Tricuspid Valve

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

Lindsey Justice, DNP, RN, CPNP-AC,
Nurse Practitioner, Cardiac Intensive Care Unit,
Cincinnati Children's Hospital Medical Center

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

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

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

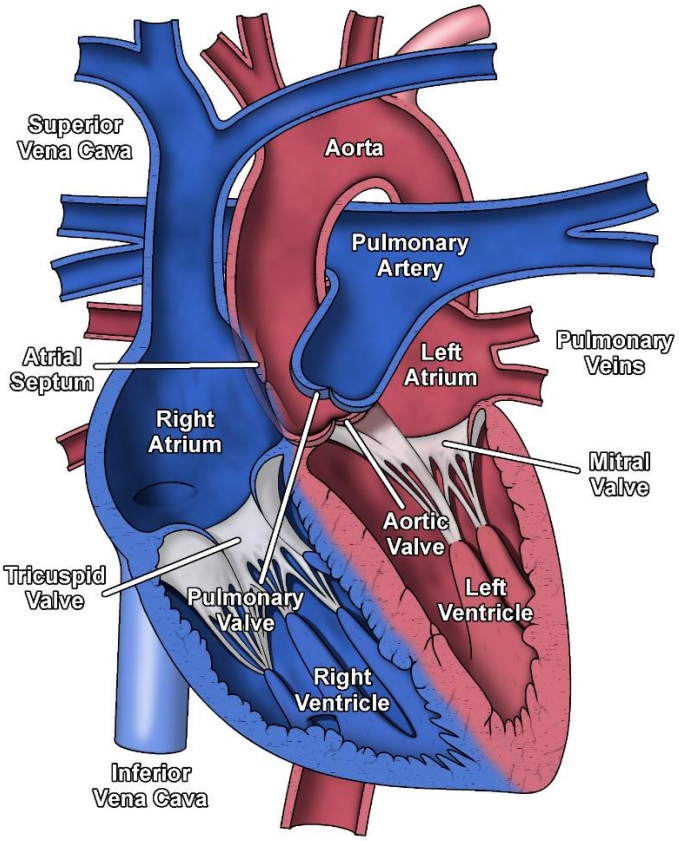
Embryology

- 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
  - 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)



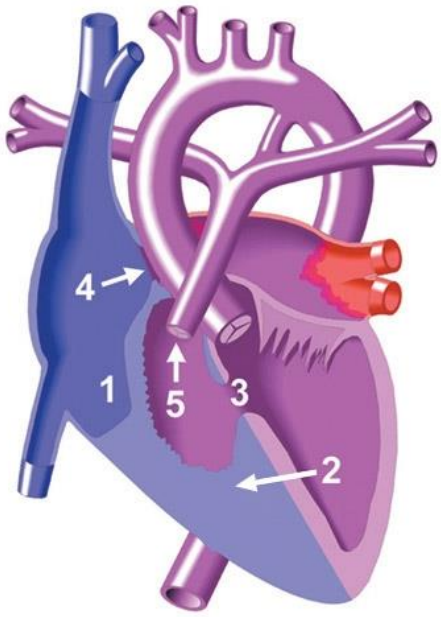
Normal Heart

- 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

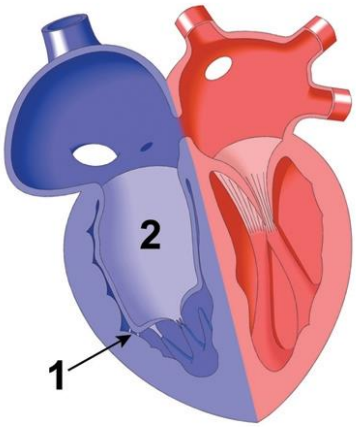
    Type III: used to describe patients with more complex lesions, such as truncus arteriosus or atrioventricular septal defect, and malposed great arteries, occurrence 3-6%



Tricuspid Atresia

- Tricuspid Stenosis
  - Valvar abnormalities
    - Large annulus
      - Thickened leaflets are thickened with
      - Commissures 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



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/atresia
                - 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) shunt/central aortopulmonary shunt)

        - Necessary to augment pulmonary blood flow
    - 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
    - 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
    - Surgical repair
        - Commissurotomy, not very successful in infants
        - Interventions less successful in infants
    - Valve Replacement
        - Bioprosthetic valve preferred over 7 years of age
        - High rate of thrombosis of mechanical valve
- Tricuspid Regurgitation
    - May resolve without intervention
        - Relatively normal tricuspid valve structure
        - Insufficiency related to ventricular or papillary muscle dysfunction
    - 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)
    - Intervention indicated based on symptoms of patient. Type of intervention based on age of patient at presentation
    - Adolescents and adults:
        - Repair or replacement depending on degree of regurgitation and anatomy of the valve
    - 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
                - Neonatal Ebstein's in severe CHF
                - In place of single ventricle palliation

- See Alternative Surgical Strategies below

- Pediatric surgical interventions
  - Valve Repair
    - Preferable to valve replacement
    - May not be possible due to:
      - Extensive, atrialized RV
      - Anterior leaflets adherent to ventricular wall
      - Absent chordae and papillary muscles
  - Valve Replacement
    - Limited availability of prosthetic valve
      - Size
      - Type
    - No growth of valve
    - Requires anticoagulation
    - See Alternative Surgical Strategies below
- 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
      - Septectomy
      - Neo-aortic reconstruction
      - Placement of arteriopulmonary shunt
      - 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
      - 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
      - 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
      - Limited cardiac output through right heart structures
      - Decrease the risk of postoperative RV failure
      - Decrease volume stress on a marginal tricuspid valve
      - 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)
    - Valve function
    - Arrhythmias
    - Exercise tolerance

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