anomaly (discussed below)
  - Secondary to:
    - Other lesions, such as severe stenosis or atresia of the RV outflow tract
    - Intrauterine or perinatal event resulting in RV and papillary muscle dysfunction
    - Surgical complication or residual lesion such as AVSD
    - Systemic RV dysfunction and tricuspid valve regurgitation in previous corrected congenital lesions (s/p Senning or Mustard for transposition of the great arteries)
    - Systemic RV dysfunction with tricuspid regurgitation in congenital defects such as LTGA

• **Ebstein’s Anomaly** (For Defect Document for Ebstein’s Anomaly)
  - Abnormal attachment of the posterior and septal tricuspid leaflets in the RV
    - Downward displacement of the valve leaflets
    - Varying degrees of adherence to the RV wall
  - Abnormal anterior leaflet
    - Normally attached at the annulus
    - Enlarged, “sail-like”
    - Often abnormally tethered to the RV wall
    - May obstruct the RV outlet
  - Abnormal tricuspid valve orifice (See illustration below. Number 1 illustrates the displaced tricuspid valve)
    - Displaced downward into the RV at the junction of the inlet and the trabecular components of the RV
- Inlet portion of the RV integrated into the right atrium (Number 2 illustrates the RA with the integrated inlet portion of the RV)
- Functional RV
  - Trabecular and outlet portions of the RV
  - Small RV cavity

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**Ebstein’s Anomaly**


- May be associated with Wolfe-Parkinson-White (WPW) or other atrial reentry tachycardia

**Physiology**

- Tricuspid Atresia (See Defect Document on Tricuspid Atresia) (See above illustration on Tricuspid Atresia)
  - Obligatory right-to-left shunt at the atrial level (Number 4 on illustration)
  - Requires communication between the systemic and pulmonary circulations
    - Ventricular septal defect (VSD) (Number 3 on illustration)
    - Patent ductus arteriosus
  - Pulmonary blood flow
    - May be restricted
    - Associated lesions
      - Valvar: pulmonary valve stenosis, pulmonary annular hypoplasia, pulmonary atresia (Number 5 on illustration)
      - Severe hypoplasia of the right ventricular outflow tract (RVOT)
  - Position of great arteries:
    - Normally related
    - Blood flow: Systemic venous blood from the right atrium (RA) shunts right to left, mixes with pulmonary venous return, flows across the mitral valve (MV) into the left ventricle (LV)
- Variations:
  - VSD and a patent RV outflow track:
    - Blood ejected into the aorta and shunts left to right across the VSD to the pulmonary artery. (Number 2 on illustration) Pulmonary blood flow determined by:
      - Size of the VSD
      - Degree of outflow track obstruction
      - No obstruction –
        - Pulmonary overcirculation
        - Neonate: may develop as pulmonary vascular resistance decreases in the newborn period
      - Restrictive/intact VSD/some degree of pulmonary stenosis/ataresia
        - Cyanosis
        - Neonate: adequate pulmonary blood flow dependent on PDA
  - Transposed
    - Blood flow: Systemic venous blood from RA shunts to the left atrium (LA) and empties into the LV. Size of the VSD and presence/degree of pulmonary outflow obstruction may compromise systemic or pulmonary flow
      - Unrestricted pulmonary blood flow
        - Pulmonary overcirculation
        - Neonate: May develop as pulmonary vascular resistance decreases in the newborn period
      - May see congestive heart failure
      - Obstruction to systemic blood flow
        - From restrictive VSD or infundibular narrowing
        - Severe obstruction: May result in hypotension, shock, and metabolic acidosis

- Tricuspid Stenosis
  - Symptoms resemble that seen in tricuspid atresia
  - Physiology depends on:
    - Size of the RV
    - Presence/size of a VSD
    - Degree of obstruction to pulmonary blood flow

- Tricuspid Regurgitation
  - Rare to be seen as an isolated condition at any age
  - Symptoms usually present in the newborn period
    - Cyanosis
    - Congestive heart failure
  - Physiology depends on degree of valve dysfunction

- Ebstein’s Anomaly (See Defect Document on Ebstein’s Anomaly)
- Wide spectrum of pathology (See illustration above)
  - Hemodynamic compromise related to:
    - Extent of downward displacement of the leaflets
    - Severity of tricuspid regurgitation
    - Degree of RV outlet obstruction
    - Reduced chamber capacity of the RV
    - Degree of myocardial dysfunction
    - Presence of other associated cardiac abnormalities
- Mild displacement with minimal insufficiency
  - Symptoms may be absent
- More severe displacement with increased insufficiency
  - Cyanosis from:
    - Right-to-left shunt at the atrial level due to RA pressure > LA pressure
    - Decreased RV filling
      - Abnormal contraction pattern of the atrialized portion of the RV
      - Blood flows back into true RA instead of forward to true RV during ventricular systole
    - Elevated PVR in the neonatal period
    - Obstruction to pulmonary blood flow due to varying degrees of pulmonary stenosis or possibly atresia
  - Neonate:
    - Increased PVR
    - RV may be unable to generate enough antegrade pulmonary blood flow
    - Pulmonary blood flow may be dependent on a PDA
- Intact atrial septum
  - No cyanosis
  - Symptoms of increased RA pressure
    - Hepatosplenomegaly
    - Severe regurgitation and/or decreased RV function
      - Low cardiac output
      - Shock
      - Cardiovascular collapse

**Procedures/Interventions**
- Tricuspid Atresia (See Defect Document on Tricuspid Atresia for illustrations on procedures)
  - Requires staged palliation for lesions that result in single-ventricle anatomy
  - Decreased pulmonary blood flow
    - Defects: tricuspid atresia with absence of/restrictive VSD and/or pulmonary stenosis/atresia
    - Surgical placement of an aortopulmonary shunt (modified Blalock-Taussig (BT) shuntcentral aortopulmonary shunt)
• Necessary to augment pulmonary blood flow
  o Excessive pulmonary blood flow
    • Defects: tricuspid atresia with normally related great vessels and no pulmonary stenosis, or tricuspid atresia with transposition of the great arteries
    • Pulmonary artery banding to restrict pulmonary blood flow
    • Ligation of the pulmonary artery with placement of an aortopulmonary shunt to control pulmonary blood flow
  o Restricted systemic blood flow
    • Anastomose main pulmonary artery (MPA) to the aorta (Damus-Kaye Stansel operation)
      • Alleviate subaortic obstruction
      • Promote coronary flow
      • Provide pulmonary blood flow with placement of an aortopulmonary shunt

• Tricuspid Stenosis
  o Surgical repair
    • Commissurotomy, not very successful in infants
    • Interventions less successful in infants
  o Valve Replacement
    • Bioprosthetic valve preferred over 7 years of age
    • High rate of thrombosis of mechanical valve

• Tricuspid Regurgitation
  o May resolve without intervention
    • Relatively normal tricuspid valve structure
    • Insufficiency related to ventricular or papillary muscle dysfunction
  o Surgical interventions
    • Generally not indicated for valve repair (especially in neonates and infants)
    • May be necessary for management of RV outflow tract obstruction
    • May be necessary if result of surgical complication (AVSD repair, VSD repair with damage to TV apparatus)

• Ebstein’s Anomaly (See Defect Document on Ebstein’s Anomaly)
  o Intervention indicated based on symptoms of patient. Type of intervention based on age of patient at presentation
  o Adolescents and adults:
    • Repair or replacement depending on degree of regurgitation and anatomy of the valve
  o Neonates and infants
    • Palliation: single ventricle palliation
      • Ebstein’s malformation and severe pulmonary stenosis, pulmonary atresia, or absent pulmonary valve
    • Repair
      • One-and-a-half ventricle repair
        o Neonatal Ebstein’s in severe CHF
        o In place of single ventricle palliation
• See Alternative Surgical Strategies below
  o Pediatric surgical interventions
    ▪ Valve Repair
      • Preferable to valve replacement
      • May not be possible due to:
        o Extensive, atrialized RV
        o Anterior leaflets adherent to ventricular wall
        o Absent chordae and papillary muscles
    ▪ Valve Replacement
      • Limited availability of prosthetic valve
        o Size
        o Type
      • No growth of valve
      • Requires anticoagulation
      • See Alternative Surgical Strategies below
  o Alternative surgical strategies
    ▪ Single ventricle palliation
      • Ebstein’s anomaly in severe CHF, pulmonary atresia, or absent pulmonary valve
      • Critically ill neonates with severe tricuspid regurgitation
      • Initial procedure
        o Septectomy
        o Neo-aortic reconstruction
        o Placement of arteriopulmonary shunt
        o RV exclusion by placement of a fenestrated patch on the tricuspid valve annulus
          ▪ Alleviates poor RV function
          ▪ Alleviates tricuspid regurgitation
          ▪ Alleviates RV outflow tract obstruction
        o Subsequent procedures
          ▪ Glenn/hemi-Fontan
          ▪ Completion Fontan
    ▪ One-and-a-half ventricle repair
      • May be considered to decrease the risk of volume stress on a marginal tricuspid valve and RV
      • Indications
        o Tricuspid valve apparatus or RV cavity appear not conducive to a two-ventricle repair
          ▪ RV diastolic volumes between 45% and 90% of predicted normal
          ▪ RV dysfunction
          ▪ Severe tricuspid valve disease
        o Limited cardiac output through right heart structures
        o Decrease the risk of postoperative RV failure
        o Decrease volume stress on a marginal tricuspid valve
        o Delaying operation allows:
- Potential development of the RV
- Decrease in regurgitation of TV
- Allows decrease in PVR
  - Surgical procedure
    - Bidirectional Glenn/hemi-Fontan
      - Connect right superior vena cava (SVC) to right pulmonary artery (RPA)/main pulmonary artery
    - Downsizing or patch closure of the atrial septal defect - depends on function of RV
    - Allows SVC circulation to drain directly to the lungs and the hypoplastic RV/TV carries only inferior vena cava (IVC) circulation
- Two ventricle repair
  - TV repair
    - Attempted in patients with enough muscular RV to maintain CO
    - Transposes the tricuspid valve leaflets to the tricuspid annulus
    - Eliminate the atrialized ventricle
    - Minimize tricuspid regurgitation
    - Relieve RV outflow obstruction
  - TV replacement
- Transplant
  - Neonates with Ebstein’s malformation and severe pulmonary stenosis, pulmonary atresia, or absent pulmonary valve
  - Neonatal Ebstein’s in severe CHF in place of single ventricle palliation
  - Severe TV anomalies
  - Hypoplastic RV
  - Associated pulmonary valve anomalies
- Arrhythmias
  - Ebstein’s Anomaly
    - Associated with WPW and atrial tachyarrhythmias
    - Recurrent SVT
      - Causes decreased cardiac output
      - Will require treatment preoperatively or postoperatively
  - Surgical or catheter ablation
  - Medication
    - Neonatal period
      - May control the SVT
      - Allow time for patient to grow
    - May still be required following ablation if persistent SVT/atrial tachycardia
  - Anti tachycardic (ICD) pacemaker
Specific considerations and routine care

- Respiratory distress & failure
  - Lung hypoplasia
    - May result from cardiomegaly
    - Seen especially in neonatal period
  - Possible pulmonary insufficiency
  - May require increased respiratory support
    - Positive Pressure Ventilation

- Ventricular dysfunction
  - Congestive heart failure
    - May result in both left and right ventricular dysfunction
    - Due to tricuspid insufficiency
      - Causes RV dilation
      - Underdeveloped LV
  - Right ventricular dysfunction and failure very common
    - Due to hypoplasia
    - Strategies should be aimed at right ventricular afterload reduction

- Valve Regurgitation
  - Strategies should be aimed at decreasing PVR

- Arrhythmias
  - Atrial tachycardias most common
  - Electrical and/or pharmacologic management can be used to restore atrioventricular synchrony

- Routine care
  - Periodic routine post-operative monitoring
  - Focus examination for:
    - Atrial arrhythmias
    - RV afterload reduction
    - Decreasing PVR

Long-term problems/complications and routine care

- Tricuspid atresia (See Defect Document on Tricuspid Atresia)
  - Single ventricle monitoring
    - Balanced pulmonary and systemic circulation
    - Ventricular function
    - Arrhythmias
  - Single ventricle complications
    - Repeat operations
    - Potential Fontan failure/ventricular function
    - Neurodevelopmental problems

- Valve replacement/palliation
  - Monitoring for function
    - Re-stenosis
    - Regurgitation with serial echocardiograms
  - Long-term growth restriction of prosthetic valves
  - Anticoagulation (See both Peds/Neo and Adult Guidelines on Anticoagulation)
• Ebstein’s Anomaly (See Defect Document for Ebstein’s Anomaly)
  o Valve function
  o Arrhythmias
  o Exercise tolerance

References:


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