National Education Curriculum

Specialty Curricula

Cardiac

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Section I: Anatomy and Physiology of the Heart

Rationale: Accurate assessment and performance of echocardiograms requires sonographers to assemble a comprehensive knowledge of the anatomy, embryology, physiology, pathophysiology and echocardiographic appearances of the heart and surrounding structures. An understanding of these is essential for the performance of high quality examinations and is requisite to quality patient care.

1. Describe the heart anatomy
2. Explain the blood flow through the coronary arteries
3. List the events that lead to myocardial contraction
4. Compare and contrast the anatomy of aorta and vena cava
5. Describe the location and function of each portion of the cardiac conduction system
6. Describe the anatomy of the pericardium, the myocardium, and endocardium
7. Explain the relational anatomy of the heart in the thoracic cavity
8. Describe the location of the apex in relationship to other structures in the heart
9. Explain how body habitus influences the position of the heart in the thoracic cavity

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I. Anatomy and Physiology of the Heart

- A. Chambers and Related Septa
  - 1. Morphologic and anatomic features
    - a. Right atrium (RA)
      - i) Eustachian valve
      - ii) Chiari network
      - iii) Crista terminalis
    - b. Right ventricle (RV)
      - i) Moderator band
    - c. Left atrium (LA)
      - i) Left atrial appendage (LAA)
    - d. Left ventricle (LV)
    - e. Interatrial septum (IAS)
      - i) Septum primum
      - ii) Septum secundum
      - iii) Fossa ovalis
    - f. Interventricular septum (IVS)
      - i) Inlet
      - ii) Trabecular
      - iii) Infundibular (outlet)
      - iv) Membranous
    - g. Coronary sulcus
    - h. Atrioventricular groove
    - i. Crux of the heart
- B. Valves and Related Apparatus

1. Cardiac valves
    a. Atrioventricular valves (AV)
        i) Mitral valve (MV)
        ii) Anterior leaflet
        iii) Scallops (A1, A2, A3)
        iv) Posterior leaflet
        v) Scallops (P1, P2, P3)
        vi) Papillary muscles
        vii) Chordae tendinae
        viii) Annulus
        ix) Tricuspid valve (TV)
        x) Septal leaflet
        xi) Posterior leaflet
        xii) Anterior leaflet
        xiii) Papillary muscles
        xiv) Chordae tendinae
        xv) Annulus
    b. Semilunar valves
        i) Aortic valve (AV)
        ii) Right coronary cusp (RCC)
        iii) Left coronary cusp (LCC)
        iv) Non-coronary cusp (NCC)
        v) Sinus of Valsalva
        vi) Annulus
        vii) Commissures
        viii) Subvalve, supravalve, LVOT
        ix) Pulmonic valve (PV)
        x) Leaflets
        xi) Subvalve
        xii) Supravalve
        xiii) Infundibulum RVOT

C. Arterial-Venous System
    1. Aorta (Ao)
        a. Origination and termination
        b. Anatomic location
        c. Segments
            i) Aortic annulus
            ii) Sinus of Valsalva

iii) Sinotubular junction
iv) Ascending Ao
v) Ao Arch
vi) Descending Ao
vii) Abdominal Ao

d. Major branches
i) Brachiocephalic (innominate)
ii) Left common carotid
iii) Left subclavian

2. Pulmonary artery
a. Branches
i) Main pulmonary artery (MPA)
ii) Pulmonary artery bifurcation
iii) Right pulmonary artery (RPA)
iv) Left pulmonary artery (LPA)

b. Coronary arteries
i) Left coronary artery
- Left anterior descending (LAD)
- Circumflex

ii) Right coronary artery (RCA)
iii) Congenital variations
iv) Flow patterns
v) Flow reserve

3. Superior vena cava (SVC) and inferior vena cava (IVC)
4. Pulmonary veins
5. Coronary venous system
a. Coronary sinus
b. Differentiation from descending Ao
c. Atrioventricular groove

6. Vessel wall layers
a. Tunica intima
b. Tunica media
c. Tunica adventitia

D. Layers of the Heart
1. Pericardium
a. Visceral
b. Parietal

2. Myocardium

a. Layers
    i) Subepicardium
    ii) Midwall
    iii) Subendocardium
3. Endocardium

E. Relational Anatomy
1. Mediastinum anatomy
2. Heart position
    a. Levocardia
    b. Mesocardia
    c. Dextrocardia
    d. Levoposition
    e. Dextroposition
    f. Mesoposition
3. Thoracoabdominal situs
    a. Solitus
    b. Inversus
    c. Ambiguous
4. Atrial situs
    a. Solitus
    b. Inversus
    c. Ambiguous
5. Atrioventricular connection
6. Ventriculoarterial connection
7. Great vessel relationship

F. Pulmonary Versus Systemic Circulation
1. Components of the pulmonary circulation
    a. Right atrium (RA)
    b. Right ventricle (RV)
    c. Main pulmonary artery (MPA) and branches
    d. Pulmonary capillaries
    e. Pulmonary veins
2. Components of the systemic circulation
    a. Left atrium (LA)
    b. Left ventricle (LV)
    c. Aorta (Ao) and branches
    d. Systemic capillary network
    e. Cerebral, peripheral and abdominal veins

f. SVC
g. IVC
h. Hepatic veins

Section II: Basic Embryology

1. Sequence the formation of the heart beginning with the primitive heart tube through the development of the six aortic arches
2. Describe cardiac septation including atrial and ventricular
3. Describe the formation of the atrioventricular valves and the semilunar valves
4. Compare fetal and neonatal circulation

II. Basic Embryology of the Heart

A. Primitive Heart Tube

1. Development sequence of heart tube
2. Primitive vascular tube
   a. Sinus venosus
   b. Cardiac loop
   c. Bulbus cordis/truncus arteriosus
3. Bulboventricular loop
   a. Dextrolooping (D-loop)
   b. Levolooping (L-loop)
4. Regions of heart tube
   a. Sinus venosus
   b. Primitive atria
   c. Atrioventricular canal
   d. Primitive ventricle
   e. Bulbus cordis
   f. Truncus arteriosus
   g. Aortic sac and arches
5. Septation
   a. Endocardial cushion
   b. Atrioventricular canal
   c. Truncoconal region
   d. Atrial septum formation
      i) Septum primum
      ii) Septum secundum
   e. Interventricular septum (IVS) formation
6. Six aortic arches
   a. Maxillary arteries branches from external carotid artery (ECA) - 1st pair
   b. Stapedial arteries - 2nd pair
   c. Proximal common carotid artery (CCA) and distal internal carotid artery (ICA) - 3rd pair
   d. Ao Arch (L) - left 4th pair

e. Proximal right subclavian (R) - right 4th pair
f. Rudimentary - 5th pair
g. (L) Left pulmonary artery and ductus - left 6th pair
h. Right pulmonary artery - right 6th pair

7. Cardiac valve formation
   a. The atrioventricular valves
   b. The semilunar valves

B. Comparison of Fetal and Postnatal Circulation

1. Prenatal circulation
   a. Umbilical vein
   b. Ductus venosus
   c. Eustachian valve
   d. Foramen ovale
   e. Ductus arteriosus
2. Neonatal circulation
   a. Ligamentum arteriosum
   b. Fossa ovalis
   c. Ligamentum venosum
   d. Ligamentum teres

Section III: The Echocardiographic Examination

1. Describe a 2-D echocardiographic examination using proper nomenclature
2. List equipment controls commonly used for each mode
3. Identify common M-mode patterns associated with cardiac disease
4. Associate the best Doppler approach based on location of various cardiac diseases
5. Apply pertinent Doppler formulas to the cardiac setting
6. Explain provocative, positional and breathing maneuvers that affect venous inflow and cardiac output
7. Explain the appearance of echocardiographic artifacts on an image
8. Describe myocardial segmentation
9. Differentiate between all normal anatomy visualized on an echocardiogram
10. Describe the coronary arterial system

III. The Echocardiographic Examination

A. Basic Imaging Principles
   1. Tomographic planes
   2. Nomenclature
   3. Image orientation
   4. Technical quality

B. Scanning Views
   1. Parasternal
      a. Long-axis
         i) Ascending aorta
         ii) Aortic root
         iii) Aortic valve (AV)
         iv) Left ventricular outflow tract (LVOT)
         v) Left atrium (LA)
         vi) Inlet portion of left ventricle
         vii) Interventricular septum IVS
         viii) Mitral valve (MV)
         ix) Descending thoracic Ao
      b. Long-axis right ventricular inflow tract (RVIT)
         i) Inlet portion of the right ventricle (RVIT)
         ii) Tricuspid valve (TV)
         iii) RA
         iv) RA appendage
      c. Long-axis right ventricular outflow tract (RVOT)
         i) Outlet portion of RV
         ii) PV
         iii) Main pulmonary artery (MPA)
      d. Short-axis

e. High parasternal
f. Level of aortic valve, great vessels and atria
g. TV
h. RVOT
i. PV
j. MPA bifurcation
k. Coronary arteries
l. RCA
m. LMA
n. LAD
o. Circumflex artery
p. Left atrial appendage (LAA)
q. Left ventricle
r. Level of the mitral valve leaflets (basal-level)
s. Papillary muscles (mid-level)
t. Anterior leaflet
u. Apex
v. Posterior leaflet IVS
w. RV
x. Left ventricle papillary muscles (mid-level)
y. Anterior lateral
z. Posterior Medial
aa. IVS
bb. RV

2. Apical views
   a. 4-chamber
      i) Ventricles and atria
      ii) MV
      iii) TV
      iv) Pulmonary veins
      v) Coronary sinus (posterior tilt)
      vi) IVS
      vii) IAS
   b. Apical 4-chamber with aortic valve (apical 5-chamber)
      i) LV
      ii) LA
      iii) LVOT (anterior tilt)
      iv) AV (anterior tilt)

v) Proximal aortic root (anterior tilt)
vi) Descending thoracic Ao

c. 2-chamber
i) LV
ii) LA
iii) MV
iv) Left atrial appendage (LAA)
v) Descending thoracic Ao

d. Long-axis 3-chamber
i) LV
ii) LA
iii) LVOT
iv) MV
v) AV

3. Subcostal views
a. 4-chamber view
i) Chambers and atria
ii) MV
iii) TV
iv) IAS
v) IVS

b. 4-chamber with aortic valve
i) LVOT
ii) AV

c. Short-axis views
i) Left ventricle
ii) Mitral valve cusps (basal-level)
iii) Papillary muscles (mid-level)
iv) Apex
v) IAS
vi) Origin of coronary arteries

d. Inferior vena cava (IVC)/RA
i) Hepatic veins

e. Bicaval view
i) SVC
ii) IVC
iii) IAS

4. Suprasternal views

a. Long-axis of aortic arch
    i) Ascending
    ii) Transverse
    iii) Descending
    iv) Branch vessels
b. Short-axis of aortic arch
c. SVC
d. Pulmonary veins

5. Other acoustic windows
    a. Mid clavicular
    b. Right parasternal
    c. Posterior

C. Echocardiography Anatomy and Physiology

1. Orientation of images
    a. Terminology
    b. Display
2. Coronary artery distribution
3. Left Ventricle (LV)
    a. Dimensions, area, volumes
    b. LV mass, wall thickness
    c. Function
        i) Global systolic
        ii) Regional systolic
            - 16 segments – regional wall motion
            - 17 segments – perfusion
        iii) Diastolic
    d. Interdependence of LV and RV
    e. LV wall segments
        i) Subdivisions of heart
            - Base
            - Mid
            - Apical
        ii) Normal anatomic location of walls in planes
            - Parasternal long-axis
            - Parasternal short-axis sweep
            - Apical 4-chamber
            - Apical 2-chamber
            - Apical long-axis

f. Subdivision of LV
    i) Left ventricular inflow tract (LVIT)
        - Anatomic location
        - Functional location
        - Imaging views
    ii) Left ventricular outflow tract (LVOT)
        - Anatomic location
        - Functional location
        - Imaging views

4. Right ventricle (RV)
    a. Dimensions, area, volumes
    b. Global systolic function
    c. Echo findings with RV volume overload
    d. Echo findings with RV pressure overload
    e. Moderator band
    f. Subdivision of RV
        i) Right ventricular inflow tract (RVIT)
            - Anatomic location
            - Imaging views
        ii) Right ventricular outflow tract (RVOT)
            - Anatomic location
            - Imaging views
5. Left atrium (LA)
    a. Dimensions, area, volumes
    b. LA function
    c. Left atrial appendage
    d. Pulmonary veins
6. Valves
    a. Atrioventricular valves
        i) Composition
        ii) Function
        iii) Mitral valve
            - Anatomic location
            - Leaflet names and characteristics
        iv) Tricuspid valve
            - Anatomic location
            - Leaflet names and characteristics
    b. Semilunar valves

i) Composition
ii) Function
iii) Aortic valve
- Anatomic location
- Leaflet names and characteristics

iv) Pulmonic valve
- Anatomic location
- Leaflet names and characteristics

7. Great vessels
   a. Aorta
      i) Anatomic segments and location
      ii) Imaging views
   b. Pulmonary artery
      i) Anatomic location
      ii) Imaging views
8. Coronary sinus
   a. Anatomic location
   b. Characteristics
   c. Imaging views
      i) PLAX
      ii) Apical 4-chamber (posterior tilt)
   d. Differentiation of coronary sinus from surrounding structures
      i) Location
      ii) Sonographic appearance
9. Coronary arteries
   a. Anatomic locations
   b. Branching patterns
   c. Perfusion patterns
   d. Factors that influence coronary flow
      i) Heart rate (HR)
      ii) Diastolic aortic pressure
      iii) Left ventricular diastolic pressure
      iv) Degree of coronary stenosis
   e. Imaging views

D. Echocardiography Modalities
1. Two-Dimensional (2-D) echocardiographic examination
   a. Equipment controls
      i) Depth

ii) Dynamic range
iii) Edge enhancement
iv) Focal zones
v) Gain
vi) Harmonics
vii) Line density
viii) Processing curves
ix) Sector size
x) Transducer frequency
xi) Transmit power
xii) Zoom

b. Transducer manipulations
i) Rotation
ii) Angulation
iii) Sweep/Tilting (heel-toe)
iv) Probe pressure
v) Sliding

2. M-mode instrumentation
a. Equipment controls
i) Depth
ii) Gain
iii) Sweep speed

b. M-mode technique
i) 2-D guided cursor placement
ii) Advantages/disadvantages
iii) Clinical application
- Chamber and wall measurements
- Fractional shortening
- Findings with pathology

iv) Color M-mode
- Timing of events
- Clinical application

c. Imaging planes

3. Color Doppler
a. Equipment controls
i) Gain
ii) Color maps
iii) Invert

iv) Scale/PRF
v) Sector size
vi) Sector position
vii) Filters

b. Color characteristics
i) Flow directionality
ii) Flow disturbances
iii) Flow profiles
iv) Pulse wave mechanism
v) Advantages/disadvantages
vi) Frame rate
vii) Line density

c. Imaging planes

d. Clinical application
i) Valvular regurgitation
ii) Localization of stenotic valves and shunts

4. Spectral Doppler

a. Equipment controls
i) Gate position
ii) Gate size
iii) Angle correction
iv) Angle steering
v) Invert
vi) Transmit power
vii) Gain
viii) Reject/wall filter
ix) Velocity/range scale
x) Baseline shift
xi) Sweep speed

b. Doppler characteristics
i) Pulsed wave versus continuous wave
ii) Doppler equation
iii) Flow directionality
iv) Display
- Timing (systolic, diastolic)
- Direction (positive, negative)
- Quality (normal, disturbed)

v) Doppler tissue imaging (TDI)

- Low wall filter setting (high pass velocity setting)
- Low gain settings
- Color flow assignment
- Frame rate
- Scale adjustment
- Sample size or placement
- Respiratory variation
- Clinical use and formulas
- Waveforms
  - e' - early diastolic annular velocity
  - a' - annular velocity during atrial contraction
  - s'- systolic annular velocity

c. Standalone dedicated CW transducer
   i) Clinical application

d. Imaging planes

e. Clinical application
   i) Valvular stenosis or regurgitation
   ii) Assessment of intracardiac pressures
   iii) Shunts
   iv) Systolic function
   v) Diastolic function

5. Three Dimensional imaging

a. Transducer
   i) Matrix-array

b. Equipment controls
   i) Real time
   ii) Live
   iii) Simultaneous multiplane mode
   iv) Triplane mode
   v) Volume size
   vi) Single beat, multibeat
   vii) Zoom
   viii) Full volume
   ix) Gated
   x) Cropping
   xi) Rendering
   xii) 2D tomographic slices
   xiii) Color flow

xiv) Artifacts (over gain, under gain, stitch artifact, breathing)

xv) Limitations

6. Speckle tracking echocardiography (STE)
    a. Physics of speckle tracking
    b. Longitudinal, circumferential and radial strain
    c. Rotation, twist and torsion
    d. Global longitudinal strain and global circumferential strain vs segmental
    e. Clinical utility of STE

E. Provocative Maneuvers
1. Physical maneuver
    a. Valsalva maneuver
    b. Mueller
    c. Isometric handgrip
    d. Straight leg raising
    e. Squatting
    f. Inspiration
    g. Expiration
    h. Tilt table
2. Pharmacologic
3. Clinical application of each

F. Recognition of Technical Errors
1. Artifacts
    a. Reverberation
    b. Side lobes
    c. Near field clutter
    d. Echo drop out or shadowing
    e. Mirror image
    f. Translational motion
    g. Aliasing
2. Instrumentation errors
    a. Overgain or undergain
    b. Improper focal location
    c. Improper processing curves
    d. Improper set-up or software package
    e. Inappropriate PRF/scale (Doppler)
    f. Inappropriate PW gate size or position
    g. Inappropriate sweep speed (M-mode/spectral Doppler)
    h. Improper ECG lead placement

Section IV: Principles of Cardiac Hemodynamics and Cardiac Cycle

1. List the factors that affect blood flow
2. Describe the relationship between pressure and velocity as it relates to the Bernoulli equation
3. Describe clinical applications and pitfalls of common cardiac Doppler hemodynamics
4. List normal pressures of cardiac chambers and great vessels
5. Relate phases of the cardiac cycle to electrocardiographic events
6. Describe spectral Doppler physiology as it relates to valvular and pulmonary vein flow

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IV. Principles of Cardiac Hemodynamics

A. Fluid Dynamics
   1. General description
      a. Flow and related terms
      b. Power, work and energy
      c. Potential and kinetic energy
      d. Hydrostatic pressure
      e. Volumetric flow
      f. Velocity
      g. Capacitance
      h. Compliance
      i. Fluid viscosity

B. Derivations of Equations
   1. General description
      a. Resistance equation
      b. Volumetric flow equation (continuity equation)
         i) Conservation of mass - flow proximal to valve must equal flow across the valve
      c. Pressure flow relationships
         i) Poiseuille's law
         ii) Bernoulli principle
            - Conservation of energy
            - Bernoulli equation
               - Assumption
                  - Flow acceleration + viscous friction are negligible
                  - No energy transfer
               - Modified Bernoulli equation
               - Simplified Bernoulli equation
               - Clinical use
                  - Valvular stenosis (aortic, pulmonic)
                  - Pulmonary artery pressure
         iii) Reynolds number

- Point at which turbulence occurs

C. Physiology and Hemodynamic Information Derived from Doppler

1. Pressure and flow resistance
    a. Stroke volume (SV)
2. Cardiac output (CO)
3. Cardiac index (CI)
    a. Measured in liters/minute/meter2
4. Delta pressure (dp) over deltatime (dt) mmHg/msec
    a. Time in milliseconds (msec) it takes the LV to generate 32 mmHg; first derivative of LV pressure rise
5. Continuity equation - based on conservation of mass
    a. Aortic valve area (AVA)
        i) Pitfalls
    b. Mitral valve area (MVA)
        i) Mitral Valve Area
        ii) Pitfalls
6. PISA
    a. Mitral regurgitation
        i) Regurgitant flow rate
        ii) Effective regurgitant orifice area (EROA)
        iii) Regurgitant volume (RV)
        iv) Pitfalls
    b. Mitral stenosis
        i) Mitral valve flow rate
            - Flowrate in cc/sec
        ii) Mitral valve area (MVA cm2)
        iii) Pitfalls
7. Pressure half time
    a. Time it takes for mitral valve gradient to fall by half its initial value
    b. Mitral valve area (MVA)
        i) MVA cm2 ---- 220/halftime (msec)
        ii) Halftime = deceleration time x 0.29
    c. Aortic insufficiency (AI)
    d. Pitfalls
8. Regurgitant fraction (RF)
    a. RF aortic valve (AV)
    b. RF mitral valve
    c. Pitfalls

9. Qp:Qs ratio
    a. Used to evaluate shunt flow
    b. Pitfalls

D. Normal Pressures
1. Chamber and great vessel pressure
    a. LA
    b. RA
    c. RV
    d. LV
    e. PA
    f. Aorta
    g. Arterioles
    h. Capillaries

E. Cardiac Cycle
1. Electrical characteristics
    a. Conduction system
        i) Sinoatrial (SA) node
        ii) Intra-atrial tracts
        iii) Atrioventricular (AV) node
        iv) Bundle of His
        v) Right and left bundle branches (Purkinje fibers)
    b. Electrocardiogram
        i) P wave
        ii) QRS complex
        iii) ST segment
        iv) T wave
2. Mechanical characteristics
    a. Systole
        i) Pressure changes within chambers
        ii) Valve opening and closing
    b. Diastole
        i) Pressure changes within chambers
        ii) Valve opening and closing
3. Ventricular diastole
    a. Isovolumic relaxation time (IVRT)
        i) LV pressure falls rapidly
        ii) Follows aortic valve closure
        iii) IVRT ends when LV pressure falls below LA pressure and MV opens
    b. Passive filling/early filling
        i) Rapid filling

ii) Pressure equalization
iii) Factors that increase passive filling
- Mitral/tricuspid regurgitation
- Atrial/ventricular septal defect

iv) Factors that decrease passive filling
- Increased heart rate
- Mitral stenosis
- Increased ventricular end-diastolic pressure

c. Diastasis
d. Atrial systole (atrial contraction)
i) Atrial pressure exceeds ventricular pressure
ii) MV opens and late LV filling occurs

4. Ventricular systole
a. Isovolumic relaxation time
b. Isovolumic contraction time (IVCT)
c. Rapid ejection
d. Reduced ejection
e. Factors that influence ventricular systole
i) Ventricular function
ii) Afterload
iii) Preload
iv) Frank-Starling's law

F. Pulmonary Veins
1. Vein characteristics
2. Anatomic location
3. Doppler interrogation
4. Waveform characteristics
a. Systole
b. Diastole

G. Cardiac Catheterization
1. Clinical utility
a. Intracardiac pressure
b. Oxygen saturation
c. Coronary artery assessment
d. Other
2. Normal pressure tracings
a. Right heart
b. Left heart

Section V: Ventricular Function

1. Define ventricular function terminology
2. Identify the etiologies, signs and symptoms, and risk factors of ischemic heart disease
3. Classify the types of wall motion abnormalities
4. Describe 2-D, M-mode and Doppler features associated with ischemic heart disease
5. Identify formulas associated with assessment of ventricular function
6. Differentiate ways to assess global LV function and regional LV quantification

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V. Ventricular Function

A. Determinants of Left Ventricular Performance
   1. Contractility
      a. Inotropic state of myocardium
      b. Best determinants
      c. Limitations
   2. Heart rate
   3. Preload
      a. Define
      b. Frank-Starling relation
         i) Left ventricular end diastolic volume (LVEDV) versus left ventricular end diastolic pressure (LVEDP)
         ii) SV increases as end-diastolic volume increases
   4. Afterload
      a. Define
      b. Determinants
         i) Ventricular volume and pressure
         ii) Atrial resistance
         iii) Aortic impedance
         iv) Mass of blood in aorta
         v) Viscosity of blood
      c. LaPlace's Equation
         i) Wall stress = Pressure * LV radius/wall thickness
         ii) Inverse relation to LV systolic function
   5. Coordinated LV contraction

B. Wall Motion Abnormalities
   1. Hypokinesis
   2. Akinesis
   3. Dyskinesis
   4. Hyperkinesis
   5. Wall motion scoring index

C. Global LV Systolic Performance

1. Ejection phase indices
    a. Ejection fraction (EF)
    b. Fractional shortening (FS)
    c. Velocity of circumferential fiber shortening (Vcf)
    d. CO and SV
    e. Tissue Doppler imaging (Sm)
    f. Speckle tracking echocardiography (STE)
2. Non-ejection phase indices
    a. Systolic time intervals
        i) Pre-ejection time
    b. LV dP/dt
    c. Acceleration time of aortic flow
    d. Pressure/volume analysis
3. Indirect methods
    a. Mitral E-point septal separation (EPSS)
    b. Aortic root motion
    c. Descent of cardiac base
    d. Sphericity Index of LV
    e. Velocity time integrals (VTI) of outflow
        i) LVOT
        ii) RVOT
4. Potential limitations
    a. Endocardial dropout
    b. Influence of load conditions
    c. Regional wall motion abnormalities
    d. Some methods require no major shape distortion
    e. Oblique measurements/angles
    f. Limited frame rate of 2-D imaging
    g. LV foreshortening
    h. Translation artifact
    i. Dyssynchrony of contraction
5. Improve endocardial delineation
    a. Harmonic imaging
    b. Contrast agents (enhanced cardiac ultrasound)

D. Assessment of Ventricular Systolic Function
1. Fractional shortening
    a. Technique
        i) M-mode
        ii) 2-D
    b. Calculations

c. Normal values
d. Limitations
    i) Less accurate in the presence of wall motion abnormalities
    ii) Load dependent

2. Stroke volume
3. Cardiac output and index
4. Ejection fraction
    a. Characteristics
        i) Ratio of SV to DV
        ii) Most commonly used clinical index of LV function
        iii) Influenced by preload, afterload and heart rate
        iv) Visual estimated valid with interobserver variability
    b. Calculations
    c. Normal values
    d. Limitations
        i) Due to altered loading conditions
            - MR: %EF normal with intrinsic myocardial dysfunction
            - AS: %EF decreased with normal myocardial function
            - Severe anemia
            - Hemodialysis patients
5. Left ventricular mass
    a. M-mode method
        i) Penn formula
        ii) ASE formula
        iii) Normal values
    b. 2-D method
        i) Area-length
        ii) Truncated ellipsoid
        iii) Normal values
        iv) Limitations
6. Change in pressure by change in time (dP/dt)
    a. Characteristics
        i) Relatively load-independent
    b. Calculations
    c. Appropriate measurement placement
    d. Normal values
7. LV volumes
    a. Simpson's rule and modified Simpson's rule
        i) Biplane more accurate
        ii) Single plane

- Suitable if LV symmetric

iii) Quick method (regression equation)

b. End systolic volumes (ESV)

i) Most reproducible volume measure

ii) Insensitive to cardiac loading

iii) Powerful predictor of cardiac events

iv) Normal values

c. End diastolic volumes (EDV)

i) Endocardium more difficult to image at end diastole

ii) More variable than ESV

iii) Normal values

d. Technical considerations

i) Image optimization

ii) Both atrioventricular valves imaged

iii) Avoid aorta and coronary sinus

iv) Selection of precise time in cardiac cycle for measurement

v) ED frame with largest LV cavity just after MV closure

vi) ES frame with smallest LV cavity just before MV opening

vii) Enhanced cardiac imaging

8. Tissue Doppler imaging

a. Characteristics

b. Appropriate technique

E. LV Quantification

1. Prognosis

a. CAD

b. Acute MI

c. Cardiomyopathy

i) Volume/mass ration

2. Methods for regional function

a. 17 segment wall score

b. Tissue Spectral Doppler imaging

i) Strain

3. Considerations

a. Hypertension

i) LV mass predicts morbidity/mortality

ii) LV wall thickness/cavity dimension predicts morbidity/mortality

b. Interdependence of LV and RV

c. Global RV systolic function

4. Associated conditions

a. LVH

b. Ischemic heart disease
c. DCM
d. HCM
e. RCM
f. RV pressure and volume overloads
g. Valvular heart disease
h. Pericardial disease
i. Transplant rejection
j. Takotsubo Cardiomyopathy
k. Chemotherapy cardiotoxicity

5. Therapeutic implications
   a. Angiotensin-converting enzyme inhibitors
   b. Timing of surgery in volume overload states (MR, AI)
   c. Timing of surgery in pressure overload states (AS)

F. Assessment of Ventricular Diastolic Function
1. General principles
   a. Four phases of diastole
      i) Isovolumic relaxation
      ii) Early rapid diastolic filling
      iii) Diastasis
      iv) Late diastolic filling caused by atrial contraction
   b. Parameters of diastolic function
      i) Relaxation
         - IVRT
         - Maximum rate of pressure decline (dP/dt)
         - Time constant of relaxation (tau)
      ii) Compliance
         - dP/dV
         - Chamber stiffness constant
      iii) Ventricular diastolic pressures
      iv) Ventricular diastolic filling curves
2. Echocardiography evaluation
   a. 2-D imaging
      i) Chamber dimensions
      ii) Wall thickness
      iii) Automated endocardial border tracing
   b. Doppler imaging
      i) Assessment of MV inflow & normal values
         - E-wave/A-wave ratio
         - Deceleration time (DT)

- IVRT
- Mitral E-wave velocity (E)
- Mitral A-wave velocity (A)
- Mitral A-wave duration
- Time from mitral valve opening to E-wave velocity

ii) Assessment of pulmonary vein flow patterns

- Systolic wave velocity (S)/velocity time integral (VTI)
- Diastolic (D) wave velocity/VTI
- Systolic to diastolic ratio (S/D)
- Atrial reversal wave (AR) velocity
- AR duration
- AR duration minus Mitral A-wave duration

iii) Assessment using tissue Doppler

- Peak early diastolic myocardial velocity (e')
- E to e' ratio

iv) Assessment using color M-mode

- Velocity of propagation of early filling (Vp)

3. LV filling patterns
   a. Technical factors
      i) Sample volume location
      ii) Doppler modality (PW versus CW)
      iii) Intercept angle
   b. Physiologic factors
      i) Respiration
      ii) Heart rate
      iii) PR interval
      iv) Age
      v) Preload
      vi) Afterload
      vii) Exercise
      viii) Valsalva
      ix) LV systolic function
      x) Atrial contraction function
      xi) Diastolic function
      xii) Cardiac function
   c. Normal Doppler values
      i) AR
      ii) AR duration – A duration
      iii) DT
      iv) E/A

v) e’
vi) IVRT
vii) Vp using color M-Mode

d. Abnormal relaxation
   i) Doppler values
      - AR
      - AR duration < Mitral A duration
      - DT
      - E/A
      - e’
      - IVRT
      - Vp
   ii) Significance
      - Initial stage
      - No increase in mean LA pressure
      - Difficult to assess with elevated heart rate
      - Seen in
        - Systemic hypertension (HTN)
        - Coronary artery disease (CAD)
        - Cardiomyopathies
        - Elderly

e. Pseudonormalization
   i) Doppler values
      - AR
      - AR duration – Mitral A duration
      - DT
      - E/A
      - e’
      - IVRT
      - S/D
      - Vp
   ii) Significance
      - Spectral Doppler of mitral inflow appears normal
      - Underlying relaxation abnormality with elevated LA pressure
      - Preload reduction may unmask relaxation abnormality
        - Valsalva
        - Nitroglycerin
      - Seen in:
        - Dilated cardiomyopathy (DCM)

- Restrictive cardiomyopathy (RCM)
- Ischemic cardiomyopathy

f. Restrictive/Noncompliance

i) Doppler values

- AR
- AR duration – Mitral A duration
- DT
- E/A
- e'
- IVRT
- S/D
- Vp

ii) Significance

- Late stage
- Severe decrease of LV compliance
- Elevated mean LA pressure at rest
- May reverse with preload reduction maneuvers

g. Estimation of LV filling pressures

i) Normal LV filling pressures

- Abnormal relaxation pattern except with severe LVH

ii) Elevated LV filling pressures

4. RV Quantification

a. Dimensions and volumes

i) Basal, mid and base-apex dimensions

ii) Systolic and Diastolic Volumes

iii) RV wall thickness

b. Systolic function

i) Tricuspid Annular Plane Systolic Excursion (TAPSE)

ii) Tricuspid Annular s' velocity

iii) Fractional Area Change

c. Other quantitative assessment

i) Pulmonary vascular resistance (PVR)

ii) RV Index of Myocardial Performance (RIMP)

d. RV diastolic function

i) Tricuspid inflow E/A

ii) e', a', and s' velocity

iii) E/e' ratio

Section VI: Coronary Artery Disease (CAD)

1. Correlate ventricular wall segments to coronary artery distribution
2. Differentiate complications associated with myocardial infarction

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VI. Coronary Artery Disease (CAD)

A. Coronary Artery Anatomy and Function
   1. Coronary arteries
   2. Flow distribution
   3. Artery location
   4. Relation between coronary arteries and myocardial wall segments
   5. Flow measurements and limitations
   6. Coronary flow reserve
   7. Myocardial perfusion with contrast enhanced echo
   8. Anatomy
   9. Normal imaging in PSAX
   10. Left main at left posterior sinus
   11. 4 o'clock position
   12. RCA at right aortic sinus
   13. 11 o'clock position SAX aortic valve level
   14. Normal imaging in PLAX
   15. RVOT view

B. Coronary Artery Abnormalities
   1. Anomalous origins and course
   2. Aneurysms
      a. Kawasaki disease
      b. Atherosclerotic aneurysms
      c. Polyarteritis
      d. Syphilis
      e. Infection
      f. Trauma
      g. Other
   3. Fistulas
      a. Incidence in general population
      b. Emptying pathway
         i) Cardiac chambers
         ii) PA
         iii) Coronary sinus
      c. Artery affected
         i) RCA 50%

ii) LCA 45%
iii) Both 5%
d. Continuous murmur
e. Echo feature
i) Dilated coronary artery
ii) Chamber enlargement
iii) Color Doppler may detect shunt flow

C. Myocardial Ischemia
1. Coronary artherosclerosis
2. Sequence of events in ischemia (ischemic cascade)
3. Relation of wall motion and thickness to artery perfusion
4. Echocardiographic findings
a. Regional wall motion abnormalities
i) Confounding factors
- Conduction or pacing abnormalities
- Translocation motion
- Loading conditions
- Altered imaging planes

ii) Diastolic function changes
iii) Stress echocardiography

D. Myocardial Infarction (MI)
1. Detection and location of MI
a. Regional wall motion abnormality
i) Wall motion score index
ii) Relationship of wall motion abnormalities to coronary artery anatomy
b. Complications and associated findings
i) Myocardial stunning and hibernation
ii) Myocardial rupture
iii) Acute ventricular septal rupture
iv) Papillary muscle dysfunction/rupture
v) Mural thrombi
- MR
- Dilated LV

vi) Papillary muscle dysfunction
vii) Aneurysm
viii) Pseudoaneurysm
ix) Post MI pericardial effusion (Dressler's syndrome)
x) Ischemic cardiomyopathy
xi) RV infarction

E. LV Post Infarction/Follow-Up

1. Characteristics
2. Echocardiographic features
    a. Recovery of function
    b. Effect of revascularization
    c. LV thrombus
3. Myocardial remodeling
    a. Infarct expansion
    b. Global dilatation

Section VII: Valvular Heart Disease

1. Describe common etiologies of valvular heart disease
2. Discuss signs and symptoms associated with valvular heart disease
3. Describe key echocardiographic findings associated with valvular heart disease
4. Explain the methods for estimation of right atrial and right ventricular systolic pressure
5. Describe typical echocardiography views utilized in assessing valvular stenosis and regurgitation
6. List the parameters used in qualitative and quantitative assessment of valvular heart disease
7. Discuss the challenges of stenotic valve assessment in the presence of reduced LV function or significant regurgitation
8. Describe common locations of vegetation formation
9. Identify the signs and symptoms of infective endocarditis
10. List examples of mechanical and bioprosthetic valves
11. List common complications of prosthetic valves
12. Differentiate between advantages of using mechanical versus bioprosthetic valves
13. Describe Doppler calculations used to assess prosthetic valves

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VII. Valvular Heart Disease

A. Mitral Stenosis (MS)

1. Etiology
   a. Rheumatic
   b. Mitral annular calcification (MAC) and calcific MS
   c. Congenital (parachute)
   d. Drug-induced valvulopathy
   e. Other
2. Pathophysiology
   a. Narrowing of MV orifice with obstruction to diastolic forward flow
   b. MV gradient and LA pressure increase to maintain cardiac output
   c. Increased LA pressure eventually leads to LA enlargement, “backwards” failure and pulmonary hypertension
3. Clinical presentation
   a. Signs and symptoms
      i) Dyspnea
      ii) Fatigue
      iii) Chest pain
      iv) Hemoptysis
      v) Palpitations
      vi) Ortner’s syndrome (hoarseness)
   b. Physical findings
      i) Atrial fibrillation
      ii) Edema
      iii) Hepatic congestion
      iv) Pulmonary edema
      v) RV lift

vi) Apical diastolic thrill

c. Auscultatory findings

i) Loud S1

ii) Opening snap

iii) Low-pitched diastolic rumble

iv) Holosystolic murmur (with MR)

v) Accentuated P2 (pulmonary hypertension)

4. Echocardiographic features

a. Thickened and calcified mitral leaflets and subvalvular apparatus

b. Doming of anterior mitral leaflet during diastole ("hockey-stick" appearance)

c. Immobility of posterior leaflet

d. "Fish-mouth" orifice in short-axis view

e. Commissural fusion

f. Decreased mitral opening

g. LA enlargement

h. LV small with normal function (pure MS)

i. PHTN

i) Secondary RA and RV enlargement

ii) Elevated TR velocity

j. Candle-flame appearance of LV inflow tract (Color Doppler)

k. Coexisting valvular lesions

l. Spontaneous contrast or thrombus in LA, LAA

5. Quantitation

a. Pressure gradients (CW Doppler)

i) Peak (initial gradient)

ii) Mean

b. MV area

i) 2-D planimetry (short-axis view)

ii) PHT method

iii) Continuity method

iv) PISA

c. Values

d. Degree of pulmonary hypertension (TR velocity)

e. Technical considerations and pitfalls

i) Should not use PHT method immediately after balloon valvuloplasty, with severe AR, or with decreased LV compliance

ii) Should not use continuity method if significant AR or MR

iii) Angle correction may be required with PISA method

iv) Mean gradient dependent on heart rate and cardiac output

6. Role of hemodynamic stress testing

a. Useful in patients with symptoms out of proportion to resting hemodynamics or those with sedentary lifestyle
b. Substantial rise in mean gradient and PA pressure with exercise indicates hemodynamically significant MS

7. Role of TEE
   a. Assessment for spontaneous contrast, LA, LAA thrombus
   b. Evaluation of morphology and hemodynamics with suboptimal surface study
8. Role of echocardiography in percutaneous balloon mitral valvuloplasty
   a. Patient selection
      i) MV score index
      ii) Severity of MR
      iii) Absence of LA, LAA thrombus
      iv) Minimal or no commissural calcium
   b. Echo guidance
      i) Transseptal puncture
      ii) Balloon positioning
      iii) Assessment of results
      iv) Detection of complications
   c. Complications
      i) MR
      ii) Cardiac perforation and tamponade
      iii) ASD
9. Related testing
   a. ECG
      i) Atrial fibrillation
      ii) LA enlargement
      iii) RVH and right axis deviation (with pulmonary hypertension)
   b. Chest x-ray
      i) LA enlargement with normal LV size
      ii) Pulmonary venous hypertension
      iii) Right heart enlargement
      iv) Pulmonary edema
   c. Cardiac catheterization
      i) Angiography to identify underlying CAD
      ii) Increased pressure gradient across MV
      iii) Balloon valvuloplasty may be performed
10. Common treatment
   a. Medical
      i) Antibiotic prophylaxis
      ii) Treatment for heart failure
      iii) Anticoagulation if clinically indicated

b. Surgical
    i) Balloon valvuloplasty
    ii) Open or closed commissurotomy
    iii) MV replacement

B. Mitral Regurgitation (MR)
1. Etiology
    a. MV prolapse (MVP)
    b. Flail mitral leaflet
    c. Ruptured chordae
    d. Mitral annular dilatation or calcification
    e. Papillary muscle dysfunction or rupture
    f. Rheumatic or myxomatous valve
    g. Drug-induced valvulopathy
    h. Congenital malformation
    i. Endocarditis or perforation
    j. Regional or global LV remodeling
2. Pathophysiology
    a. Backward flow of blood through an incompetent mitral valve during ventricular systole
    b. Chronic mitral regurgitation
        i) Progressive increase in degree of MR leading to LV volume overload
        ii) Impaired forward cardiac output due to volume of regurgitant flow
        iii) Heart compensates for volume overload with LV dilatation and hypertrophy
        iv) Increased LA pressure causes LA dilatation
        v) Increased LVEDP and LA pressure eventually lead to “backwards” failure and pulmonary hypertension
    c. Acute mitral regurgitation
        i) Rapid development of significant MR
        ii) LV unable to compensate for increased volume
        iii) Marked increase in LA pressure in non-compliant left atrium
        iv) Increased LA pressure leads to increased pulmonary capillary pressure and pulmonary edema
3. Clinical presentation (chronic and acute MR)
    a. Signs and symptoms
        i) Progressive dyspnea
        ii) Fatigue and exhaustion
        iii) Palpitations
        iv) Atypical chest pain
    b. Physical findings
        i) Atrial fibrillation (chronic)
        ii) Elevated JVP
        iii) Hepatomegaly

iv) Peripheral edema

v) Displaced, active LV impulse

vi) Pulmonary edema (acute)

c. Auscultatory findings

i) Holosystolic murmur (chronic)

ii) Early systolic murmur (acute)

iii) S3, S4 with heart failure

iv) Accentuated P2 with pulmonary hypertension

4. Echocardiographic features

a. Functional anatomy

i) Mitral annulus (dilated, calcified)

ii) Mitral leaflets (thickened, flail, prolapse)

iii) Chordae tendineae (elongated, ruptured)

iv) Papillary muscle (displaced, ruptured)

v) Ventricular myocardium (remodeled)

b. LV dilatation

c. LA enlargement

d. Color flow jet of MR

e. Evidence of pulmonary hypertension

i) Elevated TR velocity

ii) Dilated IVC/hepatic veins

5. Qualitative and semi-quantitative assessment

a. Color flow imaging

i) Jet size and configuration

- Jet area to LA area ratio
- May be deceptive with acute MR: maximal jet area is limited by small receiving chambers and rapid equalization of pressures
- Central jets may appear larger because blood cells are entrained on all sides of the jet
- Interpret in context of jet geometry and surrounding solid boundaries

ii) Direction and eccentricity of jet

iii) Color M-mode (to establish timing)

iv) Vena contracta

- Narrowest portion of jet that occurs at or just downstream from the orifice
- Obtain in high resolution zoomed view
- Image in multiple planes perpendicular to the commissural line (usually parasternal or apical long-axis views)
- Measure width of the “neck” of the jet at the time of its largest diameter
- Not valid in the presence of multiple jets

b. Pulsed-wave Doppler

i) Mitral inflow E wave velocity

- Predominant early filling with severe MR
- Elevated “E” velocity
- Must rule out co-existing stenosis

ii) Pulmonary vein flow abnormalities

- Usually see predominant systolic forward flow
- Blunting of systolic forward flow occurs with increasing severity of MR
- High LA pressure and atrial fibrillation may also cause diminished systolic forward flow
- Severe MR may cause systolic flow reversal
- Eccentric jet may selectively enter one pulmonary vein

c. Continuous-wave Doppler

i) Density of CW Doppler signal proportional to regurgitant volume

ii) Peak MR velocity dependent on pressure gradient between LV and LA

- Decreased velocity indicates elevated LA pressure, but does not in itself indicate severity of MR

iii) Duration of MR signal is typically holosystolic

iv) Shape of MR Doppler signal

- Symmetrical shape: reflects mild MR
- Asymmetrical shape: reflects severe MR (due to rapid increase in LA pressure)

v) Assess RVSP: indirect clue to severity of MR and compensation to volume overload

6. Quantitative parameters

a. Methods

i) Continuity

ii) PISA

b. Parameters

i) Regurgitant volume

ii) Regurgitant fraction

iii) Regurgitant orifice area

c. Values

d. Pitfalls

i) Learning curve of operator

ii) Data acquisition errors

iii) Continuity method invalid if mild or greater aortic insufficiency or significant shunts are present

iv) Difficult to calculate accurate regurgitant volume by continuity method in the presence of valvular stenosis, dense annular calcification or prosthetic valve

7. Related testing

a. ECG

i) LA enlargement

ii) LV enlargement

iii) Left axis deviation

iv) Atrial fibrillation

v) RVH with pulmonary hypertension

b. Chest x-ray

i) Cardiomegaly (chronic)

ii) Pulmonary congestion

iii) PA and right heart enlargement (late)

iv) Pulmonary edema

c. Cardiac catheterization

i) Angiography to identify underlying CAD

ii) Increased right heart pressures

d. Role of TEE

i) Evaluation of morphology, assessment of hemodynamics and quantitation with suboptimal surface study

ii) Intra-operative evaluation

8. Common treatment

a. Medical

i) Antibiotic prophylaxis

ii) Treatment for heart failure

iii) Anticoagulation if clinically indicated

b. Surgical MV Repair

i) Role of echo in timing of surgery

ii) MV repair

- Resection
- Chordal transfer, reattachment
- Artificial chordate
- MV annuloplasty ring
- MitraClip percutaneous repair
- Alfieri stitch

iii) MV replacement (MVR)

C. Mitral Valve Prolapse (MVP)

1. Etiology

a. Myomatous disease

b. Marfan syndrome

c. Flail leaflet

2. Clinical presentation

a. Signs and symptoms as for MR

b. Physical presentation

i) Most patients asymptomatic

ii) Palpitations

iii) Chest pain

iv) Dyspnea

v) Fatigue

c. Auscultatory findings

i) Mid-systolic click

ii) Mid-to-late systolic murmur if MR present

iii) Click and murmur occur earlier in systole with preload reduction (Valsalva, standing)

iv) Click and murmur occur later in systole with increased ventricular volume (squatting)

3. Echocardiographic features

a. Diagnosis based on parasternal long-axis view

b. Systolic displacement of one or both mitral leaflets into the LA, below the plane of the mitral annulus

c. Leaflets may be thickened or myxomatous

d. Elongated chordae

e. LA and/or LV enlargement with significant MR

f. Mitral regurgitation, typically eccentric and late-systolic

g. Progression of MR severity over time

4. Related testing

a. ECG

i) Normal in asymptomatic patients

b. Chest x-ray

i) Normal in asymptomatic patients

c. Cardiac catheterization

i) Rarely performed for diagnostic purposes

5. Common treatment

a. None unless significant MR is present

b. Endocarditis prophylaxis

i) Serial echo as indicated

D. Aortic Stenosis (AS)

1. Etiology

a. Degenerative calcification

b. Congenital aortic valve disease

c. Post inflammatory disease

d. Rheumatic

e. Irradiation

f. Endocarditis

2. Pathophysiology

a. Narrowing of AV orifice with obstruction to systolic forward flow

b. Decreased AV orifice causes increased gradient

c. Heart responds to increased afterload with increased contractile function and LVH

d. Elevated LVEDP eventually leads to LA enlargement, “backwards” failure and pulmonary hypertension

3. Clinical presentation
    a. Signs and symptoms
        i) Angina
        ii) Dyspnea, exertional dyspnea
        iii) Paroxysmal nocturnal dyspnea
        iv) Congestive heart failure
        v) Syncope, presyncope
    b. Physical findings
        i) Narrow pulse pressure
        ii) Slow rising, late peaking carotid pulse (parvus et tardus)
    c. Auscultatory findings
        i) Harsh systolic ejection murmur (crescendo-decrescendo) that may radiate to carotids
        ii) Diastolic murmur if AR present
        iii) S3, S4 with heart failure
4. Echocardiographic features
    a. Aortic valve 2-D appearance
        i) Cusp number
        ii) Presence/location of raphe
        iii) Systolic “doming” of AV if bicuspid
        iv) Eccentric closure
        v) Thickened/calcified leaflets
        vi) Restricted mobility
    b. Aorta
        i) Post-stenotic dilatation
        ii) Aortoseptal angle
        iii) Possible coarctation with BAV
    c. LV
        i) LVH
        ii) LVOT obstruction
        iii) Normal or small LV size
        iv) Hyperdynamic LV function (until late)
    d. LA enlargement
    e. Color Flow
        i) Turbulent forward flow
        ii) Associated aortic regurgitation
5. Quantitation
    a. Pressure gradients (CW Doppler)
        i) Peak instantaneous
        ii) Mean
        iii) Utilize multiple imaging windows to obtain Doppler signal most parallel to flow

- Apical
- Subcostal
- Right parasternal
- Right supraclavicular
- Suprasternal
- Left parasternal

iv) Use nonimaging CW Doppler probe

- Smaller footprint allows optimal positioning and angulation
- Non-imaging transducer has higher signal-to-noise ratio than duplex transducer

v) Assessment unreliable in the presence of subaortic obstruction

- Bernoulli invalid with stenosis in series
- Cannot measure LVOT velocity

vi) Alternative method for peak gradient:

- MR velocity
- LVSP – LVOT gradient – Systolic BP = AV gradient

b. AV area

i) 2-D planimetry

ii) Continuity method

c. Values

d. Dimensionless index

i) LVOT and AV velocity or VTI ratio

ii) May be used if unable to measure LVOT diameter

6. Low output-low gradient AS

a. Small calculated valve area with limited excursion of leaflets, but mean gradient is low in the setting of LV systolic dysfunction

b. Two possible explanations

i) True anatomically severe aortic stenosis

ii) Functionally severe aortic stenosis

c. Low-dose pharmacologic stress echo

i) Stroke volume increases with increase in contractility

ii) Doppler data obtained at each infusion stage

iii) True stenosis: No change or minimal increase in AVA

iv) Functional stenosis: Valve area increases

7. Related testing

a. ECG

i) LVH

ii) LA enlargement

b. Chest x-ray

i) LVH

ii) LA enlargement

iii) Post-stenotic dilatation of ascending aorta
iv) AV calcification

c. Cardiac catheterization
i) Determines peak-to-peak gradient (different from echo peak instantaneous)
ii) Determines mean gradient
iii) Angiography to identify underlying CAD
iv) Increased right heart pressures

d. Role of TEE
i) Evaluation of morphology with suboptimal surface study
ii) Preferred method for planimetry of AV area
iii) Difficult to obtain accurate Doppler data

8. Common treatment
a. Medical
i) Antibiotic prophylaxis
ii) Treatment for heart failure
iii) Anticoagulation if clinically indicated

b. Surgical
i) Balloon valvuloplasty (in children)
ii) AVR

E. Aortic Regurgitation (AR)
1. Etiology
a. Aortic root abnormalities
i) Aortic root dilatation
ii) Aortic dissection

b. Aortic leaflet abnormalities
i) Congenital (bicuspid)
ii) Degenerative valve disease
iii) Rheumatic valve disease
iv) Endocarditis
v) Drug-induced valvulopathy
vi) Miscellaneous other

2. Pathophysiology
a. Backward flow of blood through an incompetent AV during ventricular diastole
b. Chronic
i) Progressive increase in AR leading to LV volume overload
ii) Heart compensates for volume overload with LV dilatation and hypertrophy
iii) Forward stroke volume increased by Frank-Starling mechanism
iv) Effective forward cardiac output maintained with normal function
v) LVEDP begins to rise as ejection fraction and cardiac output decrease
vi) Increased LVEDP and LA pressure eventually leads to “backwards” failure and pulmonary hypertension

c. Acute

i) Rapid onset of AR leads to volume overload

ii) LV unable to compensate for increased regurgitant volume

iii) Leads to elevated left ventricular end-diastolic pressure (LVEDP) and acute pulmonary edema

3. Clinical presentation

a. Signs and symptoms

i) Patients often asymptomatic until AR becomes significant

ii) Exertional dyspnea

iii) Angina

iv) Fatigue

v) Palpitations, tachycardia

vi) Lightheadedness, syncope

b. Physical findings

i) Apical impulse displaced due to LV enlargement (chronic)

ii) Wide pulse pressure (low diastolic BP)

iii) Tachycardia (acute)

iv) Pulmonary edema (acute)

c. Auscultatory findings

i) Early, blowing, diastolic decrescendo murmur

ii) Austin-Flint murmur

iii) Secondary systolic ejection murmur

iv) S3, S4 with heart failure

4. Echocardiographic features

a. Functional anatomy

i) Aortic root abnormalities

- Aortic root dilatation
- Annulo-aortic ectasia
- Sinus of Valsalva aneurysm (ruptured or unruptured)
- Aortic dissection

ii) Aortic leaflet abnormalities

- Bicuspid aortic valve
- Endocarditis
- Post-inflammatory disease
- Calcific disease
- Post valve surgery

b. LV dilatation (chronic)

c. Reduced LV systolic function (late)

d. Normal LV size with hyperdynamic systolic function (acute)

e. LA dilatation (chronic)

f. Associated findings
   i) Diastolic flutter of anterior mitral valve leaflet
   ii) Jet lesion on septum, anterior mitral valve leaflet
   iii) Premature closure of mitral valve with diastolic MR (acute)
   iv) Premature opening of aortic valve (acute)

g. Color flow jet of AR

h. Evidence of pulmonary hypertension
   i) Elevated TR velocity
   ii) Dilated IVC/hepatic veins

5. Qualitative and semi-quantitative assessment

a. Color flow imaging
   i) Direction of flow (central jet, eccentric jet)
   ii) Color and color M-mode helpful in demonstrating timing and thickness of jet and diastolic reversal in descending thoracic aorta
   iii) Regurgitant jet width to LVOT width ratio
      - Parasternal long-axis view
   iv) Regurgitant jet area to LVOT area ratio
      - Parasternal short axis view immediately below the aortic valve (within 1 cm of the valve)
   v) Vena contracta
      - Narrowest portion of jet that occurs at or just downstream from the orifice, measured at the time of its largest diameter
      - Image in multiple planes perpendicular to the commissural line (usually parasternal long-axis view)
      - Not valid in presence of multiple jets

b. Pulsed-wave Doppler
   i) Mitral inflow
      - Premature cessation of flow across mitral valve
      - Fluttering motion due to turbulence of posteriorly-directed jet
      - Restrictive mitral inflow pattern caused by high LVEDP
   ii) LVOT velocity
      - Increased velocity and VTI due to increased flow volume
   iii) Descending thoracic aorta
      - Mild AR: mild early diastolic flow reversal
      - Duration and velocity of reversal increase with increasing severity of AR
      - Severe AR: holodiastolic flow reversal
   iv) Abdominal aorta diastolic flow reversal

c. Continuous-wave Doppler
   i) Density of CW Doppler signal proportional to regurgitant volume
   ii) Duration of AR signal is holodiastolic
   iii) Pressure half-time of AR signal

iv) Assess RVSP

6. Quantitative parameters
   a. Methods
      i) Continuity
      ii) PISA
   b. Parameters
      i) Regurgitant volume
      ii) Regurgitant fraction
      iii) Regurgitant orifice area
   c. Values
   d. Pitfalls
      i) Continuity method invalid if multivalvular regurgitant lesions or significant shunts present
      ii) Difficult to calculate accurate regurgitant volume with continuity method in the presence of valvular stenosis, dense annular calcification, or prosthetic valve
7. Role of TEE
   a. Evaluation of morphology and hemodynamics with suboptimal surface study
   b. Intra-operative evaluation
8. Related testing
   a. ECG
      i) LVH (chronic)
      ii) Left axis deviation (chronic)
      iii) AV block may be seen in the presence of infective endocarditis
      iv) Sinus tachycardia (acute)
   b. Chest x-ray
      i) Cardiomegaly (chronic AR)
      ii) Widened mediastinum with aortic dissection
   c. Cardiac catheterization
      i) Angiography to identify underlying CAD
      ii) Increased right heart pressures
9. Common treatment
   a. Medical
      i) Antibiotic prophylaxis
      ii) Treatment for heart failure
      iii) Anticoagulation if clinically indicated
   b. Surgical repair
      i) Echo predictors of surgical outcome
      ii) AV repair or replacement
      iii) TAVR

F. Tricuspid Stenosis (TS)
   1. Etiology

a. Rheumatic
b. Congenital
c. Carcinoid
d. Drug-induced valvulopathy
e. Eosinophilic endomyocardial disease
f. Miscellaneous other

2. Pathophysiology
   a. Rarely occurs as an isolated lesion
   b. Narrowing of tricuspid valve orifice with obstruction to diastolic forward flow
   c. Increased RA pressure eventually leads to RA enlargement and signs of right heart failure
3. Clinical presentation
   a. Signs and symptoms
      i) Dyspnea
      ii) Fatigue
      iii) Right upper quadrant abdominal pain
   b. Physical findings
      i) Jugular venous distention with cannon “a” waves
      ii) Hepatomegaly
      iii) Ascites
      iv) Peripheral edema without pulmonary congestion
   c. Auscultatory findings
      i) Tricuspid opening snap
      ii) Diastolic rumble (may be accentuated with inspiration)
      iii) Absence of normal respiratory splitting of S2
      iv) Murmur of TR
4. Echocardiographic features
   a. Valve thickened and/or calcified
   b. Restricted leaflet motion
   c. Diastolic doming of the tricuspid valve
   d. RV size may be increased if concomitant TR
   e. RA dilatation
   f. Inferior vena cava (IVC) enlargement
   g. Increased antegrade velocities with turbulent flow
   h. TR often present
   i. Co-existing valve lesions
   j. Best to obtain Doppler data during apnea (end-expiration)
5. Quantitation
   a. Pressure gradients (CW Doppler)
      i) Peak (initial gradient)
      ii) Mean

b. TV area (rarely used clinically)
   i) Continuity method
   ii) PISA
   iii) Severe stenosis
c. Degree of pulmonary hypertension (TR velocity)
d. Technical considerations and pitfalls
   i) Continuity method invalid if multivalvular regurgitant lesions or significant shunts present
   ii) Angle correction may be required with PISA method
   iii) Mean gradient dependent on heart rate, cardiac output, and respiratory cycle

6. Related testing
   a. ECG
      i) RA enlargement
      ii) Atrial fibrillation
      iii) RVH
   b. Chest x-ray
      i) RA enlargement
      ii) Dilated SVC
   c. Cardiac catheterization
      i) Elevated right heart pressures
      ii) Gradient between RA and RV
7. Common treatment
   a. Medical
      i) Antibiotic prophylaxis
      ii) Treatment for heart failure
      iii) Anticoagulation if clinically indicated
   b. Surgical
      i) Commissurotomy
      ii) TV replacement

G. Tricuspid Regurgitation (TR)
1. Etiology
   a. Functional
   b. Pulmonary hypertension due to left heart failure
   c. Cor pulmonale
   d. Primary pulmonary hypertension
   e. DCM
   f. RV infarction
   g. Anatomic
   h. Rheumatic
   i. Congenital
   j. Carcinoid

k. Drug-induced valvulopathy
l. Endocarditis
m. Injury
n. Miscellaneous other

2. Pathophysiology
   a. Backward flow of blood through incompetent tricuspid valve during ventricular systole
   b. Progressive increase in degree of TR leading to RV volume overload
   c. Impaired forward cardiac output due to volume of regurgitant flow
   d. Heart compensates for volume overload with RV dilatation and hypertrophy
   e. Increased RA pressure causes RA dilatation
   f. Process eventually leads to right heart failure
3. Clinical presentation
   a. Signs and symptoms
      i) Usually asymptomatic for a long period
      ii) Weakness
      iii) Fatigue
      iv) Findings of heart failure
   b. Physical findings
      i) Jugular venous distention with prominent “v” wave
      ii) Hepatomegaly
      iii) Pulsatile liver
      iv) Hepatojugular reflux
      v) Peripheral edema
      vi) Ascites
      vii) Atrial fibrillation
      viii) Hyperdynamic RV impulse
   c. Auscultatory findings
      i) Holosystolic, blowing murmur; may be accentuated with inspiration
      ii) Diastolic flow rumble
      iii) Right-sided S3
      iv) Paradoxic splitting of S2
      v) Loud P2
4. Echocardiographic features
   a. Functional versus anatomic cause
   b. RV volume overload (RVVO)
   c. Increased RV and RA size
   d. Paradoxic septal motion
   e. Dilated IVC and hepatic veins
5. Qualitative and semi-quantitative assessment
   a. Color flow imaging

i) Jet size and configuration
- Jet area to RA area comparison
- May be deceptive in acute regurgitation: maximal jet area is limited by small receiving chambers and rapid equalization of pressures

ii) Direction and eccentricity of jet

iii) Color M-mode (to establish timing)

iv) Vena contracta
- Narrowest portion of jet that occurs at or just downstream from the orifice, measured at the time of its largest diameter
- Not valid in presence of multiple jets

v) PISA
- Validated only in small studies
- Rarely needed clinically

b. Pulsed-wave Doppler

i) Tricuspid inflow E wave velocity
- Predominant early filling with severe TR
- Elevated E velocity
- Must rule out co-existing stenosis

ii) Hepatic vein flow abnormalities
- Diminution of systolic forward flow occurs with increasing severity of TR
- High RA pressure and atrial fibrillation may also cause diminished systolic forward flow
- Severe TR may cause systolic flow reversal

c. CW Doppler

i) Density of CW Doppler signal proportional to regurgitant volume

ii) Velocity of TR is not related to severity of TR

iii) Duration of TR signal typically holosystolic

iv) Shape of TR Doppler signal
- Symmetrical shape: reflects mild TR
- Asymmetrical shape: reflects severe TR (due to rapid increase in RA pressure

v) Use of the Bernoulli equation to assess RVSP may be unreliable if the regurgitant orifice is not “restrictive”

6. Related testing

a. ECG

i) RA enlargement

ii) RBBB

iii) Atrial fibrillation

b. Chest x-ray

i) RA and RV enlargement

ii) Prominent SVC

c. Cardiac catheterization

i) Right heart hemodynamics

7. Common treatment
   a. Medical
      i) Often none (may be well tolerated for years)
      ii) Antibiotic prophylaxis
      iii) Treatment for right heart failure
      iv) Anticoagulation if clinically indicated
   b. Surgical
      i) Annuloplasty
      ii) TVR

H. Pulmonary Stenosis (PS) - See Section Congenital Heart Disease

I. Pulmonary Regurgitation (PR)
   1. Etiology
      a. Pulmonary hypertension
      b. Endocarditis
      c. Congenital abnormalities
      d. Carcinoid
      e. Rheumatic
   2. Pathophysiology
      a. Backward flow of blood through an incompetent pulmonary valve during ventricular diastole
      b. Pathologic regurgitation is infrequent and is typically seen in the presence of significant right heart structural abnormalities
      c. Severe hemodynamic compromise due to isolated severe PR is rare
   3. Clinical presentation
      a. Signs and symptoms
         i) May be well tolerated for years
         ii) Dyspnea
         iii) Fatigue
      b. Physical findings
         i) Palpable RV impulse (right sternal border)
         ii) Jugular venous distention
         iii) Hepatomegaly
         iv) Peripheral edema
         v) Ascites
      c. Auscultatory findings
         i) Low pitched diastolic murmur
         ii) Loud P2 with PHN
         iii) Systolic ejection murmur with severe PR due to increased flow
   4. Echocardiographic features
      a. Anatomic cause: abnormalities
         i) Cusp number

ii) Cusp motion
iii) Valve structure
b. Dilated RV, RA and PA
c. RVVO (paradoxical ventricular septal motion)
d. Diastolic flutter of PV
e. Evidence of PHTN

5. Qualitative and semi-quantitative assessment
a. Color flow imaging
i) Jet size and spatial orientation
ii) Few parameters have been validated
b. PW Doppler
i) RVOT velocity
- Increased velocity due to increased flow volume

ii) Evidence of flow reversal in PA
c. CW Doppler
i) Density of CW Doppler signal proportional to regurgitant volume
ii) Duration of PR signal is holodiastolic
iii) Steep deceleration slope
- Also influenced by RV diastolic compliance and pressure

iv) Assess PAEDP

6. Related testing
a. ECG
i) RVH
ii) Right axis deviation
iii) RBBB
b. Chest x-ray
i) RV enlargement
ii) Enlarged PA
c. Cardiac catheterization
i) Right heart hemodynamics
ii) Pulmonary angiography shows diastolic flow reversal into RV

7. Common treatment
a. Medical
i) Often none (may be well tolerated for years)
ii) Antibiotic prophylaxis
iii) Treatment for right heart failure
iv) Anticoagulation if clinically indicated
b. Surgical
i) Annuloplasty
ii) PV replacement

J. Infective Endocarditis
    1. Etiology
        a. Microbial infection of the endothelial lining of the heart
        b. Most common bacterial organisms
            i) Streptococcus viridans
            ii) Staphylococcus aureus
    2. Pathophysiology/natural history
        a. Occurs primarily on cardiac valves, but may occur on other endocardial surfaces or intracardiac devices
        b. High mortality/complication rate
        c. Higher-risk patients
            i) Prosthetic heart valves
            ii) Native valve disease
            iii) Congenital heart disease
            iv) IV drug users
    3. Clinical presentation
        a. Signs and symptoms
            i) Fever
            ii) Fatigue/weakness
            iii) "Flu-like" symptoms
            iv) Weight loss
        b. Physical findings
            i) Vascular phenomena
                - Petechiae
                - Splinter hemorrhages
                - Osler's nodes
                - Janeway's lesions
            ii) Embolic events (systemic or pulmonary)
            iii) Clubbing
            iv) Splenomegaly
            v) Findings consistent with valve obstruction/regurgitation
        c. Auscultatory findings
            i) New or changing murmur
        d. Diagnostic criteria
            i) Duke – major and minor
    4. 2-D/M-mode features
        a. Echo hallmark: vegetation
        b. Common locations
            i) Atrial side of MV and TV
            ii) Ventricular side of AV and PV
            iii) Secondary jet lesions

c. Less common locations
    i) Chordae
    ii) Mural endocardium
    iii) Eustachian valve
    iv) Pacemaker wire
d. Echogenic intracardiac mass
    i) Typically irregular shape
    ii) Pedunculated or sessile
    iii) Rarely impair valve motion
    iv) Often flutter or vibrate
e. New valvular regurgitation
f. Secondary effects of valvular regurgitation
g. Abscess
h. New partial dehiscence of prosthetic valve

5. Complications
    a. Cusp or leaflet rupture/flail
    b. Perforation
    c. Abscess
    d. Aneurysm
    e. Fistulae
    f. Dehiscence of prosthetic valve
    g. Pericardial effusion
    h. Hemodynamic compromise
        i) Regurgitation (acute or chronic)
        ii) Valvular stenosis
        iii) Shunt
        iv) CHF
    i. Embolization
        i) Cerebral
        ii) Systemic
        iii) Pulmonary
6. Differential diagnosis
    a. Nonbacterial thrombotic endocarditis
    b. Active versus healed vegetations
    c. Lambl’s excrescences and valve strands
    d. Tumors, thrombi
    e. Sutures, strands on prosthetic sewing rings
    f. Focal, nonspecific thickening or calcium deposits
7. Related testing
    a. ECG

i) New conduction abnormalities
ii) New arrhythmia
iii) Evidence of ischemia or MI (coronary artery embolism)

b. Laboratory testing
i) Anemia
ii) Positive blood cultures

c. Chest x-ray
i) Evidence of CHF

d. Role of TEE
i) Greater sensitivity than TTE
ii) Intraoperative imaging

8. Common treatment

a. Medical
i) Prevention – endocarditis prophylaxis
ii) Antimicrobial therapy
iii) Treatment for heart failure
iv) Anticoagulation if clinically indicated

b. Surgical
i) Valve repair or replacement
ii) Role of echo in timing of surgery

K. Valvular Heart Disease Associated with Systemic Conditions

1. Systemic lupus erythematosus

a. General
i) Chronic inflammatory disease
ii) Valvular involvement often clinically silent

b. Echo findings
i) Mild, nonspecific valve thickening
ii) Libman-Sacks endocarditis (nonbacterial thrombotic)
- Typically only mitral or aortic valves involved
- Usually small with little independent motion
- Irregular borders

iii) Valve regurgitation
iv) Valve stenosis (rare)
v) Sensitivity of TTE low; TEE useful

2. Antiphospholipid syndrome

a. General
i) Hypercoagulability syndrome
ii) Valve lesions further increase risk for thromboembolic complications
iii) Superadded bacterial endocarditis may occur

b. Echo findings

i) Nonspecific valve thickening

ii) Libman-Sacks endocarditis (nonbacterial thrombotic)

3. Rheumatoid arthritis

a. General

i) Autoimmune disorder

ii) Immune system attacks the joints, specifically the synovium (thin layer of cells that line and lubricate the joints)

b. Echo findings

i) Valvular lesions similar to rheumatoid nodules

ii) Leaflet fibrosis, thickening, retraction

4. Ankylosing spondylitis

a. General

i) Chronic systemic inflammatory disorder

ii) Primarily affects the axial skeleton (hips and shoulders)

b. Echo findings

i) Nonspecific thickening of aortic and mitral leaflets

ii) Thickening of base of anterior mitral leaflet ("subaortic bump")

iii) Increased echogenicity of posterior aortic wall

iv) Aortic root dilatation

L. Prosthetic Valves

1. Classification and characteristics

a. Autograft (Ross procedure)

i) Native pulmonary valve sewn into aortic position

ii) Central flow

iii) Valve retains ability to grow

iv) Trivial or no regurgitation

v) Very low mean gradient

b. Homograft

i) Human donors

ii) Stentless

iii) Typically harvested as a block of tissue (aortic valve, root, and ascending aorta) and trimmed as needed

iv) Central flow

v) Trivial or no regurgitation

vi) Very low mean gradient

c. Bioprosthetic (tissue)

i) Stented

- Common examples (porcine)
- Common examples (bovine pericardium)
- Central flow
- Trivial or no regurgitation

ii) Stentless

- Intended for aortic or pulmonary position
- Common examples (porcine)
- Very low mean gradient

d. Mechanical

i) Bileaflet

- Two semicircular disks attached to a rigid valve ring by small hinges
- Common examples
- Two large lateral and one small central orifice create central flow
- Least obstructive mechanical prosthesis; allows more laminar blood flow
- Normal leakage volume at margin of central discs and at the periphery between disc and sewing ring

ii) Tilting (single) disk

- Single circular disk rotates within a rigid annulus, with the disk secured by lateral or central metal struts
- Common examples
- Low velocity, normal leakage volume; predominantly central with smaller peripheral jets

iii) Ball cage

- Circular sewing ring with several metal struts that "cage" a small hollow ball
- Blood flows around the spherical occluder
- Changes in chamber pressure causes ball to move back and forth
- Least optimal hemodynamics
- Normal closing volume regurgitation
- High profile limits use in patients with small ventricles

iv) TAVR

- Placements
- Size
- Perivalvular leaks

v) Valve conduits

- Mechanical or tissue prosthesis attached to a vessel graft or conduit

2. Tissue versus mechanical prostheses

a. Tissue prostheses

i) Advantages

- Less obstructive; better hemodynamics
- Fewer complications due to thromboembolism
- May not require anticoagulation

ii) Disadvantages

- Less durable than mechanical valves

- Leaflets may become regurgitant or stenotic due to tissue degeneration and calcification
- Limited availability of homografts

b. Mechanical prostheses

i) Advantages

- Extremely durable

ii) Disadvantages

- Inherently more obstructive
- Vulnerable to thromboembolism
- Require anticoagulation
- May experience catastrophic failure

3. Prosthetic valve dysfunction

a. Prosthetic stenosis/obstruction

i) Prostheses are inherently somewhat obstructive

ii) Gradient varies with:

- Valve type
- Valve size
- Anatomic position
- Cardiac output

iii) Etiology of obstruction

- Leaflet changes: thickening or calcification
- Thrombus or pannus
- Patient-prosthesis mismatch

b. Prosthetic regurgitation

i) Etiology

- Leaflet degeneration
- Prolapse or flail segment
- Thrombus or pannus
- Endocarditis
- Abnormalities in disk or poppet seating or motion
- Dehiscence

ii) Normal prosthetic regurgitation

- Leakage volume
  - Built-in transvalvular regurgitation of short duration
  - Decreases incidence of thrombosis
  - Volume increases with valve size and decreasing heart rate
  - Low velocity color profile
  - Multiple color jets related to multiple orifices
- Closure volume
  - Volume displaced by occluder during closure

- Ball cage prosthesis has largest closing volume

iii) Abnormal prosthetic regurgitation

- Transvalvular (prosthetic) regurgitation
  - Abnormal leakage through the valve
  - Tissue valves tend to leak with age and calcification
  - May see trivial leakage with a normal tissue prosthesis
  - Thrombus on mechanical valves may cause regurgitation
- Perivalvular (periprosthetic) regurgitation
  - Usually eccentric, high velocity and turbulent
  - Leakage from outside the sewing ring
  - Usually due to valve bed abnormalities or endocarditis

c. Thromboembolism

i) Higher risk with:

- Mechanical valves
- Atrial fibrillation
- Large LA
- LV dysfunction

d. Hemolysis/anemia

i) Mechanical trauma to red blood cells

ii) Some degree present with all mechanical prostheses

iii) Periprosthetic leaks, eccentric regurgitant jets, and obstruction are common culprits

e. Endocarditis

i) Higher risk of SBE in patients with prosthetic valves

ii) Prophylaxis recommended

f. Pannus

i) Fibrous ingrowth of tissue

ii) Rarely affects tissue prostheses

iii) May cause obstruction or regurgitation

iv) Disc and bileaflet valves are particularly prone

g. Prosthetic structural failure

i) Poppet/cage variance

ii) Strut fracture

iii) Valve degeneration or calcification of tissue valves

h. Valve bed abnormality – often a complication of endocarditis

i) Pseudoaneurysm

ii) Ring abscess

iii) Fistula

iv) Dehiscence

4. Echocardiographic assessment

a. Should include assessment of all cardiac chambers and other valves for incidental evidence of normal or abnormal prosthetic valve function
b. Imaging challenges
    i) Acoustic shadowing or artifacts from mechanical prosthesis or struts
    ii) May require multiple, unconventional imaging planes and extensive image adjustment
    iii) Consider use of TEE, especially for mitral prostheses
c. Patient should have baseline echo within 30 days of valve replacement to "fingerprint" the prosthesis
d. Serial studies
    i) Compare to baseline post operative echo
    ii) Assess for obstruction, regurgitation or complications
e. Clues to prosthetic dysfunction
    i) High or increasing gradient
    ii) Decreased valve area
    iii) Increased intensity of CW Doppler signal
    iv) Progressive chamber dilatation
    v) Recurrent or unexplained pulmonary hypertension
    vi) Flow convergence on LV side of MV
    vii) Intermittent flow variations

5. 2-D assessment
    a. Cardiac chamber size and function
    b. Stability of sewing ring
        i) Stable versus excessive or rocking motion
    c. Evidence of obstruction or regurgitation
        i) Reduced occluder, cusp or leaflet motion
        ii) Abnormal thickening or calcification of leaflets
        iii) Flail leaflet
    d. Extraneous echoes
        i) Pannus
        ii) Thrombus
        iii) Suture material
        iv) Vegetation
        v) Remnants of native valve apparatus
    e. Valve bed abnormalities
        i) Abscess
        ii) Pseudoaneurysm
        iii) Fistula
6. Color flow Doppler
    a. Identify presence and severity of valvular or perivalvular regurgitation
7. Doppler assessment (general comments)
    a. Assess prosthesis as if a stenotic native valve

b. Use multiple transducer positions (parallel to flow)

c. Indicate window from which the optimal Doppler signals were obtained

d. Average 3-5 beats in sinus rhythm, 8-10 in atrial fibrillation

8. Doppler assessment of aortic valve prosthesis

a. Measure peak prosthetic velocity

b. Measure mean gradient (more useful than maximal gradient)

i) If LVOT velocity is elevated (high-output state), LVOT mean gradient should be subtracted from prosthetic mean gradient

c. Calculate prosthetic effective orifice area (EOA)

i) Use continuity equation as if calculating AV area

ii) Measure LVOT diameter in parasternal long-axis view

- Measure LVOT diameter (insertion of sewing ring to ventricular septum and anterior mitral leaflet) during systole
- May use sewing ring diameter of prosthesis, but less accurate
- Note use of measured LVOT diameter or sewing ring approximation in report

iii) Measure LVOT velocity and VTI with PW Doppler

- Place PW sample volume below region of flow convergence (usually about 1 cm below prosthesis)
- Trace signal to obtain velocity and VTI

iv) Measure peak prosthetic velocity and VTI with CW Doppler from multiple windows

v) Calculate LV stroke volume and EOA

d. Dimensionless velocity index (DVI) (former DOI)

i) May use if unable to measure LVOT diameter

e. Intraventricular gradients

i) May occur after surgery for AVR

ii) Due to hemodynamic change – reduction of afterload in a small, hypertrophied ventricle

f. Interpretation of Doppler data

i) Need initial post-operative or pre-dismissal echo, then serial echo

ii) Consider valve type and size

iii) Consider EOA

9. Doppler assessment of MV prosthesis

a. Measure E and A velocities of mitral inflow by CW Doppler

b. Measure mean gradient by CW Doppler

c. Calculate prosthetic effective orifice area (EOA) using continuity equation (if no significant MR or AR)

d. Measure pressure half-time

e. Calculate pulmonary artery pressure

f. Interpretation of Doppler data

i) Compare to baseline post-operative echo

ii) Consider valve type and size

iii) Consider mean gradient in relation to heart rate

iv) Excessive increase in mean gradient or PAP with exercise

v) Consider EOA

10. Doppler assessment of tricuspid valve prosthesis
    a. CW Doppler (average 5-10 signals due to respiratory variation)
        i) E and A velocities
        ii) Mean gradient
        iii) Pressure half-time
        iv) Pulmonary artery pressure
11. Assessment of prosthetic/periprosthetic valve regurgitation
    a. Surface echo can usually adequately assess aortic prosthetic regurgitation, but not mitral prosthetic regurgitation
    b. Two-dimensional echo
        i) Assess chamber sizes and function
        ii) Determine etiology of regurgitation
    c. Color Flow Doppler
        i) Determine whether regurgitation is valvular or perivalvular
        ii) Note presence of eccentric jet
        iii) Look for flow convergence on the ventricular side of MV prosthesis
        iv) Use multiple imaging planes and non-standard views
    d. PW Doppler
        i) Mitral deceleration time (AR)
        ii) Assess diastolic flow reversal VTI in descending or abdominal aorta (AR)
        iii) Note presence of systolic flow reversals in pulmonary veins (MR)
    e. CW Doppler
        i) Note velocity, intensity and duration of regurgitant signal
        ii) Note change in velocity, gradient or pressure half-time from prior echo
        iii) Calculate PA pressure
    f. Quantitate regurgitation if possible
12. Role of transesophageal echocardiography (TEE)
    a. May be useful if surface study suboptimal
    b. Difficult to assess mitral prosthetic regurgitation by surface echo due to shadowing
    c. TEE probe is on atrial side of MV prosthesis, so mitral prosthetic regurgitation can be more clearly defined
    d. Intraoperative assessment
13. TAVR techniques
    a. Types of valves
    b. Echocardiography's role
    c. Technical considerations

Section VIII: Cardiomyopathies

1. Discuss the echocardiographic features associated with hypertrophic cardiomyopathy (HCM), dilated cardiomyopathy (DCM), restrictive cardiomyopathy (RCM) and arrhythmogenic RV dysplasia
2. Differentiate between the echocardiographic features of cardiomyopathies
3. Differentiate between primary and secondary etiology in DCM
4. List common infiltrative systemic myocardial diseases

VIII. Cardiomyopathies

A. Hypertrophic Cardiomyopathy
   1. Epidemiology
      a. Prevalence
      b. Associated risk factors for sudden death
      c. Genetics
   2. Etiologies
   3. Clinical presentations
      a. Signs and symptoms
      b. Physical findings
      c. Auscultatory finding
   4. Types/varied patterns
      a. Classic - asymmetric septal hypertrophy (ASH)
      b. Midventricular
         i) With apical aneurysm
         ii) Without apical aneurysm
      c. Apical
      d. Concentric
      e. Other
   5. Provocative maneuvers
      a. Standing
      b. Valsalva
      c. Amyl nitrate
   6. 2-D features
      a. Small or normal LV cavity
      b. Normal or hyperdynamic LV function
      c. Extent of hypertrophy
      d. Mitral apparatus
         i) Systolic MV anterior motion (SAM)
            - One or both leaflets
            - Degree
            - Mechanism
              - Venturi effect

- Anterior papillary muscle displacement and elongation
- Other situations
  - MV repair
  - AV replacement
  - Hypovolemia
  - Endogenous and exogenous catecholamines

ii) Elongated anterior mitral leaflet (AML)

iii) Increased area of AML

iv) Calcified mitral annulus

e. LA enlargement

f. Narrowed LVOT diameter

g. Altered texture of myocardium

h. Basal septal endocardial thickening (contact lesion)

7. M-mode features
   a. Normal or small LV size
   b. Extent of hypertrophy
   c. Normal or increased shortening fraction
   d. LA enlargement
   e. Abnormal MV motion
      i) Reduction of E-F slope
      ii) SAM of leaflet
      iii) Mitral annular calcification
   f. B-bump
   g. AV mid-systolic closure
   h. Color M-Mode of MV and LVOT obstruction
8. Doppler features
   a. Diastolic dysfunction assessment
      i) Characteristics
         - Impaired relaxation
         - Decreased compliance
         - Increased LVEDP
         - Decreased EA ratio
         - Normal filling pattern
           - Normalization by MR
           - Normalization by increased LA pressure

      ii) Poor correlation between DT and LA pressure

      iii) Presumed cause of symptoms in patients with no obstruction

   b. MR with or without SAM present
      i) MR likely seen with obstructive SAM

      ii) Eccentric

- Posterolateral
- Late systolic

iii) Primary MR versus secondary MR

c. Level of obstruction (LVOT versus midcavity)

i) Doppler wave patterns

- Late peaking
- Reliably assessed by Bernoulli equation

ii) Provocative maneuvers

9. Limitations and pitfalls

a. Differential diagnosis

i) Chronic hypertension

ii) Cardiac amyloid

iii) Pheochromocytoma

iv) Freidreich's ataxia

v) Inferior MI with previous LVH

b. Dynamic LVOT obstruction not specific

i) Hyperdynamic LV systolic function

- Anemia
- Volume depletion
- Catecholamines

ii) Hypertensive HCM in elderly

iii) Postoperative period

iv) Sub aortic obstruction post AVR

v) Acute anteroapical MI with pre-existing basal septal hypertrophy

10. Related testing

a. ECG

b. Laboratory testing

c. Correlative imaging

11. Common treatments

a. Role of echo

i) Evaluation of therapy

ii) Pacemaker implementation and therapy

iii) Septal ablation

iv) Myotomy/myectomy

B. Dilated Cardiomyopathy

1. Epidemiology

a. Prevalence

b. Genetics

2. Etiologies

3. Clinical presentations

a. Signs and symptoms
b. Physical findings
c. Auscultatory finding

4. 2-D features
   a. Chamber enlargement
   b. Hypokinesis of LV
   c. Regional wall motion abnormality
   d. Mural thrombi in apex or aneurysm
   e. Spontaneous echo contrast
5. M-mode features
   a. Chamber dilation
   b. Hypokinesis of LV
   c. Decreased LV wall thickening
   d. Decreased fractional shortening
   e. Increased EPSS
   f. Reduced aortic root motion
   g. Reduced ejection fraction
6. Doppler features
   a. MR and TR assessment
   b. Diastolic dysfunction assessment
   c. RV systolic pressure assessment
   d. LVEDP assessment
7. Prognostic role of echo
   a. EF
   b. DT
   c. RV function
8. Related testing
   a. ECG
   b. Laboratory testing
   c. Correlative imaging
9. Common treatments

C. Restrictive/Infiltrative Cardiomyopathy
   1. Etiologies of restrictive cardiomyopathy
      a. Primary
         i) Idiopathic
      b. Secondary
         i) Amyloid heart disease
         ii) Hemochromatosis
         iii) Heart muscle disease
            - Post-irradiation heart disease

- Carcinoid heart disease
- Doxorubicin/daunorubicin toxicity
- Progressive systemic sclerosis

iv) Eosinophilic endomyocardial disease

2. Classifications of infiltrative CM
   a. Infiltrative
      i) Interstitial
         - Amyloid
         - Hemochromatosis
         - Sarcoid
         - Malignancy
      ii) Storage
         - Glycogen
         - Lipid
3. Clinical presentations
   a. Signs and symptoms
   b. Physical findings
   c. Auscultatory finding
4. 2-D features
   a. Diffuse speckling or granular appearance of myocardium (amyloid)
   b. Pericardial or pleural effusion
   c. Bi-atrial enlargement
   d. Small to normal LV cavity
   e. Possible LVH
   f. Normal systolic function
5. M-mode features
   a. Normal LV size
   b. Concentric LVH
   c. Decreased LV shortening fraction
   d. Pericardial or pleural effusion
6. Doppler features
   a. MR and TR assessment
   b. Diastolic dysfunction assessment including TDI
   c. Minimal or absent respiratory variation in Doppler flows
7. Differential diagnosis
   a. Constrictive pericarditis
8. Related testing
   a. ECG
   b. Laboratory testing
   c. Correlative imaging

9. Common treatment

D. Arrhythmogenic RV Dysplasia
   1. Etiology
   2. Characteristics
      a. Loss of RV myocardium with fatty or fibro-fatty replacement of tissue
      b. Associated with ventricular arrhythmia
      c. Associated with sudden death in young
   3. Clinical presentations
      a. Signs and symptoms
      b. Physical findings
      c. Auscultatory finding
   4. 2-D features
      a. Dilated RV
      b. Aneurysm, outpouching of the RV
      c. Focal RV wall thinning
      d. RV wall motion abnormality
      e. Abnormal RV muscle composition
   5. Other arrhythmogenic RV cardiomyopathies
      a. RVOT tachycardia
      b. Benign extrasystoles
      c. Uhl's anomaly
      d. Biventricular dysplasia
   6. Related testing
      a. ECG
      b. Laboratory testing
      c. Correlative imaging
   7. Common treatment

Section IX: Systemic and Pulmonary Hypertensive Heart Disease

1. Explain the pathophysiology of systemic hypertensive disease
2. Explain the pathophysiology of pulmonary hypertensive disease
3. List classifications of pulmonary hypertension
4. Describe the echocardiographic features associated with systemic and pulmonary hypertensive disease
5. Define Eisenmenger's Syndrome

IX. Systemic and Pulmonary Hypertensive Heart Disease

A. Systemic Hypertension
   1. Etiology and risk factors
   2. Pathophysiology
      a. Increased afterload leads to left ventricular hypertrophy (LVH) and increased mass
      b. Ventricular hypertrophy leads to diastolic dysfunction
   3. 2-D and color flow features
      a. Increased LV mass and mass index
      b. Concentric LVH
      c. Normal or hyperdynamic ejection fraction (early)
      d. Dynamic LVOT or mid-cavitary obstruction
      e. LA enlargement due to increased left ventricular end-diastolic pressure (LVEDP)
      f. Aortic root dilatation
      g. MAC with associated mitral regurgitation
      h. AV sclerosis with associated AR
   4. Diastolic abnormalities
      a. Abnormal relaxation
         i) Early
         ii) Minimal or no symptoms at rest
      b. Pseudonormalization
         i) Later
         ii) Minimal or no symptoms at rest
         iii) Symptoms with mild/moderate exertion
      c. Restrictive filling
         i) Significantly elevated LV pressure
         ii) Symptoms at rest or with minimal exertion
   5. Prognostic value of echocardiography
   6. Complications of hypertension
   7. Treatment

B. Pulmonary Hypertension (PHTN)
   1. Definition
      a. Pulmonary artery systolic pressure (PASP) > 25 mm Hg at rest or > 30 mmHg with exercise

2. Etiology
    a. Primary
    b. Secondary
3. Pathophysiology
    a. Response of RV to chronic pressure overload is right ventricular hypertrophy (RVH) and dilatation
4. Classification
    a. Pulmonary arterial hypertension
    b. Pulmonary venous hypertension
    c. PHTN associated with hypoxemia
    d. PHTN due to chronic thrombotic and/or embolic disease
    e. Miscellaneous
5. Echocardiographic features
    a. RV enlargement and hypertrophy
    b. Dilated PA and branches
    c. RA enlargement
    d. Decreased RV systolic function (late)
    e. D-shaped LV in short axis view
    f. Paradoxical septal motion
    g. Mid-systolic closure of PV
    h. Dilated IVC and hepatic veins
    i. TR and PR secondary to annular dilatation
    j. Elevated PASP and PAEDP
6. Eisenmenger's Syndrome
    a. Reversal of intracardiac shunt from left-to-right to right-to-left secondary to severe pulmonary hypertension
7. Cor pulmonale
    a. Acute (pulmonary embolism)
    b. Chronic
8. Prognostic value of echocardiography
9. Treatment

Section X: Pericardial Diseases

1. List key etiologies of pericardial disease
2. Explain the pathophysiology and hemodynamics of cardiac tamponade and constrictive pericarditis
3. Describe key echocardiographic features of cardiac tamponade and constrictive pericarditis
4. Explain Doppler criteria associated with cardiac tamponade and constrictive pericarditis
5. Describe key echocardiographic features of congenital absence of the pericardium

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X. Pericardial Diseases

- A. Normal Pericardium
  - 1. Structure
  - 2. Function
- B. Pericarditis/Pericardial Effusion
  - 1. Etiologies
  - 2. Clinical presentations
    - a. Signs and symptoms
    - b. Physical findings
    - c. Auscultatory finding
  - 3. Echocardiographic Features
    - a. Echo-free space
    - b. Variable size and location
  - 4. Differentiation from pleural effusion, epicardial adipose, and other posterior echo-free spaces
  - 5. Treatment
- C. Cardiac Tamponade
  - 1. Etiologies
  - 2. Clinical presentation
    - a. Signs and symptoms
      - i) Chest pain
      - ii) Dyspnea
    - b. Physical findings
      - i) Tachycardia
      - ii) Narrow pulse pressure
      - iii) Pulsus paradoxus
      - iv) Elevated jugular venous pressure (JVP)
      - v) Beck's triad
    - c. Auscultatory findings
    - d. ECG findings
    - e. Correlative imaging
  - 3. Hemodynamics
    - a. Accumulation of pericardial fluid causes pericardial pressure to rise and compromise systemic venous return in right atrium

b. Ventricular filling is impaired and cardiac output is decreased
c. Intrapericardial pressure is influenced by both volume of fluid and rate at which it accumulates
d. Dissociation of intrathoracic and intracardiac pressures due to insulating effect of effusion
e. Relatively fixed combined cardiac volume (ventricular interdependence)

4. Echocardiographic features
   a. Pericardial effusion
   b. RA or RV early diastolic chamber collapse
   c. IVC plethora
   d. Reciprocal changes in ventricular volumes (septal shift)
   e. "Swinging heart" if large effusion
5. Doppler features
   a. Respiratory variation in velocities
   b. Inspiration
      i) Decreased mitral E velocity
      ii) Decreased pulmonary venous diastolic forward flow
      iii) Increased tricuspid E velocity
   c. Expiration (reciprocal changes)
      i) Increased mitral E velocity
      ii) Increased pulmonary venous diastolic forward flow
      iii) Decreased tricuspid E velocity
      iv) Decrease or loss of hepatic vein diastolic filling with marked expiratory reversal
   d. Technical caveats
      i) Use of respirometer preferred
      ii) Typical respiratory patterns are reversed in a mechanically ventilated patient
      iii) Changes may not be seen in patients with high filling pressures, ASD or RVH
6. Echo-guided pericardiocentesis
   a. Treatment of choice
      i) Locate optimal site of puncture
      ii) Determine depth of pericardial effusion and distance from puncture site to effusion
      iii) Confirm position of needle by use of agitated saline
   b. Pigtail catheter introduced and left in for several days with intermittent drainage
   c. Low complication rate

D. Constrictive Pericarditis
1. Etiology
   a. May develop in the aftermath of virtually any pericardial injury or inflammation
2. Clinical presentations
   a. Signs and symptoms
      i) Dyspnea
      ii) Chest pain
   b. Physical findings

i) Jugular venous distention
ii) Edema
iii) Ascites
iv) Kussmaul sign
v) Pulsus paradoxus
vi) Hepatosplenomegaly

c. Auscultatory findings
i) Pericardial knock

d. ECG findings

e. Correlative imaging

3. Pathophysiology and hemodynamics
a. Thickened and/or fibrotic pericardium externally constrains diastolic filling of the heart
b. Elevated diastolic pressures
c. Rapid early diastolic filling that stops abruptly as limit of ventricular expansion is achieved
d. Fibrotic pericardium prevents full transmission of intrathoracic pressure changes (dissociation of intrathoracic and intracardiac pressures)
e. Exaggerated ventricular interdependence

4. 2-D/M-mode features
a. Thickened pericardium
b. Normal LV size and EF
c. Respiratory variation in ventricular size
d. Enlarged atria
e. Flattening of LV posterior wall in diastole
f. Abrupt posterior motion of ventricular septum in early diastole (septal bounce)
g. Dilated IVC and hepatic veins
h. Ascites

5. Doppler features
a. Respiratory variation in velocities
b. Mitral inflow pattern typically restrictive
c. Inspiration
i) Decreased mitral E velocity
ii) Decreased pulmonary venous diastolic forward flow
iii) Increased tricuspid E velocity

d. Expiration (reciprocal changes)
i) Increased mitral E velocity
ii) Increased pulmonary venous diastolic forward flow
iii) Decreased tricuspid E velocity
iv) Decreased or loss of diastolic filling with marked expiratory reversal

e. Tissue Doppler
i) e' velocity relatively normal or accentuated in constriction

f. Technical caveats

i) Use of respirometer preferred
ii) Typical respiratory patterns are reversed in a mechanically ventilated patient
iii) Changes may not be seen in patients with high filling pressures, ASD or RVH

6. Differentiating constriction from other disorders
   a. Restriction
      i) Lack of respiratory variation
      ii) Tissue Doppler e' velocity is reduced
      iii) E/e' ratio is increased
      iv) Color M-mode shows prolonged slope
   b. COPD or obesity
      i) Prominent increase in SVC forward flow during inspiration
      ii) Mitral inflow not typically restrictive
      iii) Hepatic vein flow not characteristic of constriction
7. Pitfalls
   a. High filling pressure
   b. Sample volume placement (translational motion)
   c. Atrial fibrillation
   d. Localized constriction
8. Treatment
   a. Pericardiectomy

E. Other Pericardial Disease
1. Tumors
   a. Types
      i) Primary
      ii) Metastatic
   b. Characteristics
   c. Echocardiographic features
      i) Variable
      ii) Most often multiple
   d. Associated findings
      i) Pericardial effusion
2. Pericardial cyst
   a. Characteristics
   b. Echocardiographic features
      i) Echolucent space
      ii) Frequently adjacent to RA

F. Congenital Absence of Pericardium
1. General
   a. Uncommon
   b. More frequent in males

2. Types
    a. Total absence
    b. Partial absence
3. Echocardiographic features
    a. Unusual echo windows
    b. Cardiac hypermobility
    c. Marked shift of cardiac chambers to the right
    d. Abnormal ventricular septal motion
4. Complications
    a. Herniation or strangulation of left atrial appendage with partial absence

Section XI: Cardiac Tumors/Masses

1. List benign and malignant cardiac tumors
2. Describe echocardiographic criteria used for evaluating tumors
3. Differentiate cardiac tumors from cardiac structures
4. List potential sources of artifactual echoes within the chambers

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XI. Cardiac Tumors/Masses

A. Thrombus
   1. Echocardiographic characterization
      a. Location and site of attachment
      b. Size
      c. Degree of mobility
      d. Shape
      e. Co-existing cardiac abnormalities
   2. Potential for embolization
      a. Size
      b. Protrusion into cavity
      c. Mobility
   3. Technical suggestions
      a. Use high-frequency, short-focus transducers
      b. Decrease depth of field
      c. Move focal zone nearer apex
      d. Use nonstandard views
      e. Contrast enhanced echocardiography
   4. Pitfalls
      a. Artifact
      b. Must differentiate from prominent trabeculations, papillary muscles, anomalous chords
      c. Laminated (non-protruding) thrombi more difficult to appreciate
   5. LV thrombus
      a. Predisposing conditions
         i) Underlying regional wall motion abnormalities
         ii) LV aneurysm
         iii) Diffuse LV systolic dysfunction
      b. Echocardiographic features
         i) Contour distinct from endocardial border
         ii) Often more echogenic than underlying myocardium
         iii) Located in region of abnormal wall motion
   6. LA and LAA thrombus
      a. Predisposing factors
         i) Atrial fibrillation

ii) MS
iii) Prosthetic MV
iv) LA enlargement

b. Association with spontaneous echo contrast
c. TEE more sensitive than TTE
d. Clinical significance
i) Increased risk of thromboembolism
ii) Relative contraindication to balloon mitral valvotomy
iii) Relative contraindication to cardioversion

7. RA thrombus and thrombus-in-transit
a. Predisposing factors
i) Atrial fibrillation
ii) RA enlargement
iii) Catheters, pacemakers
b. Clinical significance
i) Embolization (pulmonary or paradoxical)
ii) Thrombi on catheters, pacemakers have potential for infection
c. Often a “popcorn” appearance
d. Need to distinguish from
i) Congenital remnants (Eustachian valve, Chiari network)
ii) Reverberation artifacts
iii) Lipomatous hypertrophy of atrial septum

8. RV thrombus
a. Relatively uncommon

9. Treatment
a. Anti-coagulation
b. Thrombolytics
c. Thrombectomy

B. Primary
1. Benign
a. Myxoma
b. Papillary fibroelastoma
c. Fibroma
d. Rhabdomyoma
e. Lipoma
f. Hemangioma
g. Miscellaneous others

2. Malignant
a. Sarcomas
b. Mesothelioma

c. Malignant lymphoma
d. Miscellaneous others

3. Characteristics
   a. Etiology
   b. Clinical presentation
      i) Signs and symptoms
      ii) Physical findings
      iii) Auscultatory finding
   c. Common locations
4. Echocardiographic features
   a. Occur more frequently in right heart
   b. Sessile, with intracavitary extension

C. Secondary (Metastatic)
1. Most often metastasize to pericardium with associated effusion
2. Myocardial invasion, intracavitary extension
3. Occur more frequently in right heart
4. Tumors invading right heart from IVC
   a. Renal cell carcinoma
   b. Hepatoma
   c. Uterine tumors
5. Characteristics
   a. Etiology
   b. Clinical presentations
      i) Signs and symptoms
      ii) Physical findings
      iii) Auscultatory findings

D. Role of Echocardiography
1. Location
2. Tumor characteristics
   a. Morphology
   b. Single or multiple
   c. Size
   d. Degree of mobility
   e. Point of attachment
3. Effect on cardiac and valvular function
4. Limitations
   a. Inability to determine histology
   b. Limited acoustic access in some patients
   c. Limited field of view (mediastinum, adjacent structure)
   d. Differential diagnosis

E. Differentiation from Other "Masses"
   1. Extracardiac
      a. Cysts
         i) Mediastinal
         ii) Pancreatic
         iii) Pericardial
         iv) Bronchogenic
      b. Hematoma
      c. Thymoma
      d. Teratoma
      e. Diaphragmatic or hiatal hernia
      f. Aorta
   2. Intracardiac
      a. Crista terminalis
      b. Congenital remnants
      c. Trabeculations
      d. Moderator band
      e. Papillary muscles
      f. Redundant chordae
      g. Myxomatous tissue
      h. Vegetations
      i. Tuberculomas
      j. Lipomatous hypertrophy of the atrial septum
      k. Mitral annular calcification
      l. Chiari network
      m. Eustachian valves
      n. Catheter or pacemaker wire
      o. Others
   3. Artifacts
      a. Side lobes
      b. Reverberation
      c. Improper gain settings

Section XII: Diseases of the Aorta

1. Describe echocardiographic features associated with Marfan syndrome
2. List the etiologies of aortic aneurysm and dissection
3. Describe two classifications of dissection
4. Explain the echocardiographic features of aortic aneurysm, and sinus of Valsalva aneurysms and dissection.

XII. Diseases of the Aorta

- A. Marfan Syndrome
  - 1. Etiology
  - 2. Clinical presentation
    - a. Signs and symptoms
    - b. Physical findings
      - i) Cardiac manifestations
      - ii) Non-cardiac manifestations
    - c. Auscultatory findings
  - 3. 2-D features
    - a. Aortic root dilatation
      - i) Dilated aortic annulus
      - ii) Dilated Sinuses of Valsalva
    - b. Dilated ascending aorta
    - c. Annuloaortic ectasia
    - d. Aortic dissection
    - e. Myxomatous mitral valve and mitral valve prolapse
  - 4. Doppler features
    - a. AR
    - b. MR
  - 5. Related testing
    - a. ECG
    - b. Laboratory testing
    - c. Correlative imaging
  - 6. Management
- B. Aortic Aneurysm
  - 1. Definition: dilatation of aorta with intact vessel layers (intima, media, adventitia)
  - 2. Etiology
    - a. Atherosclerosis
    - b. Medial degeneration
      - i) Annuloaortic ectasia
      - ii) Marfan syndrome
      - iii) Associated with bicuspid aortic valve
      - iv) Other heritable disorders

c. Trauma
d. Syphilis
e. Mycotic
f. Noninfectious aortitis
g. Aortic dissection with dilatation of persisting false lumen

3. Clinical presentation
   a. Signs and symptoms (often asymptomatic)
   b. Physical findings
   c. Auscultatory finding
4. Classification
   a. Morphology
      i) Fusiform
      ii) Saccular
   b. Location
      i) Ascending
      ii) Arch
      iii) Descending
      iv) Thoracoabdominal
5. Goal of imaging
   a. Confirm or exclude diagnosis
   b. Measurement of diameter and length
   c. Determine involvement of aortic valve or arch vessels
   d. Differentiate from aortic dissection
   e. Detect mural thrombus
6. Echocardiographic features
   a. Aortic dilatation
   b. Aortic regurgitation
   c. Diastolic flutter of mitral valve with significant AR
   d. Flow in false lumen if dissected
7. Role of TEE
8. Complications
   a. Aortic regurgitation
   b. Aortic rupture
      i) Pericardium
      ii) Left pleural space
   c. Aortic dissection
   d. Thromboembolism
   e. Compression of adjacent structures
9. Related testing
   a. ECG

b. Laboratory testing
c. Correlative imaging

10. Management
    a. Medical
    b. Surgical
        i) Time of operation controversial
        ii) Depends on many factors: cause, size, rate of change, other surgical risk
    c. Endovascular stenting
    d. Serial imaging

C. Aortic Dissection
1. Definition
    a. A tear in the intima layer of the aorta resulting in blood flowing into the aortic wall. As blood flows though the tear, the intima and media layers separate resulting in dissection. In some instances, dissection may occur due to hemorrhage within the media which then results in separation of the media and intima.
2. Etiology: same as for aneurysm
3. Clinical presentation
    a. Signs and symptoms
        i) Chest pain, typically radiating to the back, sudden onset, unremitting
        ii) Shock/hypotension
    b. Physical findings
    c. Auscultatory finding
4. Classifications
    a. Stanford Types A and B
    b. DeBakey Types I, II, III
5. Echocardiographic features
    a. Intimal flap
    b. True lumen versus false lumen (2D and color flow)
    c. Possible LV enlargement
    d. Thrombus in the false lumen
    e. Effusion
    f. Increased aortic wall thickness (Intramural hematoma)
    g. Aortic regurgitation due to
        i) Aortic root dilatation
        ii) Asymmetric dissection causes incomplete coaptation
        iii) Dissection into commissure causes incomplete coaptation
        iv) Intimal flap prolapses through AV
    h. Pericardial and/or pleural effusion
    i. Compression of LA
    j. RWMA with coronary ostia involvement
6. Goal of imaging

a. Confirm or exclude diagnosis
b. Determination of location, characteristics and extent of dissection
c. Presence, severity, and mechanism of AR
d. Presence of pericardial or pleural effusion
e. Involvement of coronary arteries or branch vessels
f. Detection of rupture

7. Role of TEE
   a. Diagnostic procedure of choice
   b. Advantages
   c. Limitations
8. Differential diagnosis
   a. Reverberations, catheters
   b. Mirror-image artifacts
   c. Hemiazygous sheath
   d. Thoracic aortic aneurysm with thrombus
   e. Adjacent vein (Left brachiocephalic vein)
9. Related testing
   a. ECG
   b. Laboratory testing
   c. Correlative imaging
10. Management
    a. Medical
    b. Surgical
    c. Endovascular stenting

D. Sinus of Valsalva Aneurysm
1. Types and location
   a. Right coronary sinus (most common)
   b. Non-coronary sinus
   c. Left coronary sinus
   d. Many variations
   e. May present as ruptured or unruptured
2. Etiology
   a. Congenital
   b. Trauma
   c. Infective endocarditis
   d. Syphilis
   e. Marfan syndrome
3. Clinical presentation
   a. Signs and symptoms (ruptured)
      i) Dyspnea

ii) Chest pain
iii) Gradual onset and progression of symptoms

b. Physical findings
i) Wide pulse pressure

c. Auscultatory finding
i) Almost continuous murmur

4. Associated congenital anomalies
5. Echocardiographic features
a. Round or fingerlike (windsock) outpouchings
b. Dilatation of one or more sinuses
c. Abnormal dilatation of one or more of the sinuses; most often the right
d. TV systolic flutter
e. High velocity systolic/diastolic flow pattern with color and CW Doppler
f. Compression of adjacent structures
6. Complications
a. Rupture into cardiac chamber
b. RVOT obstruction
c. Endocarditis
d. AR
e. TR
f. Erosion into ventricular septum
g. Obstruction of adjacent structures
i) Coronary artery compression
ii) SVC obstruction
iii) TS
7. Serial evaluation of unruptured aneurysm
8. Related testing
a. ECG
b. Laboratory testing
c. Correlative imaging
9. Management
a. Medical
b. Surgical
10. Other Aortic Pathology
a. Aortic atherosclerosis
b. Aortic pseudoaneurysm
c. Penetrating aortic ulcer
d. Aortic hematoma
e. Traumatic injury of the aorta

Section XIII: Normal and Altered Electrical Activation

1. Describe normal electrical pathways
2. Define normal electrical wave forms seen on ECG
3. Describe echocardiographic findings associated with altered electrical activation
4. Identify abnormal ECG tracings
5. Recognize abnormal ECG tracing

XIII. Normal and Altered Electrical Activation

- A. Normal Electrical Activation
  - 1. Electrical pathways
    - a. Sino atrial (SA) node
    - b. Atrio ventricular (AV) node
    - c. Bundle of His
    - d. Left and right bundle branches
    - e. Purkinje fibers
  - 2. Electrical waves, intervals and segments
    - a. P wave
    - b. QRS
    - c. T wave
    - d. U wave
    - e. P-R interval
    - f. Q-T interval
    - g. QT segment
    - h. ST segment
- B. Altered Electrical Activation
  - 1. Electrocardiographic features
    - a. Left bundle branch block (LBBB)
    - b. Right bundle branch block (RBBB)
    - c. Wolff-Parkinson White (WPW)
    - d. Ischemic changes
      - i) ST segment depression
      - ii) ST segment elevation
      - iii) Q waves
    - e. Arrhythmias
      - i) Premature ventricular contraction
      - ii) Premature atrial contraction (PAC)
      - iii) Sinus arrhythmia
      - iv) Supraventricular tachycardia
      - v) Quadrigeminy, trigeminy, bigeminy
      - vi) Atrial flutter

vii) Atrial fibrillation
viii) Ventricular tachycardia
ix) Ventricular fibrillation
x) Junctional

f. Heart block
   i) First-degree AV block
   ii) Second-degree AV block
   iii) Wenckebach, mobitz I, II
   iv) Complete heart block

g. Pacemaker
   i) Single chamber
   ii) Dual chamber
   iii) Bi-ventricular pacemakers

Section XIV: Pediatric and Adult Congenital Heart Disease

1. Describe echocardiographic findings associated with various types of congenital heart disease
2. Discuss clinical signs and symptoms associated with congenital heart disease
3. Explain how the segmental approach is utilized to identify and characterize congenital heart disease
4. Describe how the echocardiography exam is tailored for each type of congenital heart disease listed in this outline
5. List the parameters used in qualitative and quantitative assessment of congenital heart disease for each type described in this outline
6. Describe common surgical repairs seen in the pediatric and adult patients with congenital heart disease

Notes:

1. Due to the depth of this subject, rare forms of congenital heart disease are not discussed in this section.
2. When discussing image orientation the following apply:
   A. The term "orientation mark" refers to the orientation ridge on the transducer: When imaging congenital heart disease in the pediatric echocardiography laboratory, images are usually oriented in the "anatomically correct" position on the display monitor. Thus, anterior and superior anatomical structures are usually displayed at the top of the display monitor, and rightward anatomical structures are usually displayed on the left side of the display monitor. The parasternal long axis view is the only view displayed with the apex (inferior structure) to the left side of the display monitor
   B. The phrase "image sector" refers to the image as it is seen in the sector window on the display monitor
3. When describing "angling" or "angled", these terms are referring to angling the face of the transducer (non-cord) end.
4. All numerical values in this section are from the following two references;
   A. Hugh D. Allen, MD, David J. Driscoll, MD, Robert E. Shaddy, MD, Timothy E. Feltes, MD, Moss and Adams' Heart Disease in Infants, Children and Adolescents: Including the Fetus and Young Adult, 7th Edition . Lippincott, Williams & Wilkins. Philadelphia, Pennsylvania. 2008.
   B. Lai, W, Mertens L, Cohen M, Geva T. Echocardiography in pediatric and congenital heart disease: From Fetus to Adult. Wiley-Blackwell, Hoboken, New Jersey. 2009.

XIV. Pediatric and Adult Congenital Heart Disease

A. Segmental Approach to Cardiovascular Anatomy
   1. Right atrial morphology
      a. Receives suprahepatic portion of IVC
      b. Right atrial appendage (RAA); broad-based, triangular
      c. Pectinate muscles within chamber
      d. Receives coronary sinus (if intact)
   2. Left atrial morphologic features
      a. Left atrial appendage (LAA); thin, narrow-based
      b. Septum primum attachments
      c. Smooth walled; pectinate muscles limited to the LAA
   3. Common atrium
      a. Absent septum secundum, septum primum
      b. Associated with isomerism of the atrial appendages (heterotaxy syndrome)
      c. Commonly associated with anomalies of systemic and pulmonary venous return
   4. Right ventricular morphology
      a. Coarsely trabeculated endocardium (including the septal surface)

b. Moderator, septal and parietal bands
c. "Septophillic" attachments of the tricuspid valve
d. Muscular ventriculo-infundibular fold (also known as the subarterial conus) between TV and semilunar valve (normally pulmonic valve)

5. Left ventricular morphology
   a. Smooth superior septal surface
   b. Fine endocardial trabeculations
   c. Fibrous continuity between MV and semilunar valve (normally aortic)
6. Great artery morphology
   a. Pulmonary artery bifurcates into the left and right branch pulmonary arteries
   b. Aorta gives rise to coronary arteries and systemic arteries, including the brachiocephalic arteries

B. Segmental Anatomy - Situs [S, D, S]
1. Atrial segment { , D, S} – first letter of segmental set
   a. S - situs solitus; morphologic RA is right-sided, morphologic LA is left-sided
   b. I - situs inversus; morphologic RA is left-sided, morphologic LA is right-sided
   c. A - situs ambiguous; associated with isomerism of the atrial appendages
2. Ventricular segment {S, , S} – second letter of segmental set
   a. D – Loop; morphologic RV on the right side
   b. L – Loop; morphologic LV on the right side
   c. X – Unable to determine looping
      i) Topology (Chirality or Hand Rule) used in complex hearts with "crisscross" or superior/inferior ventricular arrangement
3. Great artery segment {S, D, } – third letter of segmental set
   a. S – solitus; normal position of the aortic valve is rightward and posterior to the PV
   b. I – inversus; aortic valve is leftward and posterior to the PV
   c. D – d-malposed; aortic valve is rightward and anterior to the PV
   d. L – l-malposed; aortic valve is leftward and anterior to the PV
   e. A – anterior; aortic valve directly anterior to the PV

C. Relational Anatomy and Cardiac Position
1. Abdominal and viscero-atrial situs
   a. Liver
   b. Inferior vena cava
   c. Descending aorta
2. Cardiac location
   a. Levoposition
   b. Mesoposition
   c. Dextroposition
   d. Ectopia cordis
3. Cardiac orientation
   a. Levocardia

b. Mesocardia

c. Dextrocardia

4. Segmental alignment and connections

a. Atrioventricular and ventriculoarterial concordance (normal)

b. Atrioventricular and ventriculoarterial discordance (congenitally "corrected" transposition of the great vessels)

5. Assessment of cardiac structural morphology

a. Display all structures in correct anatomical position

b. Determine direction where apex is pointing with reference to axis of the heart

D. Imaging Technique

1. Parasternal long axis

a. Transducer orientation known by identifying long axis of heart LV and orienting directing the orientation mark superiorly toward the cardiac base

b. Superior direction structures should be demonstrated towards the right side of the image sector

c. Inferior direction structures should be demonstrated on the left side of the image sector

2. Parasternal short axis

a. Rotate transducer 90 degrees clockwise from parasternal long axis view

b. Right side of image sector should always display left sided structures

c. Left side of image sector should always display right sided structures

d. Orientation marker directed to the left

e. Determine systemic atrium (atrium sending blood to the aorta)

f. Determine pulmonic atrium (atrium sending blood to the PA)

g. Determine morphologic RA

h. Determine morphologic LA

i. Determine PA

j. Determine aorta

k. Determine right/left and anterior/posterior relationship of morphologic RV to morphologic LV

l. Evaluate ventricular septum

3. Apical

a. Orientation directed towards the left

b. Verify morphological RA, RV, LA, LV, atrioventricular arrangement and connections

c. Verify great arterial relationship

d. Verify left/right and anterior/posterior relationships of RA, RV, LA, LV

4. Subcostal cardiac views

a. Orientation marker for coronal views directed to the left

b. Orientation marker for sagittal views directed 90 degrees clockwise to from coronal views (caudal)

c. Verify atrioventricular arrangement and connections RA, RV, LA, LV

d. Verify great vessel arterial relationship

e. Verify left/right and anterior/posterior relationships of RA, RV, LA, LV

f. Evaluate atrial septum and ventricular septum

5. Subcostal abdominal views
   a. Orientation marker to the left for transverse views
   b. Orientation marker superior cephalad for sagittal views
   c. Identify IVC and follow course of IVC
   d. Evaluate for dilated veins posterior or lateral to the aorta which may indicate interruption of the IVC and dilated azygous and hemiazygous veins
6. Suprasternal notch views
   a. Orientation marker same as levocardia
   b. Orientation marker to left clavicle (1:00 – 2:00) for left arch
   c. Orientation marker to at 11:00 – 2:00 for right arch
   d. Orientation marker at 3:00 – 4:00 for suprasternal short axis arch same as levocardia
      i) Evaluate pulmonary venous return
      ii) Evaluate systemic venous return from superior brachiocephalic structures
      iii) Evaluate arch sidedness
      iv) Evaluate arch branching pattern using cephalad sweep

E. Anomalies of Venous Return
1. Anomalies of systemic venous return
   a. Types
      i) Left SVC draining into RA via coronary sinus
      ii) Left SVC draining into LA
      iii) Retroaortic innominate vein
      iv) Levoatrial cardinal vein
      v) Right SVC to LA
      vi) SVC aneurysm
      vii) Interruption of the IVC
      viii) Absence of hepatic segment with azygous vein continuation
      ix) Absence of the abdominal segment with hemiazygous continuation
      x) IVC draining into LA
      xi) Absence of right SVC
         - Absence of hepatic segment of IVC blood flows from IVC to azygous vein to left SVC to LA
         - Hepatic veins drain into LA
         - Common atrium
   b. Etiology
      i) Prevalence
         - Isolated anomalies occur in 0.3-0.5% of general population
         - Occurs in 2-10% of patients with other forms of CHD
         - Occurs in >70% of patients with heterotaxy syndrome
      ii) Abnormalities in regression of umbilical, vitelline, or cardinal venous segments during embryologic development

iii) Abnormalities of visceroatrial development; bilateral "sidedness" (heterotaxy) syndrome

c. Pathophysiology

i) Normal physiology of systemic venous return to the RA

- Left SVC to an intact coronary sinus with intact right SVC
- Left SVC to an intact coronary sinus with atresia of the right SVC
- Interrupted inferior vena cava with azygous continuation
- Absent RSVC with LSVC to CS

ii) Abnormal physiology with systemic and pulmonary venous mixing

- Left SVC draining into LA
- Right SVC draining into LA
- LSVC to an "unroofed" coronary sinus
- IVC draining into LA

d. Clinical presentation (pediatric and adult patients)

i) Left SVC to the coronary sinus and interrupted IVC usually present with additional structural cardiac defects but can be incidental findings

ii) Cyanosis is present with left/right SVC or IVC draining directly to LA

e. Associated lesions

i) Coarctation of the aorta

ii) Mitral valve abnormalities including congenital MS

iii) Heterotaxy syndrome with polysplenia

iv) HLHS

f. Echocardiographic evaluation (pediatric and adult patients)

i) Subcostal transverse and sagittal views of the abdominal viscera to image IVC/hepatic veins

ii) Suprasternal and high right/left parasternal views to image superior central venous anatomy

iii) Dilation of the coronary sinus in parasternal long and short axis views and in apical views may indicate drainage of left SVC into the coronary sinus

g. Common treatment (surgical repair)

i) No repair necessary if systemic venous return is physiologically normal

ii) Intra-atrial baffle repair of left/right SVC or IVC to LA

iii) Ligation of left SVC draining to LA if bilateral SVC with adequate "bridging" innominate vein

h. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluation of patency of repaired venous structures

ii) Evaluation for baffle leaks with baffle procedures

iii) Assessment of ligated venous structures

2. Anomalies of pulmonary venous return

a. Types

i) Normal pulmonary venous connections with abnormal drainage

- Extreme posterior (leftward) malinsertion of the atrial septum primum

ii) Partial anomalous pulmonary venous return (PAPVR)

- Drainage of one or more pulmonary veins anomalously to the systemic venous system or RA with normal drainage of remaining veins to the LA
  - Best defined in suprasternal/high parasternal (right and left) sagittal and axial views and in subcostal views
- Scimitar syndrome; right pulmonary veins drain infracardiac to the hepatic system or IVC
  - Best defined in suprasternal/high parasternal (right and left) sagittal and axial views as well as subcostal coronal and sagittal views

iii) Total anomalous pulmonary venous return (TAPVR)

- Supracardiac (Type 1)
  - Pulmonary venous drainage into an anomalous vertical vein and connecting superiorly to left innominate vein or SVC
  - Echocardiogram findings may demonstrate an enlarged innominate vein, SVC, RA, RV, and main pulmonary artery
- Intracardiac (Type 2)
  - Pulmonary venous directly to the posterior aspect of the RA
  - Pulmonary venous drainage into the coronary sinus
  - Demonstrated in subcostal coronal and sagittal images
- Infracardiac (Type 3)
  - Pulmonary venous drainage into a vertical vein connecting inferiorly to the portal venous system, ductus venosus, hepatic veins, or IVC
  - Evaluate for vertical vein obstruction
  - Best demonstrated in subcostal sagittal images
- Mixed type (Type 4)
  - Combination of anomalous sites of pulmonary venous return
  - Identify individual veins and their course
  - Best views are suprasternal or high parasternal coronal

b. Etiology

i) Incidence

- PAPVR: up to 25% of population may have a single right or left upper vein draining anomalously
- TAPVR: 9 in 100,000 of population

ii) Occurs mostly sporadically

iii) Failure of embryologic incorporation of one or more pulmonary veins to the LA

iv) PAPVR can be associated with Turner and Noonan syndromes

v) TAPVR can be associated with cat-eye, Holt-Oram and asplenia syndromes

c. Pathophysiology

i) Cyanosis due to mixing of systemic and pulmonary venous blood, frequently with pulmonary edema (when there is associated obstruction)

ii) Pulmonary overcirculation and congestion

iii) Pulmonary hypertension

d. Clinical presentation (pediatric and adult patients)

i) Tachypnea/dyspnea
ii) Cyanosis
iii) CHF (TAPVR)
iv) "Snowman" sign on x-ray (type 1 TAPVR)
v) "Scimitar" sign on x-ray (scimitar syndrome)

e. Associated lesions
i) HLHS
ii) Heterotaxy syndrome with asplenia
- Coarctation of the aorta

iii) Echocardiographic assessment (pediatric and adult patients)
iv) Define pulmonary venous return; trace anomalous pulmonary vein(s) to their point of termination (PAPVR)
- Look for a confluence of all four pulmonary veins posterior to the LA (TAPVR)
- Venous venophasic flow going away from the heart (superiorly or inferiorly) in a vertical vein
  - Evaluate for obstruction along anomalous drainage pathway using color and spectral Doppler

f. Common treatment (surgical repair)
i) Direct anastomosis of pulmonary vein confluence to LA (TAPVR types 1 and 3)
ii) Unroofing of coronary sinus with closure of interatrial communication (TAPVR type 2 to the CS)
iii) Intra-atrial baffle repair of pulmonary veins (PAPVR to RA and scimitar syndrome)

g. Echocardiographic assessment postoperatively (pediatric and adult patients)
i) Evaluate for flow obstruction at anastomosis of pulmonary veins to LA, pulmonary vein orifices and pulmonary veins
ii) Evaluate for residual vertical vein (in type 1 or type 3 cases)
iii) Evaluate for pulmonary venous baffle obstruction, if baffle-type repair
iv) TEE may be indicated for diagnostic quality imaging in adult patients

F. Anomalies of the Atria

1. Atrial septal defect (ASD)

a. Types:
i) Patent foramen ovale (PFO)
ii) Secundum ASD
iii) Primum ASD
iv) Sinus venosus (superior and inferior vena caval) defect
v) Coronary sinus septal defect

b. Etiology
i) Defects of the atrial septum occur in 1:1000 of population
ii) Patent foramen ovale found in 25% of population
iii) Primum and secundum type ASDs make up 67% of all ASDs
iv) Patent foramen ovale
- Overlapping of septum primum and septum secundum

- Located in the fossa ovalis region

v) Secundum ASD; mostly sporadic causes

- Most common adult congenital heart defect
- Defect within the fossa ovalis associated with deficient septum primum
- Usually due to single or multiple defects in septum primum
- Septum secundum usually well-formed

vi) Primum ASD

- Failure of septum primum to fuse with endocardial septation of the atrioventricular canal above the level of atrioventricular valves
- Located between the inferior septum primum and atrioventricular valves (AVV) atrial septum

vii) Sinus venosus defect

- Superior vena caval (SVC)
  - Approximately 8% of all ASDs
  - Sinus venosus not properly incorporated into the RA
  - Deficient wall between the SVC and right upper pulmonary
  - Vein
  - SVC overrides RA and LA
  - Located in superior and posterior portion of atrial septum
  - Associated with partial anomalous pulmonary venous return (PAPVR)
- Inferior vena caval (IVC)
  - Deficient wall between IVC and right lower pulmonary vein
  - Biatrial relationship of IVC to RA and LA
  - Located in inferodorsal portion of inferior limbic septum

viii) Coronary sinus (CS) septal defect

- Rare
- Complete or partial unroofing of CS
- Located between the inferior limbic septum, pars atrioventricularis of membranous septum and IVC orifice
- Associated with persistent left superior vena cava (LSVC) drainage to the coronary sinus (Raghib syndrome)

c. Pathophysiology

i) Interatrial communication caused by a defect in the atrial septum

ii) Initially left to right shunting (due to right ventricular compliance being less than left ventricular compliance), prolonged large-volume shunting can result in right to left shunting with elevated right sided pressures and Eisenmenger's physiology (rare with ASDs)

iii) Conditions that increase left to right shunting through ASD

- Left ventricular hypertrophy
- Cardiomyopathy
- Mitral stenosis
- Mitral regurgitation

- Mitral valve atresia
- Cor-triatriatum sinister
- Aortic stenosis
- Coarctation of the aorta

iv) Volume overload

v) Pressure overload

vi) Clinical presentation (pediatric and adult patients)

- May be symptom free for many years
- Shortness of breath (SOB) on exertion
- Recurrent respiratory infections
- Atrial arrhythmia (especially atrial fibrillation)
- Systolic murmur secondary to increased flow across the PV (relative pulmonary stenosis)through PV
- Large shunts may have diastolic murmur through tricuspid valve (TV)
- May have wide fixed-split second heart sound

d. Associated lesions

i) Secundum ASD and PFO

- Mitral valve prolapse
- Tricuspid atresia
- Hypoplastic left heart syndrome

ii) Primum ASD

- Atrioventricular canal defect
- Cleft mitral valve
- AVV regurgitation

iii) Sinus venosus defect

- Anomalous pulmonary venous return
- Right upper and lower pulmonary veins to SVC/RA/LA junction

e. Echocardiographic features (pediatric and adult patients)

i) Deficiency in atrial septal tissue

- Type of defect
- Size of defect

ii) Deficiencies in remaining atrial septal tissue “rims”

iii) Evaluate for systemic and pulmonary venous anomalies

- Note RV function
- Enlarged right atrium (RA)
- Enlarged right ventricle (RV)
- Evaluate for volume overload
  - Evaluate for diastolic ventricular septal flattening
- Evaluate for pressure overload
- Evaluate for systolic ventricular septal flattening

- Dilated TV annulus over time
- Enlarged pulmonary artery (PA)
- Enlarged pulmonic annulus over time
- Color flow Doppler (shunt directional information)
- Spectral Doppler (PW) assist with information about shunt directional information
- M-mode
  - Enlarged RV
  - Paradoxical septal motion

f. Echocardiographic evaluation (pediatric and adult patients)

i) Secundum defects best seen in the subcostal coronal view

- Shunt flow parallel to plane of insonation for optimal assessment
- Atrial septum perpendicular to plane of insonation minimizing dropout

ii) Subcostal sagittal view for optimal imaging of secundum and sinus venosus defects

iii) Apical 4-chamber view

- Septum primum defects
- AVV morphology and function in relation to ASDs
- Slightly oblique (foreshortened and more medial) apical 4-chamber projection may provide better definition of secundum ASD in adult patients with suboptimal image quality

iv) Subcostal 4-chamber (coronal) view

- Secundum defects
- Primum defects
- Sinus venosus defect - sweep posterior

v) Subcostal short-axis view at cardiac base

- Enface view of AVV

vi) Subcostal sagittal view of IVC/SVC

- Sinus venosus ASD
- Coronary sinus defects – seen very posteriorly and inferiorly

vii) Right parasternal sagittal view of atrial septum and SVC/IVC

- Sinus venosus ASD
- Presence of associated PAPVR and location of pulmonary venous connection to SVC
- Inferior-type ASD (including inferior sinus venosus)
- Secundum ASD

viii) Right parasternal axial (short axis) view

- Secundum ASD
- Retro-aortic tissue “rim”
- Presence and location of anomalous drainage of right pulmonary veins

ix) Color flow Doppler

- Useful to detect shunt direction and assist with size evaluation

- Evaluate for valvular insufficiency
- Assist with evaluation to differentiate partial AV canal from complete canal defect types

x) Pulsed-wave Doppler

- Interatrial pressure gradient
  - Useful for estimating LA pressure

xi) May need agitated saline (microcavitation) imaging

- Useful to detect transient right to left shunt through PFO or ASD
- Useful to evaluate for negative contrast in the RA with left to right shunt

xii) TEE may be indicated

- Evaluate type of defect (i.e. especially sinus venosus type)
- Evaluate for possible associated malformations (i.e., PAPVR, LSVC)
- Monitor intraprocedural progress and guidance for device closure

g. Common treatment (pediatric and adult patients)

i) Catheterization

ii) Device closure (only for select cases of secundum ASD)

iii) Surgical (all types)

- Primary (suture) closure
  - Smaller defects
- Patch repair
- Baffle-type repair (sinus venosus ASD with PAPVR)
- Warden repair of sinus venosus ASD; translocation of SVC with anastomosis to the right atrial appendage, baffle closure of ASD to SVC

iv) Echocardiographic assessment postoperatively (pediatric and adult patients)

- Evaluate for residual shunt in multiple views
- Evaluate for atrioventricular valve regurgitation
- If device closure, evaluate for device impingement on surrounding structures in multiple views
- If sinus venosus ASD repair, evaluate for SVC or pulmonary venous channel obstruction in subcostal and right parasternal sagittal views

2. Cor triatriatum (sinister)

a. Types

i) Obstructive

ii) Non-obstructive

b. Etiology

i) Occurs 3:100,000 of the population

ii) Failure of complete absorption of the pulmonary venous plexus confluence to the left atrium

iii) Results in Abnormal membrane that subdivides the left atrium into two chambers (pulmonary venous atrium and “true” LA) often resulting in an obstruction to pulmonary venous inflow

c. Pathophysiology

i) Supravalvular stenosis results in enlargement of the left atrium and possible dilation of pulmonary veins

ii) If an ASD is present, the chamber may not be dilated, as the ASD assists with decompressing chamber pressure

iii) Obstruction results in pulmonary venous and pulmonary arterial hypertension

d. Clinical presentation

i) Mild cases; incidental finding (patient may be asymptomatic)

ii) Pulmonary congestion on x-ray

iii) Shortness of breath/dyspnea

e. Associated lesions

i) ASD

ii) LSVC to the coronary sinus

iii) HLHS

f. Echocardiographic evaluation (pediatric and adult patients)

i) Parasternal long axis view defines membrane presence and point of pulmonary venous channel egress

- Membrane may stretch obliquely bisecting LA into pulmonary venous (dorsal) and "true" LA chambers.

ii) Apical 4-chamber view best aligns Doppler interrogation of fenestration gradient if present

iii) Subcostal views (coronal and sagittal), ASD may be present

iv) Evaluate for presence of right heart enlargement

- Evaluate for signs of pulmonary hypertension

v) Common treatment

- No treatment necessary if membrane non-obstructive
- Surgical resection of obstructive membrane with ASD closure

vi) Echocardiographic assessment postoperatively (pediatric and adult patients)

- Assess for residual obstructive membrane
- Assess atrial septum for residual defect

3. Supravalvular mitral stenosing ring

a. Etiology

i) No definitive data on frequency of occurrence exists

ii) Membrane located between the LA and the MV either just above the MV or in the funnel of the MV leaflets

iii) 90% of occurrences have associated left heart obstructive lesions

b. Pathophysiology

i) Membrane causing flow disturbances can result in damage to MV leaflets

ii) Significant obstruction can results in elevated LA pressure especially if no ASD

iii) Varying degrees of mitral valve regurgitation usually present

c. Clinical presentation

i) Pulmonary congestion on x-ray

ii) Cardiomegaly

iii) Shortness of breath/tachypnea

iv) Auscultation reveals diastolic rumble (additional systolic component with associated MR)

d. Associated lesions

i) Persistent LSVC

ii) VSD

iii) Coarctation of aorta (COA)

iv) ASD

v) Multiple left heart lesions (Shone complex)

e. Echocardiographic features (pediatric and adult patients)

i) Membrane seen in diastole when MV is open

- Attached at level of leaflets
- Attached at body of leaflets and stretched across MV funnel when valve is open

ii) MV leaflets appear thickened and/or myxomatous and may have limited excursion

f. Echocardiographic evaluation (pediatric and adult patients)

i) Multiple planes through MV evaluating for membrane when valve is open

ii) Measurement of mitral valve annulus in apical 4-chamber view

iii) Color Doppler helps define at which level of mitral inflow acceleration occurs

iv) Spectral Doppler

- Increased flow velocity at site of obstruction with elevated mean transvalvular gradient
- Flow profile may be normal if large ASD is present

g. Common treatment

i) Surgical resection of membrane

ii) Mitral valve repair or replacement if mitral valve significantly involved or damaged

h. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate for residual membrane

ii) Assess mitral inflow

iii) Presence and severity of MR

iv) Mitral leaflet perforations a rare complication

G. Atrioventricular Valve Anomalies

1. Atrioventricular (AV) canal defect

a. Etiology

i) Abnormal development of endocardial cushions

- 0.19:1000 live births
- 4.0-5.0% of all congenital heart defects (CHD)
- 40% of children with trisomy 21 and a CHD have an AV canal defect
- Failure of septum primum to fuse with endocardial cushions
- Located in the inferior anteriorly along the atrial septum
  - Categorized as incomplete (partial), complete or intermediate (transitional)

- Incomplete type consists of primum ASD, common atrium, cleft mitral valve (PAVC)
- Complete AV canal (CAVC) defects in the inferior portion of the atrial septum primum and inlet ventricular septum
- Intermediate (transitional) type AV canal consists of two complete AVV valve rings, primum ASD, restrictive posterior VSD

b. Pathophysiology
   i) Defect between the two atria and/or ventricles based on type of AV canal
   ii) Incomplete type same as ASD, complete type same as VSD
   iii) Volume overload
   iv) Pressure overload

c. Clinical presentation (pediatric and adult patients)
   i) Signs and symptoms
      - Incomplete (partial) type same as ASD
      - Complete type same as VSD
      - Small/restrictive shunts may be asymptomatic

d. Echocardiographic features (pediatric and adult patients)
   i) Defect located at the cardiac crux
      - Type of defect (complete, intermediate, incomplete)
      - Size of ventricles (balanced or unbalanced)
      - Note ventricular function and presence/degree of ventricular septal flattening
      - Estimate RV and PA pressure
      - May have varying degrees of AVV regurgitation
      - Color flow Doppler (directional information)
      - Left AVV component may be abnormal
        - Closely-spaced papillary muscles
        - Parachute-like attachments
        - Double-orifice left or right AVV component uncommonly occurs

e. Quantitation
   i) Estimate PA pressure by VSD gradient (if present) or right atrioventricular valve regurgitation jet
   ii) Visual evaluation of AVV regurgitation severity
   iii) Calculate left ventricular volume index if LV appears hypoplastic

f. Echocardiographic evaluation (pediatric and adult patients)
   i) Apical 4-chamber view
      - Optimal for the evaluation of atrial septum primum defects and atrioventricular valve commitment/attachments into the ventricles in CAVC defects
      - Optimal for evaluating the presence of associated LVOT using spectral Doppler
   ii) Subcostal 4 chamber view
      - Optimal for evaluating primum atrial septal defects and complete atrioventricular canal defects
   iii) Subcostal short axis view

- Enface view of common AVV
- Allows visualization of leaflet components, chordal attachments and atrioventricular valvular commitment to each ventricle

iv) Parasternal long axis view

- Allows for evaluation of presence of LVOT obstruction

v) Parasternal short axis view

- Additional imaging of the common AVV morphology

g. Evaluation of VSD component (if present)

i) Shunting lesion location

ii) Size of defect

iii) Shunt direction (left to right or right to left)

iv) Color flow Doppler

- Useful to detect shunt direction and assist with size evaluation
- Evaluate for AVV insufficiency
- Assist with evaluation to differentiate partial AV canal from complete canal

h. Related testing

i) ECG

- Counterclockwise frontal plane
- Left axis deviation
- Ventricular hypertrophy
  - Dependent upon volume of left-right shunt
  - Degree of PHTN
  - Degree of AV insufficiency

ii) Chest x-ray

- Small ASD-normal
- Large shunt – enlarged heart, increased pulmonary markings

iii) Cardiac catheterization

- Estimate right heart pressures in patients with pulmonary vascular obstructive disease

i. Common treatment (pediatric and adult patients)

i) Surgical

ii) Patch repair of ASD (and VSD, if present)

- "Australian" (single-patch) technique

iii) Suture repair of "cleft" in anterior leaflet of left atrioventricular valve (AVV)

iv) Replacement of left or right atrioventricular valve if significant stenosis or regurgitation is unrepairable

v) Resection or relief of subaortic obstruction, if present

j. Echocardiographic assessment of post-operative/repair evaluation (pediatric and adult patients)

i) Evaluate for residual shunt

ii) Evaluate for AVV stenosis and regurgitation

iii) Evaluate for PHTN
iv) Evaluate for AR
v) Evaluate for LVOT obstruction
vi) Evaluate ventricular function

2. Congenital mitral valve stenosis
   a. Types
      i) Mitral valve dysplasia with commissural fusion
      ii) Parachute mitral valve (PMV)
      iii) Double-orifice mitral valve (DOMV)
   b. Etiology
      i) Occurs rarely in isolation
      ii) Most cases associated with additional CHD
      iii) Caused by disruption and maldevelopment of left component of the endocardial cushions
      iv) Associated with hypoplasia of left heart structures
   c. Pathophysiology
      i) Mitral valve stenosis (PMV, DOMV)
      ii) Mitral valve may be regurgitant
      iii) Presence of ASD may cause underestimation of MS severity
      iv) Pulmonary hypertension
      v) Left atrial enlargement
      vi) Atrial dysrhythmias including atrial fibrillation and flutter (late)
      vii) Isolated DOMV may be clinically silent if not restrictive
   d. Clinical presentation (pediatric and adult patients)
      i) Pulmonary congestion on x-ray
      ii) Pulmonary edema
      iii) Cardiomegaly on x-ray
      iv) Shortness of breath/tachypnea
      v) Auscultation: diastolic rumble (additional systolic component with associated MR)
   e. Associated lesions
      i) ASD
      ii) Coarctation of the aorta
      iii) Subaortic stenosis
      iv) Bicuspid aortic valve
      v) Left SVC to a dilated coronary sinus
      vi) Left ventricular hypoplasia/HLHS
   f. Echocardiographic features (pediatric and adult patients)
      i) Increased inflow velocity through MV
      ii) May have varying degrees of mitral valve regurgitation
      iii) Left atrial enlargement
      iv) Right ventricular enlargement

v) Evidence of elevated pulmonary systolic pressure

g. Echocardiographic evaluation (pediatric and adult patients)

i) Image MV apparatus in PLAX view for assessment of excursion

ii) PSAX at MV leaflet tips to demonstrate effective orifice(s)

iii) PSAX at papillary muscles to define morphology chordal attachments

iv) Apical 4-chamber view for best angle of MV flow convergence and inflow interrogation (color and spectral Doppler)

v) For pediatric patients: CW Doppler of mean inflow gradient is used

vi) For adult patients: CW Doppler for pressure halftime and MVA

h. Common treatment

i) Open (surgical) commissurotomy

ii) Papillary muscle "splitting"

iii) Chordal fenestration

iv) Interchordal separation and reimplantation

v) Mitral valve replacement

i. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate for residual mitral valve stenosis or regurgitation

- Apical 4-chamber view is optimal for spectral Doppler assessment
- Mean inflow gradient should be measured in pediatric patients
- Mitral valve area should be calculated in adult patients

ii) Evaluate for valve prosthesis malfunction and presence of vegetations or thrombi in multiple views (patients with valve prostheses)

iii) Evaluate for residual ASD with multiple views in patients that have undergone ASD closure

iv) Assess ventricular size and function

3. Ebstein anomaly

a. Etiology

i) Malformation of the tricuspid valve with inferior apical and/or anterior displacement of the posterior and septal leaflets, (the anterior leaflet is usually large with abnormal attachments to the RV wall)

- 0.012:1000 live births
- Often associated with ASD
  - Shunt across ASD may be right to left and therefore patient may presents with cyanosis

b. Pathophysiology

i) Varying degrees of tricuspid regurgitation

ii) Systemic venous congestion with hepatomegaly

- May have a right to left shunting across an ASD causing central cyanosis and present cyanotic

c. Clinical presentation (adult and pediatric patients)

i) Mild cases may be clinically silent

ii) Extreme cases in neonatal form

- Cyanosis
- Cardiomegaly (wall-to-wall heart) on x-ray
- Tachypnea

iii) Split first and second heart sound

iv) Atrial dysrhythmias including SVT

d. Associated lesions

i) PFO or ASD (secundum), 30-78%

ii) Pulmonary valve stenosis

iii) Pulmonary valve atresia or "functional" atresia

- Approximately 30% will have Wolff-Parkinson-White syndrome

e. Echocardiographic features (pediatric and adult patients)

i) Abnormal TV leaflet attachments, may be associated with ASD, PFO, VSD, and PS

ii) Varying degrees of TV insufficiency

iii) RA size ("atrialized" RV)

iv) Note functional right ventricular function

v) Estimate RV and PA pressures

vi) May have paradoxical ventricular septal motion

vii) Spectral Doppler (evaluate tricuspid insufficiency and pulmonary flow)

viii) Evaluate for associated ASD/PFO

ix) M-mode

- TV closure delay
- Increased excursion of anterior TV leaflet

f. Echocardiographic evaluation (pediatric and adult patients)

i) Parasternal long axis view

- RV inflow view
- Evaluate RA size
- Evaluate tricuspid insufficiency
- Evaluate RV outflow view

ii) Parasternal short axis view

- Evaluate TV
- Evaluate degree of TV insufficiency
- Evaluate RVOT and PA
- Evaluate flow across pulmonic valve

iii) Apical view

- Evaluate RA and RV chamber size
- Evaluate placement of TV septal leaflet
- Evaluate TV for insufficiency and stenosis

iv) Subcostal views

- Coronal
  - Evaluate for presence of ASD and shunting pattern

- Septal TV leaflet displacement and attachments
- Extent of atrialized RV

- Sagittal (short axis)
  - RVOT involvement of TV (may have chordal insertions)
  - Effective RV size and function

v) Color flow Doppler
- Useful to evaluate TV insufficiency

vi) Spectral Doppler
- Evaluate TV insufficiency and pulmonic flow
- Evaluate TV for stenosis

g. Assessment for Ebstein
i) Degree of TV insufficiency
ii) RA size
iii) ASD/PFO presence
iv) RV function
v) Estimated PA pressure
vi) History of RV outflow tract or pulmonary valve obstruction

h. Related testing
i) ECG
- Tall p waves in limb lead II
- Right bundle branch block
- 30% will have Wolf-Parkinson-White syndrome

ii) Chest x-ray
- Cardiomegaly with prominent right sided border
- Decreased pulmonary blood flow

i. Common treatment
i) Mild cases = no treatment necessary
ii) Blalock-Taussig shunt
iii) Surgical repair (Cone, Danielson, Carpentier procedures)
iv) Valve replacement

j. Echocardiographic assessment postoperatively (pediatric and adult patients)
i) Evaluate for residual TV insufficiency
ii) Evaluate for iatrogenic TV stenosis
iii) Evaluate for residual shunt of ASD if history of ASD repair
iv) Evaluate RV inflow outflow post repair (stenosis)
- Evaluate RV outflow and pulmonic flow obstruction
- Evaluate for aorta-pulmonary artery shunt if history of procedure

4. Tricuspid valve atresia
a. Etiology

i) Failure of right sided atrial venous valves to regression resulting in minimal or no communication between the right atrium and right ventricle

- 0.039-0.085:1000 live births
- 2.7% of all CHDs
- Initial survival is dependent upon interatrial communication
- May occur with normally related great vessels or transposition of great vessels
- May occur with a normal PA and large VSD or pulmonary stenosis and small VSD or pulmonary atresia
- By adulthood will have gone through surgical repair
- Fontan procedure

b. Pathophysiology

i) Atresia of the TV

- Entire systemic venous return goes though interatrial communication (PFO or ASD)

ii) Ventricular volume overloading due to both inlets are committed to one ventricle

- Ventricular volume overload is dependent upon amount of pulmonary flood flow
  - Dependent upon presence of pulmonic stenosis or surgical shunts
  - When ventricle enlarges, ventricular wall stress increases, compensatory hypertrophy ensues
  - Hypertrophy – impaired diastolic function
  - Subendocardial ischemia

iii) In transposed great arteries

- Aorta may arise from hypoplastic "rudimentary" RV
- Aorta and subaortic region may be hypoplastic
- Aortic arch may have hypoplasia, coarctation or complete interruption

iv) Staged palliation resulting in total cavopulmonary anastomosis

c. Clinical presentation

i) Signs and symptoms

- Cyanotic at birth
- Dependent upon relationship of great arteries, and/or presence of pulmonary atresia/stenosis and how lesions were surgically addressed
- Dependent upon repair surgery and function of anastomoses/conduits

d. Associated lesions

i) RV hypoplasia

ii) VSD

iii) Normally related great arteries

iv) Transposed great arteries

v) Pulmonary stenosis

vi) Pulmonary atresia

vii) Hypoplastic aortic valve and ascending aorta (TGA type)

viii) Aortic arch hypoplasia and/or coarctation (TGA type)

ix) Juxtaposition of the atrial appendages

x) LSVC to the coronary sinus

e. Echocardiographic features (pediatric and adult patients)

i) Absence of normal TV apparatus

ii) Hypoplasia of the right ventricle

iii) Dominant and dilated LV

iv) Interventricular communication (VSD)

v) May have varying degrees of pulmonary outflow obstruction/hypoplasia

vi) ASD or patent foramen ovale

vii) Know what initial findings were at birth and what type of surgical correction was performed

viii) May have juxtaposed atrial appendages (TGA type)

ix) May have Fontan baffle within RA (postoperative patients)

x) Fenestration jet may be present

xi) Older adult patients may have atriopulmonary ("classic") Fontan

xii) Right atrial enlargement

xiii) Possible right atrial mural thrombi

f. Echocardiographic evaluation (pediatric and adult patients)

i) Parasternal long axis view

- Examine ventricular septum for presence of VSD
- Confirm relationship of great vessels
- Position of outlet septum
- Evaluate coronary sinus size and presence of left SVC

ii) Parasternal short axis view

- Sweep the interventricular septum for evidence of VSDs
- Evaluate systolic function of LV
- Evaluate RVOT and pulmonary artery and branches

iii) Apical 4 chamber view

- Evaluate right ventricular inlet region
- Evaluate outlet septum with anterior angle
- Evaluate LVOT and arterial connection
- Evaluate mitral inflow and for any insufficiency
- Evaluate pulmonary vein inflow

iv) Subcostal view

- Evaluate atrial septum to ensure unrestricted R-L shunting
- Define SVC and IVC connections to RA

g. Common treatment

i) Staged palliation

- First stage
  - Cases with unrestricted pulmonary outflow: PA banding

- Cases with severe PS or pulmonary atresia: modified Blalock-Taussig (B-T) shunt;
- If aortic coarctation is present, may need arch repair
- If significant sub-aortic obstruction is present (TGA-type), may need Damus-Kaye-Stansel anastomosi

- Second stage
  - Bidirectional Glenn (SVC to RPA) anastomosis
  - Directs systemic venous flow from upper extremities into pulmonary arteries
  - Patient remains cyanotic; IVC flow continues to mix at atrial level
- Third stage
  - Modified Fontan procedure (fenestrated and non-fenestrated)
  - Directs systemic venous blood flow from IVC/hepatic veins to PA without going through a ventricle
  - Performed to reduce the degree of cyanosis

h. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate cavopulmonary anastomoses for evidence of obstruction

ii) Evaluate Fontan baffle for obstruction or thrombi

iii) Evaluate Fontan baffle fenestration (if present) for shunting pattern and mean gradient

iv) Evaluate for Fontan baffle leaks (lateral tunnel-type only)

v) Evaluate ventricular function

vi) Evaluate pulmonary artery for evidence of obstruction

vii) Evaluate pulmonary veins for obstruction (lateral tunnel Fontan baffle may impinge)

viii) Evaluate MV and aortic valve for obstruction

ix) Additional views

- Conduits or anastomoses from the RA to the PA are usually seen anteromedially and superior to the atrium
  - Glenn shunt seen in suprasternal short-axis view
    - ~ Evaluate for an obstruction
    - ~ Evaluate with color and spectral Doppler
  - Fontan cavopulmonary anastomosis imaged in high parasternal and subcostal views
    - ~ Intra-atrial conduits can be imaged in parasternal and apical views
    - ~ Variations of Fontan (classic Fontan, extracardiac Fontan, fenestrated Fontan, lateral tunnel, RA-RV Fontan, and total cavo-pulmonary connection (TCPC)

5. Hypoplastic left heart syndrome

a. Etiology

i) Aortic valve atresia/hypoplasia, mitral valve atresia/hypoplasia and LV hypoplasia

- 0.103 -0.267:1000 live births
- Often associated with coarctation of aorta
- May have small atrial septal defect or intact atrial septum (poor prognosis)

b. Pathophysiology

i) "Ductal-dependent" lesion; ductus arteriosus patency must be maintained (PGE1 infusion, ductal stenting)

ii) Entire cardiac output is dedicated to right heart and routed through the PA/ductus with retrograde perfusion of the aortic arch

iii) Surgical procedure must be performed within the first week of life to create a permanent unobstructed pathway from the RV to the systemic circulation (stage 1 Norwood procedure)

iv) Pulmonary pressures must be regulated through an aorto-pulmonary shunt (stage I Norwood procedure) or a systemic venous – pulmonary artery shunt (bidirectional Glenn procedure or Fontan procedure)

v) Unobstructed interatrial communication must be maintained

vi) Staged palliation resulting in total cavopulmonary anastomosis

c. Clinical presentation

i) Pediatric patients in the neonatal period

- Varying degrees of cyanosis at birth
- At 3-5 days post birth, develop profound metabolic acidosis due to closing ductus
- Tachypnea
- Shock

ii) Palliated patients with complications (infant to adult)

- Evidence of CHF (right ventricular dysfunction/severe TR)
- Tachypnea
- Diminished distal pulses with upper/lower extremity pressure differential (aortic re-coarctation)
- Pleural effusions
- Ascites (Fontan patients with protein-losing enteropathy)

iii) Echocardiographic features (pediatric and adult patients)

- Single, dominant right ventricle
- Small LA
- Mitral valve hypoplasia or atresia
- Aortic valve hypoplasia or atresia
- Varying degrees of left ventricular hypoplasia
- May present with no definitive LV cavity
- LV may have endomyocardial fibroelastosis (EFE)
- Varying degrees of ascending and aortic arch hypoplasia
- Coarctation of the aorta

d. Echocardiographic evaluation (newborn/unpalliated patients)

i) Parasternal long axis view

- Evaluate for presence of TR
- Evaluate degree of RVH and assess RV function
- Evaluate for antegrade flow through aortic and mitral valves

ii) Parasternal short axis view

- Evaluate RV size and function
- Evaluate LV size and function
- Evaluate coronary artery origins and arrangement
- Evaluate PA branches

iii) Apical view

- Evaluate chamber sizes
- Evaluate left heart structures
  - Evaluate length and contractility of LV
  - Evaluate mitral valve morphology, annular size and function
  - Evaluate for presence and degree of antegrade aortic valve flow
  - Evaluate for presence of EFE
- Evaluate for presence/severity of TV insufficiency
- Evaluate RV function (qualitative assessment)
- Examine LV cavity with low-scale color Doppler for presence of coronary artery fistulae
- Examine pulmonary venous inflow

iv) Subcostal view (coronal and sagittal)

- Evaluate presence and degree of atrial-level shunting
- Measure mean transatrial gradient
- Evaluate systemic venous anatomy and drainage
- Assess ventricular function

v) Suprasternal notch view (long and short axis)

- Aortic arch anatomy, size and site(s) of obstruction
- Flow throughout arch

vi) Pulmonary venous anatomy and flow pattern

- Systemic venous anatomy and flow pattern
- Presence of abnormal venous structures (levoatrial cardinal veins, LSVC, anomalous pulmonary veins, etc.)

e. Common treatment

i) Staged surgical repair

- "Hybrid" procedure; stenting of the ductus arteriosus with individual flow-restricting bands on each of the PA branches
- Norwood (Stage 1) involving anastomosis of the proximal main PA to the ascending aorta, aortic arch reconstruction, systemic-to- pulmonary artery shunt, and atrial septectomy
- Glenn (superior cavopulmonary anastomosis)
- Fontan (inferior cavopulmonary baffle/conduit)
- Some patients may need tricuspid/neoaortic valve replacement or repair at some stage
- Heart transplantation

f. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate patency of shunts/conduits (stage 1)

ii) Evaluate neoaortic arch for obstruction

iii) Evaluate interatrial communication patency (must be widely patent)

iv) Evaluate for TR

v) Evaluate ventricular function

vi) Evaluate systemic inflow and outflow for obstruction

vii) Evaluate cavopulmonary anastomoses/conduits to ensure patency

viii) Tailor exam to evaluate the surgical repair

H. Ventricular Anomalies

1. Ventricular septal defect (VSD)

a. Etiologies

i) Perimembranous (infracristal, conoventricular)

- Most common type of VSD
- Failure of membranous portion of interventricular septum (IVS) to develop
- Incomplete closure of interventricular foramen
- Failure of subendocardial tissue to grow from right side of endocardial cushion and fuse with aorticopulmonary septum and muscular part of interventricular septum (IVS)
- Located in membranous/perimembranous area of IVS , in the LVOT just below the aortic valve
- Smaller/restrictive defects associated with aortic valve prolapse/AI

ii) Posterior/inlet muscular (canal-type, endocardial cushion–type, AV septum–type, inlet, juxtatricuspid)

- Deficiency of inlet portion of ventricular septum
- Marked discrepancy between the inlet and outlet dimensions of ventricular mass
- Seen with AV canal defects
- Located in the inflow portion of the IVS
- Isolated defects associated with straddling MV/TV

iii) Trabecular (muscular)

- Excessive cavitation of myocardial tissue during formation of IVS and ventricular walls

iv) Subarterial (supracristal, conal septal, infundibular, subpulmonic, subarterial, subarterial doubly committed, outlet)

- Located beneath the pulmonic and aortic valves and communicate with the RV outflow tract above the supraventricular crest and are associated with aortic regurgitation secondary to the prolapse of the right aortic cusp
- May have varying degrees of conal septal hypoplasia or aplasia

v) Malalignment (conal septal malalignment)

- Anterior malalignment
  - Associated with tetralogy of Fallot, pulmonary atresia; VSD type, truncus arteriosus
  - Conal septum malaligned anteriorly creating RVOT obstruction

- Varying degrees of associated pulmonary valve hypoplasia
- Posterior malalignment
  - Posterior displacement of conal septum causing LVOT obstruction
  - Associated with muscular aortic stenosis and/or aortic arch anomalies (arch interruption)
  - Associated with RVOT obstruction

b. Pathophysiology

i) Hole in ventricular septum

ii) Left to right shunt occurs when pulmonary vascular resistance is less than systemic vascular resistance

iii) Increased pulmonic flow

iv) May have decreased systemic CO2 due to left to right shunt

v) LA enlargement

vi) LV volume overload

- LVH
- Elevated LVEDP
  - Elevated LA pressure
  - Elevated pulmonary venous pressure
- Enlarged LV

c. Clinical presentation (pediatric and adult patients)

i) Signs and symptoms

- Small defects (restrictive)
  - May have no hemodynamic disturbance
  - High velocity jet with loud murmur
  - Subaortic defects associated with aortic valve prolapse and varying degrees of AI
- Large defects (unrestrictive)
  - Cardiomegaly on x-ray
  - Congestive heart failure
  - Low-frequency pansystolic murmur at lower left sternal edge or no systolic murmur
- Pulmonary hypertension (late, with large-volume shunts)
  - Eisenmenger's syndrome

d. Echocardiographic features (pediatric and adult patients)

i) Defect in ventricular septum

ii) Left heart enlargement with large, unrestrictive defects

iii) Color flow Doppler demonstrating direction of shunt and to estimate size

iv) Increased pulmonic flow

v) Evaluate RV size and function

vi) Evaluate for PHTN

vii) Estimate RV pressure by measuring gradient

viii) Enlarged LA and LV

e. Echocardiographic assessment (pediatric and adult patients)

i) Goals for VSD assessment

- Location
- Size (largest dimension)
- Shunt direction (i.e. left to right or right to left)

f. Echocardiographic views

i) Off-axis parasternal long axis views to align parallel to flow

- Color Doppler parallel to flow
- Sweep ventricular septum with color

ii) Parasternal short axis views

- Color Doppler parallel to flow
- Sweep the septum to look for defect
- Doppler PA and evaluate for pulmonary hypertension
- Subarterial VSDs will be seen at the level of the aortic valve in the 1 o'clock position
- Perimembranous VSDs will be seen at the level of the aortic valve in the 9 -12 o'clock position
- Muscular VSDs will be seen below the level of the aortic valve and in the muscular septum

iii) Apical views

- Evaluate LA and LV size and LV function
- Evaluate for presence of MR or TR
- In 5-chamber, evaluate for possible aortic valve prolapse and associated aortic valve insufficiency

g. Common treatment

i) Small (restrictive) VSDs

- May not require surgical closure unless subarterial or aortic valve prolapse is present
- Primary (pledgetted suture) closure

ii) Large (unrestricted) VSDs

- Pericardial or synthetic patch closure
- Intraventricular baffle-type for DORV
- If Eisenmenger syndrome present, VSD should not be closed

h. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate for residual shunt

ii) Evaluate for additional (undetected) VSDs

iii) Evaluate for AI and/or TR (associated with perimembranous VSD repair)

iv) Evaluate for pulmonary hypertension

v) Assess LVOT/RVOT for obstruction

2. Ventricular inversion (Congenitally or physiologically corrected transposition)

a. Etiology
   i) Leftward bulboventricular looping with abnormal development of the truncal septum
      - Left atrium connects to morphologic right ventricle (on left side of heart) which connects to the aorta
      - Right atrium connects to morphologic left ventricle (on right side of heart) which connects to the PA
   ii) Associated lesions
      - Perimembranous VSD
      - Ebstein anomaly of the morphologic (systemic) TV
      - RVOTO (subpulmonary stenosis)
      - Congenital complete heart block

b. Pathophysiology
   i) Normal hemodynamic physiology
   ii) Morphologic RV handles systemic cardiac output load
   iii) Morphologic LV handles pulmonary cardiac output load
   iv) RV remodels to accommodate systemic pressure load
   v) LV atrophies under pulmonary pressure load
   vi) Ebsteinoid TV can lead to malcoaptation and regurgitation
   vii) Severe volume load on RV (TR) can cause CHF in infancy
   viii) Patients with complete heart block have higher mortality
   ix) RV dysfunction may occur later in life

c. Clinical presentation (pediatric and adult patients)
   i) May be clinically silent until 3rd or 4th decade of life if no associated CHD (VSD, subPS, heart block, etc.)
   ii) CHF caused by TR and/or systemic (right) ventricular failure
   iii) Development of complete heart block
   iv) Severe progressive pulmonary stenosis (in cases with PS)

d. Echocardiographic features (pediatric and adult patients)
   i) Apical 4 chamber view demonstrates attachment of left sided AV (tricuspid) valve slightly apical to right sided AV (mitral) valve
   ii) Parasternal short axis view demonstrates leftward and anterior displacement of the aorta (L-transposition of the great vessels)
   iii) Morphologic RV is the systemic ventricle
      - To evaluate RV function dp/dt and Tei index may help

e. Echocardiographic evaluation (pediatric and adult patients)
   i) Parasternal long axis view
      - Demonstrate features of left-sided morphologic RV
      - Image septal attachments of morphologic TV
      - Confirm ventriculo-infundibular fold separating TV and AoV
      - Assess outflow tracts for obstruction
   ii) Parasternal short axis view

- Confirm ventricular morphology
- Evaluate ventricular sizes and systolic function
- Evaluate ventricular septal configuration
  - Assess for VSDs using 2D and color Doppler
- Evaluate mitral and tricuspid valve morphology, attachments and function

iii) Apical view

- Confirm ventricular arrangement
- Evaluate ventricular sizes and function
- Evaluate tricuspid valve morphology, function and competency
- Evaluate outflow tracts

iv) Subcostal view

- Coronal (4-chamber)
  - Look for atrial level defect
  - Evaluate LVOT for obstruction
  - Evaluate for VSDs
- Sagittal (short axis)
  - Evaluate ventricular function
  - Evaluate ventricular septal configuration

f. Common treatment

i) No treatment necessary for asymptomatic cases with no additional CHD

ii) VSD closure for cases with moderate to large unrestrictive VSD

iii) Relief of pulmonary/subpulmonary stenosis if present

iv) PA banding; patients with Ebsteinoid TV and significant TR

- Re-configures IVS to encourage better TV leaflet coaptation, effectively decreasing TR

v) Pacemaker insertion, if complete heart block is present or develops

vi) "Double-switch" operation

- Senning or Mustard (atrial switch) operation
- Jatene (arterial switch) operation
- Re-routes blood flow correctly; LV receives pulmonary venous return and pumps to aorta, RV receives systemic venous return and pumps to PA

g. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate systemic ventricular (RV) function (all patients)

ii) Evaluate for residual VSD (patients with VSD repair)

iii) Assess presence and degree of TR

iv) Assess arterial connections for obstruction (patients with double-switch operation)

v) Assess atrial baffles for obstruction/leaks (patients with double switch operation)

3. Single ventricle (functionally univentricular heart)

a. Types

i) Atrioventricular connections

- Absent right
- Double inlet
- Single (common) inlet
- Absent left

ii) Ventricular morphology

- Dominant LV
  - Left-sided LV
  - Right-sided LV
- Dominant RV
  - Right-sided RV
  - Left-sided RV
- Indeterminate morphology

iii) Arterial segment

- Normally related arteries
- Transposed/malposed
  - Aortic atresia

b. Etiology

i) Occurs in 1-2% of all CHD

ii) Most cases occur sporadically

iii) Some associated with genetic syndromes:

- Holt-Oram
- Noonan
- Velocardiofacial
- Trisomy 21 (rarely)

c. Pathophysiology

i) Most cases cyanotic at birth

ii) Systemic and pulmonary outputs determined by:

- Presence and degree of hypoplasia/narrowing of great arteries
- Presence of outflow tract/semilunar valve obstruction
- Patency of interatrial communication in the presence of absent or obstructed AV connection(s)

d. Clinical presentation (pediatric and adult patients)

i) Cases with unobstructed pulmonary outflow present with CHF

ii) CHF exacerbated with presence of significant atrioventricular valve regurgitation

iii) Cases with pulmonary stenosis or atresia cyanotic at birth

iv) Patients ranging from preschool-age to adult may present with signs of CHF due to deterioration of ventricular function

e. Echocardiographic features

i) Single dominant ventricle

ii) Rudimentary or hypoplastic secondary ventricle/outlet chamber

iii) May have malposed great arteries

f. Echocardiographic evaluation

i) Evaluate atrial communication

ii) Evaluate AV connections

iii) Assess ventricular morphology and function

iv) Evaluate VA connections for obstruction

- If significant PS or pulmonary atresia, ensure ductal patency

v) Evaluate arterial relationship

vi) Evaluate aortic arch for coarctation

g. Common treatment

i) Staged palliation necessary:

- Hearts with significant PS or pulmonary atresia
  - Aorta to pulmonary shunt; Glenn anastomosis; Fontan
- Hearts with unobstructed pulmonary outflow
  - PA banding; Glenn anastomosis; Fontan
- Hearts with severe aortic outflow obstruction/aortic atresia
  - Damus-Kaye-Stansel anastomosis and arch reconstruction (if needed); aorta to pulmonary shunt; Glenn anastomosis; Fontan
  - May have "Hybrid" operation; stenting of PDA with banding of individual PA branches

h. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate single ventricular function

ii) Evaluate aortic/semilunar outflow for obstruction or regurgitation

iii) Evaluate aortic arch (if arch reconstruction or coarctation repair performed)

iv) Evaluate AVV flow and presence of regurgitation

v) Evaluate pulmonary blood flow

- Sano (RV to PA) conduit from parasternal SAX
- Modified Blalock-Taussig (subclavian artery to RPA/LPA) shunt from suprasternal
- Glenn (SVC to RPA/LPA) anastomosis from suprasternal or high parasternal
- Fontan baffle patency and presence of fenestration (best imaged in parasternal SAX and apical views)

I. Semilunar valve anomalies

1. Aortic stenosis

a. Types and etiology

i) Subvalvular tunnel

- Hypoplastic LVOT

ii) Associated with hypertrophic cardiomyopathy

- Crowded LVOT with systolic anterior motion of MV chordae and apparatus
- May be isolated mid-cavitary

iii) Discrete fibromuscular subaortic membrane

- Associated with:
  - VSD or spontaneously closed VSD
  - Shone's complex
- Fibrous or fibromuscular tissue in LVOT
- May extend to anterior mitral valve leaflet
- Circumferential or partial (within LVOT)

iv) Associated with bicuspid aortic valve

- Incidence 2% of population
- Associated with COA
- Congenitally malformed leaflets
- Thickened leaflets
- Fused raphe
- Purely bicuspid (bi-leaflet with no raphe)

v) Congenitally stenotic tri-leaflet valve

- Three leaflets
- Varying degrees of commissural fusion

vi) Unicuspid

- Congenitally malformed leaflets
- Single slit-like commissure in cross-section, usually at the commissure between the left and non-coronary leaflet

vii) Quadricuspid

- Congenitally malformed leaflets
  - Varying degrees of commissural fusion
- Rare; Incidence 0.013%

viii) Supravalvular

- Narrowing of ascending aorta
- Can occur sporadically or familial (defect in elastin gene on chromosome 7)
- Associated with William's syndrome

b. Pathophysiology

i) Increased resistance to flow out of the left ventricle

ii) Concentric LV hypertrophy (LVH) develops

- Calculation of LV mass assists with evaluating the LV work load

iii) LA enlargement may also occur in patients with LVH

- Associated with abnormal LV diastolic function

c. Clinical presentation

i) Signs and symptoms

- More common in males than females
- Dependent upon severity (may be asymptomatic)
- Severity is measured on the pressure drop across the valve in systole
- Syncope (rare)

- Critical AS
  - Low cardiac output
  - Reduced ejection fraction
  - Loud systolic murmur
  - Congestive heart failure
  - Shock
  - Dependent on a patent ductus arteriosus

d. Echocardiographic features
   i) Thickened leaflets
   ii) Doming of leaflets with restriction of excursion
   iii) Concentric LVH
   iv) LV diastolic function
   - May have diastolic "fluttering" of anterior MV leaflet if significant aortic insufficiency is present

   v) Doppler pressure gradient
   vi) Ascending aortic dilation
   vii) Evaluate for subvalvular stenosis
   viii) Evaluate for supravalvular stenosis
   - Calculate Z-scores for the aortic annulus ad root, sinotubular junction, and ascending aorta

e. Echocardiographic evaluation (pediatric and adult patients)
   i) Parasternal long-axis view
   - Evaluate valve excursion
   - Color Doppler evaluate turbulent flow (level) and evaluate for AI

   ii) Parasternal short axis view
   - Evaluate number of cusps
   - Color Doppler of valve for regurgitation
   - LV function
   - Presence and degree of LVH

   iii) Apical 5 chamber view
   - Evaluate Doppler flow through LVOT and AV (subaortic stenosis/membrane, valvar AS)
   - Color Doppler evaluation for point of acceleration and presence/severity of regurgitation

   iv) Pedoff transducer (Adolescents to adults)
   - Right parasternal (valve and supravalve AS)
   - Suprasternal notch (valve and supravalve AS)
   - Apical 5-chamber (subvalve AS)

   v) Color flow Doppler
   - Evaluate level of stenosis
   - Evaluate for valvar insufficiency

- Assess for presence of associated VSD

vi) High right parasternal sagittal view

- Color Doppler demonstrates high pressure gradient of aortic/supravalvular flow
- Spectral (CW) Doppler of aortic outflow (Valvar and supravalvular AS)

f. Common treatment

i) Balloon dilation

ii) Surgical valvotomy

iii) Ross procedure (pulmonary autograft to aortic position, RV to PA allograft)

iv) Surgical valve replacement

g. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate transvalvular pressure gradient

ii) Evaluate for AI (perivalvular AI in patients with AVR)

iii) Evaluate for prosthesis stenosis/failure

iv) Evaluate LV function

v) Assess ascending aorta and arch

vi) Evaluate RV to PA connection and PA branches (Ross patients)

2. Pulmonary stenosis

a. Types and etiology

i) Pulmonary valve stenosis

- Bicommissural pulmonary valve
- Tri-leaflet valve with cusp fusion
- Valvular stenosis with reflexive infundibular hypertrophy

ii) Hypoplastic pulmonary valve annulus and conus

- Thickened domed and stenotic valve
- Narrow or hypoplastic valve annulus

iii) Supravalvular

- Discrete supravalvular membrane
- "Hourglass" deformity in main PA segment
- Branch pulmonary artery (PA) stenosis

iv) Subvalvular pulmonary stenosis

- Prominent muscle bundle at the infundibular os creating subvalvular obstruction
- Also known as a double-chamber right ventricle

b. Pathophysiology

i) Increased resistance to flow out of the right ventricle

ii) Obstruction results in increased RV systolic pressure

iii) Afterload increase results in RV hypertrophy and muscle mass

c. Clinical presentation

i) May be asymptomatic

ii) Systolic murmur

iii) Critical PS

- Cyanosis

d. Echocardiographic features
   i) Thickened leaflets
   ii) Doming of leaflets
   iii) RVH
   iv) Doppler pressure gradient
   v) Evaluate for infundibular stenosis

e. Echocardiographic evaluation (pediatric and adult patients)
   i) Parasternal long axis view
      - Evaluate RV outflow
      - Color Doppler evaluate turbulent flow (level) and evaluate for PI
      - Measure valve annular diameter
   ii) Parasternal short axis view
      - Evaluate RV outflow, pulmonic valve, pulmonary artery and branches
      - Measure valve annular diameter
      - Color Doppler
        - Evaluate for turbulence
        - Evaluate for PI
      - Spectral Doppler
        - Estimate mean pressure gradient
   iii) Subcostal view
      - RVOT and midcavitary RV
      - Measure valve annular diameter
      - Color Doppler RVOT, pulmonic valve, PA
      - Spectral Doppler RVOT, pulmonic valve, PA

f. Common treatment (pediatric and adult patients)
   i) Balloon dilation (bicuspid PV, commissural fusion)
   ii) Surgical valvotomy/commissurotomy
   iii) Transannular patch (annular hypoplasia)
   iv) Non-transannular patch (RVOTO, infundibular hypoplasia)
   v) Pulmonary valve replacement (adolescents and adults)

g. Post-operative/repair evaluation
   i) Evaluate trans-valvular pressure gradient
   ii) Evaluate for residual PS and presence/severity of PI
   iii) Assess for residual RVOT obstruction
   iv) Evaluate RV size and function
   v) Assess TR
   vi) TAPSE for measure of RV function

3. Pulmonary atresia with intact ventricular septum (PA/IVS)
   a. Etiology

i) Environmental factors

ii) Flow disturbances through the right heart during embryologic and fetal development

iii) Luminal discontinuity between the RVOT and Pulmonary artery without a VSD

- Two morphologic types
  - Hypoplastic RV, TV, RVH, coronary artery fistulae/sinusoids
  - Dilated thin walled RV, severe TR, massive RA

b. Pathophysiology

i) Entire systemic venous return crosses PFO, which is usually unrestricted. Mixing in LA causes cyanosis in newborn

ii) Pulmonary blood flow is dependent on the ductus arteriosus, making this a ductal dependent lesion

iii) Suprasystemic RV pressures with and without coronary sinusoids can result in myocardial ischemia

iv) PA flow is ductal dependent at birth unless MAPCAs (major aorto-pulmonary collateral arteries) are present (unusual with this disease)

c. Clinical presentation

i) Cyanosis at birth

ii) May present with CHF, hepatomegaly in subtype with massive TR and right heart enlargement

iii) Adolescents and adults may present with CHF due to poor LV function and development of MR

iv) Myocardial infarction may occur due to presence of RV dependent coronary circulation (RVDCC)

d. Echocardiographic features

i) Absent or diminished RVOT with no flow demonstrated from RV to PA

ii) Varying degrees of TV/RV hypoplasia usually present

iii) Coronary sinusoids/fistulae to the RV may be present

iv) PDA commonly originates from the underside of the arch with a tortuous path to pulmonary arteries

v) Collaterals or MAPCAs (Major Aorto-Pulmonary Collateral Arteries) are rare in this (PA/IVS) subtype

e. Echocardiographic evaluation

i) Assess RV and TV, using lower scale color flow imaging of RV cavity for sinusoids

ii) Assess RVOT for antegrade flow

iii) Assess branch PAs for continuity and discrepant flow

iv) Assess TV for TR

v) Assess atrial shunt flow

vi) Assess PDA

vii) Assess coronary artery for anatomy and flow

viii) Assess LV size and function for regional wall motion abnormalities

f. Common treatment

i) Staged approach needed in most cases

- Ductal stenting (stage 1)

- Systemic-to-pulmonary (Blalock-Taussig) shunt with PDA ligation (stage 1)
- Glenn: superior cavopulmonary anastomosis (stage 2)
- Fontan: inferior cavopulmonary baffle (stage 3)

ii) PV perforation and balloon dilation possible in cases with adequate TV and RV size and function

iii) Heart transplantation may be necessary in cases with RVDCC

g. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Assess left ventricular function

ii) Evaluate atrial shunting

iii) Assess presence and severity of MR

iv) Evaluate pulmonary blood flow source

- BT shunt (if present)
- Glenn anastomosis (if performed)
- Fontan baffle and mean fenestration gradient (if Fontan and/or fenestration present)
- RVOT outflow and degree of regurgitation (if valvotomy performed)

J. Aortic Arch Anomalies

1. Coarctation of the aorta

a. Types

i) Pre-ductal (arch hypoplasia)

ii) Ductal (discrete "shelf")

iii) Post-ductal and "mid-aortic"

b. Etiology

i) Luminal narrowing of the aorta from the normal diameter (usually isthmus)

ii) Flow disturbances though the left heart during embryologic and fetal development

iii) Environmental factors

iv) Occurs in 5-7% of all CHD

v) Associated lesions

- Shone's complex
  - Bicuspid aortic valve
  - Left SVC to the coronary sinus
  - Mitral valve abnormalities
- TGA

vi) Syndromic associations

- Turner syndrome
- 22q11 deletion (DiGeorge syndrome)

c. Pathophysiology

i) Narrowing of aorta at level of coarctation

ii) Increased resistance to flow at the coarctation site with drop in pressure distally

iii) Usually see a difference in blood pressure between arms and legs

iv) Collateralization occurs late, decompressing proximal arch segment

d. Clinical presentation

i) Signs and symptoms at birth

- Dependent upon severity of lesion
- Metabolic acidosis and circulatory shock due to ductal closure
- Ductal closure results in decreased flow to abdominal aorta and abdominal organs
- Increased resistance to flow through region of coarctation
  - Increases work of LV
  - Increased LVEDP
  - Pulmonary edema

ii) Signs and symptoms in adolescence and adulthood

- Asymptomatic until adolescence
- Heart murmur
- Systemic hypertension
- LVH on ECG during adult physical exam
- Repaired patients may present with symptoms of re-coarctation

iii) Echocardiographic features

- Narrowing in aortic arch
  - Pre-ductal; arch hypoplasia with juxtaductal coarctation shelf
  - Ductal; normal arch dimensions with discrete narrowing, usually in juxtaductal region of isthmus
  - Post-ductal; discrete or long-segment narrowing past ductal region of thoracic aorta (may also be abdominal)
- Post-stenotic dilation may be present
- Increased velocity seen on spectral Doppler in area of coarctation
- Doppler waveforms in abdominal aorta may be blunted with low velocity and continuous antegrade diastolic flow
- Color Doppler will reveal turbulent flow in stenotic region
- LVH
- LV function may be decreased
- Signs of elevated LVEDP
  - LAE
  - Monophasic MV inflow pattern

e. Echocardiographic evaluation (pediatric and adult patients)

i) Evaluate arch in SSN long axis for site of obstruction

- Measure arch dimensions; segmental diameters

ii) Evaluate arch for abnormal branching pattern

iii) Color flow Doppler

- Evaluate arch for turbulence

iv) Spectral Doppler

- Evaluate for increased velocities in the region of coarctation

- Use PW(proximal) and CW Doppler (obstruction site) to evaluate net arch gradient

f. Common treatment

i) Surgical repair

- End-to-end anastomosis (discrete)
- Extended end-to-end anastomosis (isthmus hypoplasia)
- Forward/reverse subclavian patch technique (pre-ductal arch hypoplasia)
- Patch augmentation (older technique rarely used currently)

ii) Balloon dilation with or without stent placement (adolescents and adults)

g. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate for recurrence or residual coarctation

ii) Assess for dilation or aneurysm formation (especially adults with history of patch augmentation)

iii) Assess ventricular function

iv) Evaluate for un-masked AV or MV obstructive lesions

2. Interrupted aortic arch

a. Types

i) Type A (30%); discontinuity between last vessel (usually left subclavian artery) and aortic isthmus

ii) Type B (70%); discontinuity between left carotid and left subclavian arteries

iii) Type C (<1%); discontinuity between carotid arteries or between the left carotid artery and the right innominate (brachiocephalic) artery

iv) All types may have aberrant left or right subclavian arteries

b. Etiology

i) Abnormal regression of arch segments during embryology

ii) Environmental factors may play role

iii) Highly associated with 22q11 deletion (DiGeorge syndrome)

iv) Associated defects

- Bicuspid aortic valve
- Posterior malalignment VSD
- Subaortic stenosis
- Left SVC to a dilated coronary sinus
- Mitral valve abnormalities
- HLHS

c. Pathophysiology

i) Interruption of aorta with ductal dependent circulation to post-ductal end-organs

ii) Ascending aorta provides coronary and cerebral perfusion

iii) VSD (posterior malalignment-type) is almost always present

iv) Ductal closure causes circulatory shock

d. Clinical presentation

i) Cyanosis as newborn

ii) Metabolic acidosis and circulatory shock due to ductal closure

iii) Ductal closure results in cessation of flow to abdominal aorta and abdominal organs

iv) Murmur

v) Repaired patients may present with symptoms of re-coarctation

e. Echocardiographic features

i) Discontinuity between ascending and descending aortic segments

ii) Large posterior malalignment VSD usually present

iii) Small, usually bicuspid aortic valve

iv) Hypoplastic ascending aorta

f. Echocardiographic evaluation

i) Parasternal long axis view to evaluate aortic valve annulus size and presence and degree of subaortic narrowing

ii) Parasternal short axis to evaluate aortic valve morphology

iii) Suprasternal or high right parasternal LAX view for long axis arch imaging and ascending aorta flow profile

iv) Suprasternal short axis to evaluate arch sidedness and type of interruption (depending on branching pattern)

v) Apical views to evaluate flow through subaortic region

g. Common treatment

i) Aortic arch repair

- Excision of ductal tissue with end-to-side arch anastomosis
- Interposition graft less commonly used

ii) VSD closure

iii) Resection of obstructive sub-aortic conus tissue

iv) Ross-Konno repair for significant aortic annular hypoplasia

v) Patients with severe subaortic stenosis and/or significant aortic valve hypoplasia may need Norwood-type arch reconstruction with DKS and Rastelli-type VSD baffle with RV to PA homograft

h. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate arch for residual obstruction

ii) Assess LVOT for obstruction

iii) Assess for residual VSD(s)

iv) Evaluate ventricular function

v) Assess pulmonary artery connection (homograft/conduit) in cases with Norwood/Rastelli type repairs

3. Vascular rings and slings

a. Types

i) Double aortic arch

ii) Right aortic arch with aberrant left subclavian artery

iii) Circumflex (left to right) aortic arch

iv) Cervical arch variants

v) Pulmonary artery sling

b. Etiology
   i) Most cases have unknown etiology
   ii) Abnormal regression of arch segments during embryologic development
   iii) Normal intracardiac anatomy in majority of cases
   iv) Chromosome 22q11 deletion found in a small percentage of cases

c. Pathophysiology
   i) Abnormal arch and PA vasculature causes encirclement and compression of trachea and esophagus
   ii) Ligamentum arteriosum completes "ring" in cases of right aortic arch with aberrant left subclavian artery
   iii) Hemodynamically benign
   iv) Tracheal and esophageal compression
   v) Complete tracheal rings present in 50-60% of pulmonary sling patients
   vi) Most cases cause dysphagia

d. Clinical presentation
   i) Stridor and wheezing
   ii) Esophageal compression with dysphagia
   iii) Recurrent upper respiratory infections (URI)
   iv) Exacerbation of URI symptoms
   v) Gastroesophageal reflux with reflex apnea

e. Echocardiographic features
   i) Right aortic arch dominance in most cases of double aortic arch
   ii) Right aortic arch with aberrant left subclavian artery in cases of arch-type vascular ring
   iii) Left pulmonary artery arising from the distal RPA in pulmonary artery sling-type
   iv) Usually normal intracardiac anatomy

f. Echocardiographic evaluation
   i) SSN short axis views to evaluate arch morphology and branching pattern in cases of double arch and right arch with aberrant left subclavian artery
   ii) SSN long axis views to assess left or right arch dominance in double arch cases
   iii) Parasternal short axis view to assess PA anatomy in cases of pulmonary artery sling

g. Common treatment
   i) Division of non-dominant arch in cases of double aortic arch
   ii) Division of ligamentum arteriosum in cases of right aortic arch with aberrant left subclavian artery
   iii) Division and reimplantation of the LPA anterior to the trachea in cases of pulmonary artery sling

h. Echocardiographic assessment postoperatively (pediatric and adult patients)
   i) Evaluate pulmonary arteries for patency in cases of pulmonary artery sling repair
   ii) Assess aortic arch for obstruction
   iii) Imaging the diaphragm helpful postoperatively if suspicion for paralysis exists

4. Patent ductus arteriosus (PDA)

a. Etiology
   i) Failure of the ductus arteriosus to close under hormonal influences after birth
   ii) Associated with prematurity
   iii) Associated with trisomy 21
b. Pathophysiology
   i) Left to right shunt from aorta to pulmonary artery
   ii) Increased blood volume flowing to lungs
   iii) Increased blood flow returning to the left heart
      - Enlarged LA
      - Enlarged LV
      - Cardiac failure
c. Clinical signs and symptoms
   i) Cardiac failure with large shunt
   ii) Failure to thrive
   iii) Small lesions may have no symptoms
   iv) Continuous murmur with accented systolic component
      - Machinery murmur
   v) Differential cyanosis
      - Difference in cyanosis/hypoxemia between right and left arms and lower extremities based on right to left shunt in patients with suprasystemic PA pressures
d. Echocardiography findings
   i) Patent ductus arteriosus communicating from aorta to pulmonary artery
   ii) Color flow Doppler demonstrates communication from aorta to pulmonary artery
   iii) Enlarged main pulmonary artery and branches
   iv) Enlarged LA
   v) LV hypertrophy
   vi) Enlarged LV
e. Echocardiographic evaluation (pediatric and adult patients)
   i) High parasternal short axis view
   ii) Parasternal short axis view of branch pulmonary arteries
   iii) Parasternal long axis aortic arch to ductus view
   iv) Utilize color flow Doppler to visualize and estimate size and direction of shunt in each view above
   v) Utilize spectral Doppler to demonstrate Doppler waveform and velocities to calculate pulmonary pressure
f. Common treatment
   i) Newborn period – ductal closure promoted by use of prostaglandin antagonists
   ii) Surgical ligation and division
g. Surgical or minimally invasive clip ligation
   i) Percutaneously closed using catheter-delivered coils or occlude device

h. Echocardiographic assessment postoperatively (pediatric and adult patients)
   i) Evaluate for residual PDA
   ii) Assess surrounding structures (LPA, RPA, descending aorta) for obstruction
      - Uncommonly, inadvertent ligation of LPA or descending aorta has been described
      - Occluder device/coil may impinge upon surrounding vascular structures
   iii) In cases of occluder device, ensure device is well-positioned
   iv) Assess ventricular function

K. Conotruncal Anomalies
   1. Transposition of the great arteries
      a. Types
         i) Dextro-transposition of the great arteries (D-TGA) – isolated ventriculo-arterial discordance
         ii) Congenitally corrected Transposition of the great arteries (CCTGA) – atrio-ventricular and ventriculo-arterial discordance; (covered in ventricular anomalies)
      b. Etiology of D-TGA
         i) Abnormal conotruncal septation resulting in ventriculo-arterial discordance
         ii) Higher incidence in maternal IDM
         iii) Environmental factors may play a role
         iv) 10th most common CHD
         v) 2nd most common cyanotic lesion
      c. Pathophysiology of D-TGA
         i) Two parallel circuits exist
         ii) Systemic venous blood returns to the RV and then is recirculated to the systemic circulation through the aorta
         iii) Pulmonic venous blood returns to the LV and then is recirculated to the pulmonary circulation through the unrepairable pulmonary artery
      d. Clinical presentation D-TGA
         i) Cyanotic at birth
         ii) Single loud second heart sound
         iii) VSD/LVOTO variants will have flow murmur
         iv) Coarctation variants may have significant metabolic acidosis
      e. Associated Lesions
         i) VSD in 40-45 % of patients
         ii) LVOTO in 20-30%
         iii) Aortic coarctation
         iv) Systemic venous anomalies
         v) Juxtaposition of the atrial appendages
      f. Echocardiographic features
         i) Ventriculo-arterial discordance
         ii) Fibrous continuity between the mitral and pulmonary valve
         iii) RV infundibulum is located between the aorta and the TV

iv) Coronary artery anatomy has a variable pattern

v) Ventricular configuration appears normal in the neonatal period

vi) Systemic morphologic RV (Mustard and Senning patients) becomes enlarged with septal flattening

g. Echocardiographic evaluation D-TGA (pediatric and adult patients)

i) Assess the abnormal ventriculo-arterial connections and arterial configuration

ii) Subcostal coronal and sagittal sweeps for determination of the relational anatomy

iii) Parasternal long axis view for semilunar annular sizes

iv) Parasternal short axis view for semilunar valve morphology, coronary artery origins

v) Parasternal short axis sweeps of the ventricular septum for VSDs

vi) Subcostal views to evaluate if restrictive atrial septal shunting is present

h. Common treatment

i) Balloon atrial septostomy

- Immediate palliation for extreme cyanosis; improves atrial level shunting

ii) Surgical repair

- Jatene arterial switch operation (ASO) with LeCompte maneuver
- Rastelli operation (D-TGA; DORV-type)
- Mustard or Senning (atrial switch) operation
  - Commonly used prior to era of successful arterial switch operations

i. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Jatene ASO with LeCompte maneuver

- Evaluate arterial anastomosis sites for obstruction
- Evaluate branch pulmonary arteries for patency (branch pulmonary arterial stenosis is a common complication)
- Evaluate coronary artery anastomoses using color Doppler
- Evaluate semilunar valves for stenosis and/or regurgitation
- Assess ventricular function

ii) Rastelli

- Evaluate VSD baffle for residual shunts
- Evaluate LV to aorta pathway
- Assess RV to PA connection
- Assess AV valve competency
- Evaluate ventricular function

iii) Mustard or Senning

- Evaluate atrial baffles for leaks or obstruction
- Assess ventricular function
- Evaluate for LVOTO (occasionally caused by ventricular septal bowing into LVOT)
- Assess TV function and presence of regurgitation

2. Tetralogy of Fallot (TOF)

a. Types

i) TOF with varying degrees of pulmonary stenosis
ii) TOF with pulmonary atresia
iii) TOF with absent pulmonary valve syndrome (APVS)
iv) TOF with complete atrioventricular canal defect (CAVC)

b. Etiology
   i) Occurs in 4-8% of all CHD
   ii) Embryologic maldevelopment of the subpulmonary conus
   iii) Results in narrowing of the infundibulum and an anterior malalignment VSD
   iv) Associated defects:
      - Right aortic arch
      - Vascular ring
      - Coronary artery anomalies
      - Systemic venous anomalies
      - Pulmonary artery discontinuity
   v) Syndromic associations:
      - 22q11 deletion (DiGeorge syndrome)
      - Holt-Oram
      - Trisomy 13, 18, 21
      - Alagille syndrome
      - Goldenhar syndrome

c. Pathophysiology
   i) Combination of obstructive pulmonary outflow and large VSD result in ventricular-level right-to-left shunt and cyanosis
   ii) Degree of cyanosis directly related to severity of pulmonary outflow obstruction
   iii) Prolonged cyanosis causes polycythemia and fingertip clubbing
   iv) Ductal patency improves pulmonary blood flow but may contribute to pulmonary over-circulation in cases with mild pulmonary outflow obstruction

d. Clinical presentation
   i) Varying degrees of cyanosis
      - Significant pulmonary outflow obstruction increases ventricular right-to-left shunt, resulting in increased cyanosis
      - Cases of TOF with APVS usually have massively dilated PA branches causing airway compression, exacerbating cyanosis
   ii) May present with over-circulation and signs of CHF (“pink TOF”)
   iii) Murmur
      - Harsh systolic murmur for typical cases
      - Harsh systolic and diastolic murmurs in TOF with absent PV
   iv) Abnormal x-ray (boot-shaped heart)
   v) Repaired adult patients may present with signs and symptoms of right heart failure from pulmonary regurgitation or residual outflow obstruction
   vi) Repaired adult patients frequently experience inter-ventricular conduction delays and ventricular dysrhythmia

vii) Unrepaired adolescent or adult patients present with profound cyanosis, polycythemia and fingertip clubbing

e. Echocardiographic features
   i) Parasternal long axis; classic features of aortic override of large anterior malalignment VSD
   ii) Right ventricular hypertrophy
   iii) Short axis view demonstrates anteriorly deviated conal septum with infundibular narrowing
   iv) Varying degrees of PV annular hypoplasia
   v) Cases of TOF with absent pulmonary valve may have right heart enlargement due to significant associated pulmonary regurgitation
   vi) Abnormalities of aortic arch sidedness and branching may be present

f. Echocardiographic evaluation
   i) Assess degree of outflow tract obstruction in parasternal long and short axis views
      - Measure pulmonary valve annulus
      - Measure main PA and PA branches
      - Assess for pulmonary artery continuity
   ii) Assess degree of aortic override of the VSD in PLAX view
   iii) Assess for coronary artery anomalies in PSAX view
   iv) Assess for aortic arch anomalies in SSN view
   v) Evaluate VSD shunting pattern in multiple views

g. Common treatment
   i) Repair dependent on anatomy
      - May need staged approach if anatomy unfavorable for repair
        - Blalock-Taussig shunt (BTS) early
        - Complete repair involving VSD closure and relief of PS at 6-months of age
   ii) Transannular-type patch repair
   iii) Non-transannular-type repair
   iv) Complete repair with RV to PA homograft/conduit
   v) Complete repair with plication of MPA and PA branches (TOF, absent PV)
   vi) Adolescent and adult patients may need revision of RV to PA connection
      - Stent placement(s) for residual obstructive RVOT and pulmonary artery lesions
      - Re-operative surgical pulmonary valve (re)placement
      - Percutaneous valve placements (Melody, Sapien)

h. Echocardiographic assessment postoperatively (pediatric and adult patients)
   i) Evaluate aorta-to-pulmonary shunt (BTS) flow using color Doppler in SSN view
   ii) Evaluate for residual VSD in multiple views (patients with complete repair)
   iii) Assess RVOT for residual obstruction and possible aneurysm formation in PLAX and PSAX views
   iv) Evaluate pulmonary valve for residual stenosis or regurgitation in PSAX and PLAX views (in cases of valve-sparing repair)
   v) Evaluate pulmonary arteries for residual narrowing or dilation in PSAX view

vi) Evaluate TV for disruption and regurgitation in PLAX and apical views (patients with complete repair)

vii) Assess ventricular function

- Particular attention to right ventricular function

3. Double outlet right ventricle (DORV)

a. Types

i) Categorized by the relationship of the great arteries, the conal morphology, and the location of the VSD

- Relationships of the great arteries
  - Aorta is to the right and posterior to the PA
  - Aorta is right and lateral to the PA
  - Aorta is right and anterior to the PA
  - Aorta is left and anterior
- VSD types
  - Subaortic (42-57%)
  - Subpulmonic (24-37%)
  - Doubly committed (3-12%)
  - Remote (9-19%)
- Conal morphology
  - Bilateral conus
  - Absent aortic conus
  - Absent subpulmonary conus
  - Bilaterally absent conus

b. Etiology

i) Maldevelopment of conotruncus resulting in both great arteries arising predominantly or entirely from the RV

ii) Occurs in 1-2% of all patients with CHD

iii) Associated syndromes (12%)

- Trisomy 13, 18
- 22q11 deletion (DiGeorge syndrome)

iv) Associated defects

- Subpulmonary or subaortic outflow tract obstruction
- Straddling atrioventricular valves (AVV)
- Atrioventricular canal defect (CAVC)
- Aortic arch anomalies
  - Coarctation
  - Arch branching malformations including vascular ring
- Systemic venous anomalies
- Juxtaposition of the atrial appendages

c. Pathophysiology

i) Highly variable depending on VSD size, location, outflow tract placement and patency and the streaming characteristics of blood flow
   - VSD physiology (pulmonary overcirculation)
   - TOF physiology (varying degrees of cyanosis)
   - TGA physiology (pulmonary overcirculation with suboptimal mixing)
   - Single ventricle physiology

d. Clinical presentation
   i) Newborn patients
      - Murmur
      - Varying degrees of cyanosis
   ii) Adolescent and adult patients post repair
      - Palliated patients present with cyanosis
      - Right heart failure
      - Ventricular dysrhythmia
      - CHF

e. Echocardiographic features
   i) All cases have malposed great artery relationship
   ii) One artery may override VSD
   iii) Subaortic or subpulmonary outflow obstruction
   iv) Frequently muscular discontinuity between AV valves and semilunar valves (bilateral conus)

f. Echocardiographic evaluation
   i) Evaluate great artery relationship in PLAX and PSAX views
   ii) Evaluate for outflow tract obstructions in multiple views
   iii) Presence and location of VSD(s) in multiple views
   iv) Assess ventricular morphology and sizes
   v) Evaluate semilunar valves for abnormalities
   vi) Assess aortic arch and pulmonary arteries
   vii) Evaluate coronary artery anatomy

g. Common treatment
   i) Complete repair:
      - Baffle closure of VSD connecting the LV to the nearest semilunar valve
      - Rastelli procedure
      - Nikaidoh procedure
      - Arterial switch operation with VSD closure
   ii) Staged single ventricle palliation for cases with:
      - Straddling atrioventricular valves
      - Remote VSD
      - Hypoplastic ventricles

h. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Evaluate great artery anastomosis sites for obstruction in multiple views

ii) Evaluate for residual VSD in multiple views (patients with complete repair)

iii) Assess outflow tracts for residual obstruction in PLAX and PSAX views

iv) Evaluate pulmonary arteries for residual narrowing or dilation in PSAX view

v) Evaluate TV for disruption and regurgitation in PLAX and apical views (patients with complete repair)

vi) Assess ventricular function

4. Truncus arteriosus

a. Types

i) Collett & Edwards classification

- Type I - A short main PA segment arises from the arterial trunk and gives rise to both branch Pas
- Type II - Both branch PAs arise from the arterial trunk adjacent to one another
- Type III –The branch PAs arise from either side of the arterial trunk, somewhat remote from each other
- Type IV – Collaterals supply the pulmonary circulation from the descending aorta; (previously known as pseudotruncus, it is now considered a variant of TOF with pulmonary atresia)

ii) Van Praagh classification

- Type I - A short main PA segment arises from the arterial trunk and gives rise to both branch Pas
- Type II – Both branch PAs arise from the arterial trunk through separate orifices
- Type III – A single branch PA arises from the ascending segment with the contralateral lung supplied by collaterals or anomalous origin of the contralateral branch
- Type IV – Truncus arteriosus with arch hypoplasia, coarctation or interruption
- Subtype A –VSD, B intact ventricular septum

b. Etiology

i) Failure of the truncus arteriosus to septate resulting in persistence of a single trunk that gives rise to the aorta, pulmonary arteries and coronary arteries.

ii) DiGeorge, velocardiofacial, and 22q11 deletion syndromes

iii) Occurs in 1-4% of patients with CHD

iv) Associated anomalies

- Right aortic arch (33%)
- Interrupted aortic arch, usually type B (19%)
- Truncal valve abnormalities (bicuspid, quadricuspid, stenotic, regurgitant)
- LSVC (12%)
- Anomalous origin of right or left subclavian artery
- Anomalous origin of a single branch pulmonary artery

c. Pathophysiology

i) Biventricular output to pulmonary and systemic arterial circulation

ii) Systemic and pulmonary vascular resistances dictate shunting

iii) As pulmonary vascular resistance drops, the pulmonary blood flow increases

iv) Pulmonary over-circulation occurs if left unrepaired

d. Clinical presentation

i) Tachypnea

ii) CHF

iii) Necrotizing enterocolitis in premature infants

iv) Murmur

e. Echocardiographic features

i) Unicuspid, bicuspid, tri-leaflet (most commonly) or quadricuspid truncal valve

ii) Commonly truncal valve stenosis (33%) and insufficiency (50%)

iii) Pulmonary artery branches arising from arterial trunk

iv) Large subarterial outlet-type VSD (usually)

v) Aortic arch anomalies

f. Echocardiographic evaluation

i) Assess truncal valve for morphology and function in PLAX, PSAX and subcostal sagittal and coronal views

ii) Assess origin and size of branch PAs, assessing flow using Doppler

iii) Assess LV size and function

iv) Assess coronary arteries for origin and course in PSAX, PLAX and subcostal views, relation to PA origins

v) Assess aortic arch for anatomy, laterality, and obstruction or interruption

vi) Assess origin of left and right carotid and subclavian arteries

vii) Assess for a PDA or aorto-pulmonary collaterals

viii) Assess VSD for shunt direction, size and timing

ix) Assess for additional VSDs

x) Assess for PFO/ASD

xi) MV, TV for straddling tissue

g. Common treatment

i) Complete surgical repair

- VSD closure
- Homograft or RV-PA Valved conduit connecting branch PAs to the RV

h. Echocardiographic assessment postoperatively (pediatric and adult patients)

i) Assess for residual VSD with sweeps in multiple views

ii) Assess conduit or homograft for stenosis, insufficiency in PLAX, PSAX

iii) Assess RVOT for aneurysm formation of the RVOT patch

iv) Assess distal anastomosis of conduit to branch PA for stenosis

v) Assess truncal valve for stenosis, insufficiency

vi) Assess ventricular function

vii) TR for RV pressures

L. Surgical Procedures

1. Alfieri

a. Edge-to-edge central "stitch" anastomosis of the mitral valve leaflets
b. Performed to control and minimize the severity of MR

2. Blalock-Taussig shunt (BTS)
   a. Provides supplemental pulmonary blood flow
   b. Performed for:
      i) Severe cases of tetralogy of Fallot
      ii) Double outlet right ventricle with severe PS
      iii) Pulmonary atresia
      iv) Hypoplastic left heart syndrome (as part of Norwood procedure)
      v) Single ventricle hearts with severe PS or pulmonary atresia
   c. Classic BTS
      i) Direct anastomosis of right or left subclavian artery to either pulmonary artery
      ii) No longer used due to subclavian artery enlargement over time with progressive pulmonary over-circulation
   d. Modified BTS
      i) Synthetic or biologic interposition tube (usually 3mm to 5mm in diameter) placed between a subclavian artery and a pulmonary artery
3. Carpentier procedure for Ebstein anomaly
   a. Longitudinal plication of "atrialized" RV and tricuspid annuloplasty
   b. Performed to minimize or alleviate TR
4. Cone procedure for Ebstein anomaly
   a. Extensive reconstruction of the tricuspid valve including:
      i) Detaching tethered septal leaflet
      ii) Creating an effective TV annular region
      iii) Re-suspending TV apparatus
   b. Plication of excessive atrialized RV
5. Damus-Kaye-Stansel
   a. Proximal main pulmonary artery and ascending aorta are joined in single ventricle cases with significant aortic or subaortic outflow obstruction
6. Danielson procedure for Ebstein anomaly
   a. Plication of "atrialized" RV
   b. TV annuloplasty or replacement
7. Fontan procedure
   a. Inferior cavo-pulmonary connection
   b. Routing of IVC blood flow to the pulmonary arteries in cases of single ventricle physiology
   c. Final stage for completion of single ventricle palliation
   d. Types
      i) "Classic" atrio-pulmonary connection (no longer performed):
         - Direct connection of RA to pulmonary arteries with PFO closure
      ii) Lateral tunnel: intra-atrial baffle
      iii) Extra-cardiac conduit

8. Glenn anastomosis (shunt)
    a. Superior cavo-pulmonary connection
        i) Routing of the SVC(s) blood flow to the pulmonary arteries in cases of single ventricle physiology
    b. Secondary stage for palliation for hearts with single ventricle physiology
    c. End-to-side anastomosis of the right SVC to the RPA; “bidirectional”
    d. If LSVC is present, an end-to-side anastomosis of the LSVC to the LPA is made
    e. Bilateral SVC will have bilateral anastomoses
9. Jatene operation
    a. Arterial switch operation (ASO) used for transposition of the great arteries
        i) Aorta and pulmonary arteries are transected
        ii) Ascending aorta is anastomosed to native pulmonary root
        iii) Pulmonary trunk is anastomosed to native aortic root
        iv) Coronary arteries are re-implanted to the root of the “neo-aorta”
10. Konno procedure
    a. Patch widening of the native LVOT in cases of significant LVOT hypoplasia
    b. In most cases, a VSD is created by incising a portion of the outlet ventricular septum
    c. A trans-annular incision is performed with placement of an aortic valve prosthesis to relieve aortic annular hypoplasia
    d. Some cases require a pacemaker postoperatively as surgical disruption of the conduction system leads to complete heart block
11. LeCompte maneuver
    a. A surgical method of positioning the branch pulmonary arteries anterior to the aorta
        i) Used during Jatene ASO when great artery relationship is anterior/posterior
    b. Pulmonary artery branches “straddle” the ascending aorta
        i) Used in rare cases of pulmonary artery compression of airways (TOF with APVS)
12. Mustard procedure
    a. Surgical procedure in which baffles are created using pericardial or synthetic material within the atria to “switch” pulmonary and systemic venous returns
    b. Previously used as primary repair in cases of D-transposition of the great arteries (no longer surgical technique of choice)
    c. Used for part of “double switch” operation in cases of CCTGA
13. Nikaidoh procedure
    a. Surgical reconfiguration of great artery arrangement in cases of DORV and PS with TGA physiology
    b. Relocates the native aorta closer to the LV for VSD closure
        i) The small pulmonary root is excised to create space for relocation of aortic root
        ii) The aorta is relocated posteriorly in the void created by removal of pulmonary root
        iii) LV outflow is routed through the RV to the aorta via a VSD baffle
        iv) Placement of RV to pulmonary artery homograft or conduit
        v) Procedure may include LeCompte maneuver of the pulmonary arteries
14. Norwood procedure

a. Surgical reconstruction of the aortic arch for cases of HLHS or single ventricle with aortic hypoplasia or atresia
    i) Pulmonary trunk is transected from the branch pulmonary arteries
    ii) Aortic arch is augmented using biologic patch material, coarctation shelf and ductal tissue are resected
    iii) Native ascending aorta and pulmonary trunk are combined (DKS) and anastomosed to the reconstructed "neo-aortic" arch
b. Systemic to pulmonary shunt is performed as secondary component of operation
    i) Modified BTS
    ii) Sano modified PA conduit
c. Atrial septectomy

15. Potts shunt
    a. Surgical aorto-pulmonary shunt by creating a fistula between the descending aorta to the LPA to increase pulmonary blood flow in cyanotic heart disease
    b. Currently used for pulmonary artery egress "pop-off" in cases of severe pulmonary hypertension and Eisenmenger syndrome
    c. No longer used as a primary palliation
16. Rastelli procedure
    a. Surgical repair of DORV with TGA
        i) LV outflow is routed through the VSD into the RV to the remote aorta via a baffle patch
        ii) Placement of RV to pulmonary artery homograft or conduit
        iii) Procedure may include LeCompte maneuver of the pulmonary arteries
17. Ross procedure
    a. Used for severe congenital aortic valve stenosis and/or regurgitation
        i) Surgical replacement of aortic root and valve with native pulmonary trunk (pulmonary autograft)
        ii) Placement of RV to pulmonary artery homograft or conduit
    b. May be used in conjunction with the Konno procedure (i.e., Ross-Konno)
18. Sano modified conduit
    a. Surgical placement of a synthetic or biologic conduit between a ventricle and the pulmonary artery bifurcation as a source of pulmonary blood flow
        i) Used in first stage of single-ventricle palliations
        ii) Conduit diameter ranges between 3mm and 5mm
19. Senning procedure
    a. Surgical procedure in which baffles are created using native atrial wall and atrial septal tissue within the atria to "switch" pulmonary and systemic venous returns
    b. Previously used as primary repair in cases of D-transposition of the great arteries (no longer surgical technique of choice)
    c. Used for part of "double switch" operation in cases of CCTGA
20. Warden procedure
    a. Surgical repair of sinus venosus ASD with PAPVR to the SVC
        i) The SVC is transected above the level of the anomalous pulmonary venous connection

ii) The SVC "stump" on the atrial side is sutured closed

iii) An intra-atrial baffle is placed from the SVC orifice to the ASD, effectively routing the anomalous pulmonary venous flow to the LA

iv) The SVC is then anastomosed anteriorly to the right atrial appendage

21. Waterston shunt
    a. Surgical aorto-pulmonary shunt by creating a fistula between the ascending aorta to the RPA to increase pulmonary blood flow in cyanotic heart disease
    b. No longer used as a primary palliation
22. Yacoub procedure
    a. Surgical valve-sparing replacement of the ascending aorta
        i) Used in cases of severely dilated or hypoplastic ascending aorta with normal aortic valve size, morphology and function
    b. A synthetic tube is used to replace the ascending aorta
    c. Coronary arteries are anastomosed directly to the tube in their typical locations

Section XV: Stress Echocardiography

1. Describe the purpose of stress echocardiography
2. Differentiate between the indications of exercise versus pharmacologic stress echocardiography
3. Name common pharmacological agents used in stress echocardiography
4. Identify the equipment necessary for stress echocardiographic examinations
5. Explain the protocol for stress echocardiography
6. Name the pre-echocardiographic and post-echocardiographic views used in stress testing
7. Identify wall motion abnormalities seen on a stress echocardiogram
8. Identify major coronary arteries associated with 16- and 17-segment perfusion model

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XV. Stress Echocardiography

- A. Definition
  - 1. Ischemia
    - a. Oxygen supply demand mismatch
  - 2. Sequence of ischemic response
    - a. Decrease perfusion
    - b. Decrease LV compliance
    - c. Increase LVEDP
    - d. Decrease LV contractility
    - e. ECG changes
    - f. Symptoms
  - 3. Indications
    - a. CAD screening
    - b. Assess perfusion pre- and post-revascularization
    - c. Determine prognosis
    - d. Evaluate valvular heart disease
  - 4. Contraindications
    - a. Aortic stenosis
    - b. Obstructive hypertrophic cardiomyopathy
- B. Types of Stress Echocardiography
  - 1. Exercise stress echocardiography
    - a. Patient preparation and ECG-lead placement
    - b. Indication
    - c. Relative contraindication
    - d. Stress testing system
      - i) Treadmill
      - ii) Bicycle
      - iii) Other
    - e. 12-lead ECG system
    - f. Blood pressure device
    - g. Emergency crash cart

h. End points
    i) Maximal exertion
    ii) Clinical ischemia
    iii) Blood pressure (hyper/hypotension)
    iv) Sustained tachycardia
    v) WMA involving two or more segments
    vi) Predicted heart rate

2. Pharmacologic stress echocardiography
    a. Patient preparation
    b. Indication
    c. Relative contraindication
    d. Infusion pump
    e. 12-lead ECG system
    f. Blood pressure device
    g. Emergency crash cart
    h. Pharmacological stress agents
        i) Effects on heart
    i. Atropine to achieve target heart rate
    j. End points
        i) Clinical ischemia
        ii) Blood pressure (hyper/hypotension)
        iii) Sustained tachycardia
        iv) Wall motion abnormality (WMA) involving two or more segments
        v) Predicted heart rate

3. Pacing

C. Clinical Applications for Stress Echocardiography
    1. Diagnose CAD
    2. Determine extent of CAD
    3. Evaluate LV function
    4. Screen preoperative and postoperative
    5. Assess valvular heart disease
    6. Evaluate myocardial viability

D. Corresponding Perfusion Zones of the Major Coronary Arteries
    1. LAD
    2. LCX
    3. ACA

E. Procedure
    1. Baseline images
        a. Parasternal long axis
        b. Parasternal short axis

c. Apical 4 chamber
d. Apical 2 chamber

2. Stress initiated
   a. Target heart rate
3. Post-exercise echocardiographic images
   a. Time limitations
   b. Patient position
   c. Time increments
4. Digital image acquisition
5. Tissue harmonic imaging
6. Contrast for LV opacification

F. Ischemic Response
1. Wall motion analysis
   a. Absence of hyperkinesis
   b. Normal function
   c. Hypokinetic
   d. Akinetic
   e. Dyskinetic
2. False negative
3. False positives

G. Advantages of Stress Echocardiography
1. Shorter imaging time
2. Lack of ionizing radiation
3. Portability
4. Immediate availability of results
5. Lower cost
6. Ancillary information
   a. Chamber size and function
   b. Valves
   c. Pericardial effusion
   d. Aortic root disease
   e. Wall thicknesses

H. Pitfalls

Section XVI: Transesophageal Echocardiography (TEE)

1. Describe the purpose and protocol of transesophageal echocardiography
2. Identify the equipment necessary for TEE
3. List common clinical indications to perform a TEE

XVI. Transesophageal Echocardiography (TEE)

- A. Transesophageal Echocardiography (TEE)
  - 1. Purpose
    - a. Unobstructed window
    - b. Higher resolution compared to TTE
  - 2. Indications
  - 3. Contraindications
  - 4. Procedure
    - a. Description
    - b. Equipment
    - c. Transducer types
    - d. Types of sedations
    - e. Patient preparation
    - f. Patient sedation
    - g. Laboratory set up /clean up
    - h. Transducer care
      - i) Cleaning
      - ii) Storage
  - 5. Advantages/limitations
  - 6. Complications
  - 7. Probe manipulation
    - a. Advance/withdraw
    - b. Rotation (0° - 180°)
    - c. Turn (right-left/medial-lateral)
    - d. Anteflex/retroflex
    - e. Flex right/left
- B. Imaging Views
  - 1. High esophageal - depth to include LV
    - a. 0° four-chamber – retroflex
      - i) LV size and function
      - ii) RV size and function
      - iii) LA and RA size, pulmonary veins
      - iv) Withdraw probe for LA appendage
      - v) MV and TV
      - vi) Anteflex to see aortic valve

b. ~60° two chamber
    i) LV size and function
    ii) LA and LA appendage
    iii) MV
c. ~120° long axis
    i) LV size and function
    ii) LA size
    iii) Mitral and aortic valve
    iv) Withdraw probe for ascending aorta

2. High esophageal – decrease depth to optimize valves
    a. ~120° long axis
        i) Mitral valve anatomy and function
        ii) Color Doppler for mitral regurgitation
        iii) Antegrade mitral flow
        iv) Aortic valve anatomy and flow
    b. ~60° two chamber
        i) Mitral valve anatomy and function
        ii) Color Doppler for mitral regurgitation
        iii) LA imaging and flow
    c. 0° four chamber
        i) Mitral valve anatomy and function
        ii) Anteflex for aortic valve anatomy and function
        iii) Atrial septum
3. Transgastric
    a. 0° short axis
        i) LV wall motion, thickness and dimensions
        ii) RV size and function
    b. ~90° long axis
        i) Mitral valve anatomy and function
        ii) Color Doppler for mitral regurgitation
        iii) LA imaging and flow
4. Transgastric apical
    a. 0° four chamber
        i) Anatomy and function
        ii) Antegrade aortic flow with color Doppler
5. Transgastric to high esophageal
    a. 0° short axis descending aorta
        i) Image aorta from diaphragm to aortic arch
6. Modify each view according to anatomy and pathology

Section XVII: Real Time 3-D Echocardiography

1. List the clinical indications for real time 3-D imaging echocardiography
2. Describe the equipment components necessary to perform real time 3-D imaging echocardiographic examinations
3. Describe advantages and limitations of real time 3-D imaging compared to 2-D echocardiography

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XVII. Real Time 3-D Echocardiography

- A. Imaging Methods and Display
  - 1. Image acquisition
    - a. 2-D reconstruction
      - i) Freehand
      - ii) Mechanical rotation
    - b. Volumetric imaging
      - i) Real time
      - ii) Volume acquisition
  - 2. Tissue depiction
    - a. Surface rendering
    - b. Volume rendering
- B. Clinical Indications
  - 1. Ventricular assessment
    - a. Function
    - b. Volume/mass
  - 2. Leaflet evaluation
    - a. MV
    - b. AV
  - 3. Congenital heart disease
  - 4. Surgical reconstruction
- C. Modality Comparison
  - 1. Advantages of real time 3-D imaging
  - 2. Limitations of real time 3-D imaging
  - 3. Advantages of 2-D
  - 4. Limitations of 2-D

Section XVIII: Enhanced Cardiac Ultrasound

1. Sequence the set-up and imaging procedure for administering contrast
2. List the various contrast agents used in the clinical setting
3. Differentiate between the types of contrast
4. Describe clinical indications and contraindications for the use of contrast
5. Explain appropriate equipment settings necessary to perform an optimal contrast study

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XVIII. Enhanced Cardiac Ultrasound

A. Use of Contrast Agents
   1. Transpulmonary agents
      a. Indications and contraindication
      b. Clinical applications
      c. Set up and supplies
   2. Agitated saline
      a. Indications and contraindication
      b. Clinical applications
      c. Set up and supplies
      d. IV insertion techniques
   3. Contrast media
      a. Bubble characteristics
         i) Size
         ii) Stability
         iii) Composition
         iv) Acoustic properties
            - Acoustic impedance
            - Resonant frequency
      b. Technical criteria
         i) Appropriate rate and concentration
            - Bolus
            - Diluted bolus
            - Continuous infusion
            - Set up and supplies
         ii) Image acquisition
            - Continuous
            - Sequential
            - Gated
         iii) Appropriate equipment settings
            - Mechanical index (MI)
            - Transducer frequency
            - Harmonics

- Dynamic range
- Receiver filter
- Transmit frequency spectrum

- Focal zone location
- Compression/dynamic range
- Gain

iv) Effects of inappropriate settings

v) Indications

- Suboptimal examination criteria

vi) Contradictions

vii) Echocardiographic modalities

- Fundamental
- Harmonic
- Color Doppler
- Power Doppler

c. Applications

i) Endocardial border definitions

ii) Myocardial perfusions

iii) Doppler signal enhancement

iv) Definition of cardiac mass

d. Informed consent

e. Patient monitoring

Section XIX: Advanced Techniques/Procedures

1. Describe advanced techniques/procedures that can be used in echocardiography
2. Describe clinical setting and application of advanced techniques/procedures
3. Develop basic understanding of technical application of each advanced technique/procedure

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XIX. Advanced Techniques/Procedures

- A. Intraoperative Echocardiography
  - 1. Approaches
    - a. TEE
    - b. Epicardial
  - 2. Indications
- B. Intravascular Ultrasound (IVUS)
  - 1. Description
  - 2. Transducer types/frequencies
    - a. Mechanical
    - b. Phased array
  - 3. Clinical uses
  - 4. Limitations
- C. Intracardiac Echocardiography (ICE)
- D. Echocardiography-Guided Interventional Procedures
  - 1. Pericardiocentesis
  - 2. Biopsy
- E. Myocardial Strain Imaging
  - 1. Definition
    - a. Positive strain
    - b. Negative strain
    - c. Color strain tracking
    - d. Units
  - 2. Clinical application
  - 3. Technical consideration
- F. Speckle Imaging
  - 1. Definitions
    - a. Tissue tracking
  - 2. Clinical application
  - 3. Technical consideration
- G. Evaluation of Ventricular Assist Device
  - 1. Transthoracic Echocardiographic Assessment of Continuous-Flow Left Ventricular Assist Devices
    - a. LVADs pump blood out of the LV and into the aorta
    - b. Increase cardiac output
    - c. Decrease intra-cardiac pressures

d. Pulsatile-flow
e. Continuous flow
f. Rotating motor
g. Four components
    i) Inflow cannula
    ii) Pump
    iii) Outflow graft
    iv) External controller/battery pack
h. Terms
    i) Bridge to transplant (BTT)
    ii) Bridge to decision/improvement
    iii) Destination therapy (DT)

2. Echo findings
    a. LV volume decreases
    b. MR decreases
    c. Mild systolic and diastolic AR
    d. AV stays closed on most beats
    e. Laminar flows
    f. Cannula velocity
3. Complications
    a. Effusions/hematomas
    b. Infections/endocarditis (ICD leads)
    c. Excessive suction (suck down/septum left)
    d. Insufficient suction (dilated LV/septum right, flow throughout AV, MR, smoke in LV)
    e. RV failure
    f. Aortic regurgitation
    g. Flow obstruction/thrombus

H. Cardiac Resynchronization Therapy (CRT)
1. Definition
    a. Dyssynchrony
        i) Interventricular
        ii) Intraventricular
        iii) Atrioventricular (AV)
    b. Biventricular pacing
    c. Cardiac reverse remodeling
    d. Responders
    e. Nonresponders
2. Patient selection
    a. Inclusion criteria
    b. Dyssynchrony indices

c. Echocardiographic methods/measurements

i) 2-D echocardiography

- LV end-diastolic and end-systolic diameters
- LV end-diastolic and end-systolic volumes (biplane method of discs)
- LV ejection fraction
- M-mode measurement of time delay between two walls: septal to posterior wall mechanical delay (SPWMD)

ii) Pulsed/continuous wave Doppler echocardiography

- RV and LV outflow tract velocities
- dp/dt derived from MR jet acceleration velocity

iii) 3D echocardiography

- LV ejection fraction
- LV volumes
- Segmental wall dyssynchrony

iv) Tissue Doppler imaging (TDI)

- Time of peak LV contraction systolic velocity
- Color TDI
- Tissue synchronization imaging (TSI)

v) Myocardial strain and strain rate

3. AV optimization

a. Technical aspects of adjusting AV delays

b. Hemodynamic changes seen with changes in AV delays

c. Methods

i) Ritter

ii) Iterative

d. Echocardiographic approach

i) Mitral inflow

- E wave
- A wave/truncation
- A wave duration
- EA fusion

ii) Aortic outflow tract

- VTI

4. V to V optimization

a. Technical aspects of adjusting ventricular delays

b. Hemodynamic changes seen with changes in V to V delay

c. Methods

i) Iterative

d. Echocardiographic approach

i) Aortic outflow

- VTI

ii) RV, LV outflow ejection time intervals

Section XX: Systemic Disease

1. List common cardiac abnormalities resulting from the following systemic diseases: amyloidosis, carcinoid, sarcoidosis, hypereosinophilia, hemochromatosis, connective tissue disorders, endocrine diseases and vasculitis
2. Discuss the pathophysiology of the aforementioned systemic diseases
3. Describe typical clinical presentations associated with cardiac involvement in systemic disease
4. Describe key echocardiographic findings associated with cardiac involvement in systemic disease

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XX. Systemic Disease

A. Amyloid
   1. General
      a. Heterogeneous group of diseases
      b. Deposition of extracellular proteins in various organs
      c. Types of amyloid
         i) Primary
         ii) Secondary
         iii) Familial
         iv) "Senile"
      d. Cardiac involvement common
   2. Cardiac manifestations
      a. Systolic or diastolic heart failure
      b. Arrhythmias
      c. Conduction disturbances
      d. Embolic events
      e. Coronary insufficiency (amyloid infiltration of intramyocardial arteries)
   3. Clinical presentation
      a. Symptoms of CHF with cardiac involvement
      b. Protein deposition in tongue, heart, kidney, GI tract, blood vessels, nerves, carpal ligaments
      c. Monoclonal immunoglobulin in urine or serum plus any of the following:
         i) Unexplained nephrotic syndrome
         ii) Hepatomegaly
         iii) Carpal tunnel syndrome
         iv) Macroglossia
         v) Malabsorption/unexplained diarrhea/constipation
         vi) Peripheral neuropathy
         vii) Cardiomyopathy
   4. Echocardiographic features
      a. Increased LV/RV wall thickness
      b. Myocardium may appear granular
      c. Normal to small LV size with gradual decline in LV function

d. Thickened cardiac valves with regurgitation (usually mild)
e. Atrial enlargement
f. Thickened inter-atrial septum
g. Thickened papillary muscles
h. Pericardial/pleural effusion
i. Spectrum of diastolic dysfunction
j. Progression of above corresponds to increasing LV wall thickness, systolic dysfunction and more advanced disease state

5. Related testing
   a. ECG
      i) Atrial fibrillation
      ii) Conduction disturbances
      iii) Low voltage
      iv) Pseudoinfarction pattern
   b. Bone marrow biopsy
6. Common treatment
   a. Medical
      i) Supportive treatment of cardiac failure
      ii) Chemotherapy
   b. Surgical
      i) Stem cell transplant
      ii) Cardiac transplantation

B. Carcinoid
1. General
   a. Malignant, metastatic endocrine tumor
   b. Primary tumors: gastrointestinal (GI) tract, bronchus, ovaries, pancreas, biliary tract
   c. Carcinoid syndrome
   d. Prognosis worse in patients with cardiac involvement
   e. Severity of cardiac involvement proportional to
      i) Serotonin
      ii) Kinins
      iii) Urinary 5-HIAA
2. Cardiac manifestations
   a. Hepatic metastases release tumor substances into the right heart
   b. Fibrous white plaque deposited that leads to fibrosis and valvular dysfunction
   c. Predominant involvement on right side (TV, PV)
   d. Tumor substances inactivated by the lungs
   e. Left-sided involvement seen with
      i) Intracardiac shunt (right-to-left)
      ii) Bronchial tumor or lung metastases
   f. May be mimicked by drug-induced valvulopathy

3. Clinical presentation
    a. Carcinoid syndrome: flushing, hypotension, diarrhea, wheezing, bronchospasm, endocardial plaque formation
    b. Heart failure (late)
4. Echocardiographic features
    a. Tricuspid valve leaflets thickened, retracted, fixed in semi-open position
        i) Usually severe TR with mild TS
    b. Pulmonary valve leaflets thickened, retracted and rigid
        i) Varying degrees of PR and PS (usually more PS than PR)
    c. Similar mitral and aortic valve characteristics if left heart is affected (may occur with PFO or bronchial tumor)
    d. Evidence of right heart volume overload
    e. Liver metastasis
    f. Myocardial metastases
    g. Pericardial effusion
5. Related testing
    a. Serotonin levels
    b. Urinary 5-HIAA
    c. Tissue biopsy
6. Common treatment
    a. Medical
        i) Counter-regulatory hormones
        ii) Chemotherapy (high-grade malignancies)
    b. Surgical
        i) Valve replacement/removal
        ii) Hepatic artery embolization

C. Hypereosinophilic Syndrome
1. General
    a. Always associated with hypereosinophilia – an abnormal formation and accumulation of extremely large number of eosinophils in blood without identifiable etiology
    b. Organ infiltration
    c. Multisystem disease: cardiac, skin, neurologic, pulmonary, GI, hepatic, renal
    d. Cardiac involvement most common cause of morbidity and mortality
2. Cardiac manifestations
    a. Endomyocardial fibrosis
    b. Loffler's cardiomyopathy
3. Clinical presentation
    a. Pulmonary and/or systemic embolization
    b. CHF
    c. Sudden death
    d. Variable symptoms: dyspnea most common

4. Echocardiographic features
    a. Normal LV and RV size and systolic function
    b. Atrial enlargement
    c. Left and/or right ventricular apical thrombus with cavity obliteration
    d. Posterior AV valves and subvalvular apparatus (most often PLMV) may become thickened and tethered with associated MR and/or TR
    e. Thick inferobasal LV wall
    f. Restrictive physiology due to decreased LV compliance
5. Related testing
    a. Labs: Persistent (at least six months) increase in eosinophil count > 1500 cells/mm3
6. Common treatment
        i) Medical
        ii) Treat CHF
        iii) Anticoagulants
        iv) Treat underlying hypereosinophilia (steroids)
    b. Surgical
        i) Thrombectomy
        ii) Mitral and/or tricuspid valve replacement
        iii) Endocardial stripping

D. Sarcoidosis
1. General
    a. Multisystem granulomatous disease of unknown cause
    b. Lungs, skin, heart commonly involved
2. Cardiac manifestations
    a. Ranges from a few asymptomatic granulomatous lesions to widespread infiltration of the myocardium
    b. Sudden death
    c. Arrhythmias
    d. Conduction abnormalities
    e. LV dysfunction and CHF
    f. Cor pulmonale with pulmonary sarcoid
3. Clinical presentation
    a. Typically present before age 40
4. Echocardiographic features
    a. Enlarged LV with decreased systolic function
    b. RWMA
    c. Posterolateral wall thinning with or without aneurysm
    d. Diastolic dysfunction (precedes systolic)
    e. Valvular dysfunction
    f. Pericardial effusion
    g. RV and RA enlargement and pulmonary hypertension (cor pulmonale)

5. Related testing
    a. ECG: conduction abnormalities, arrhythmias
    b. Biopsy
6. Common treatment
    a. Medical
        i) Pharmacologic therapy to control inflammation
    b. Surgical
        i) Pacemaker/AICD
        ii) Ventricular aneurysm resection
        iii) Transplant

E. Hemochromatosis
1. General
    a. Excess iron storage disease
    b. Iron is deposited within cells in organ tissue, interfering with organ function
    c. May be primary (hereditary): inappropriate increased absorption from GI tract or secondary (acquired): transfusion, iron therapy, etc.
    d. Peaks in fifth decade; rare before age 20
2. Cardiac manifestations
    a. Dilated cardiomyopathy
    b. Cardiac arrhythmias
    c. Degree of cardiac dysfunction reflects degree of iron overload
3. Clinical presentation
    a. Typically male > 40 years old with
        i) Diabetes mellitus
        ii) Skin disease (hyperpigmentation)
        iii) Liver disease/failure
        iv) Heart failure
4. Echocardiographic features
    a. Dilated cardiac chambers
    b. Global LV systolic dysfunction
    c. Normal wall thickness
    d. Mild valvular regurgitation
    e. Progressive diastolic dysfunction
5. Related testing
    a. Endomyocardial biopsy
    b. Serum iron levels
6. Common treatment
    a. Medical
        i) Phlebotomy
        ii) Manage organ dysfunction

F. HIV Disease/Acquired Immunodeficiency Syndrome (AIDS)

1. General
    a. Cellular immune dysfunction
2. Cardiac manifestations
    a. Dilated cardiomyopathy/CHF
    b. Pericarditis/pericardial effusion
    c. RV dysfunction and pulmonary hypertension
    d. Endocarditis
    e. CAD (may be due to antiretroviral therapy)
    f. Cardiac malignancy
3. Clinical presentation
    a. Cardiac involvement relatively common but usually clinically silent
    b. Significant cardiac abnormalities most common in terminal phase
4. Echocardiographic features
    a. Dilated LV and/or RV with systolic dysfunction
    b. Varying degrees of diastolic dysfunction
    c. Mitral regurgitation
    d. Endocarditis (bacterial and nonbacterial thrombotic)
    e. Pericardial effusion with or without tamponade
    f. Pleural effusion
    g. Constrictive pericarditis
    h. Cardiac neoplasm
    i. Pulmonary hypertension
5. Related testing
6. Common treatment
    a. Medical
        i) Treat underlying disease
        ii) Treat cardiac disease and heart failure

G. Connective Tissue Diseases
1. Rheumatoid arthritis (RA)
    a. General
        i) Chronic inflammatory disease (primarily involves joints)
        ii) Age-related
        iii) More common in women
        iv) Systemic symptoms: fatigue, myalgias, weight loss, fever, pain malaise, anorexia
        v) May be insidious or sudden onset
        vi) Limited motion and loss of function
    b. Cardiac manifestations
        i) Symptomatic cardiac disease not common
        ii) Pericarditis
        iii) Myocarditis

iv) Endocardial/valvular
v) Conduction system
vi) Aortic and pulmonary

c. Echocardiographic features
i) Pericardial
- Effusion
- Constriction

ii) Myocardial
- Global LV dysfunction
- Regional wall motion abnormalities (rare)
- Diastolic dysfunction
- Nodules in myocardium

iii) Valvular
- Valve thickening and regurgitation
- Valvular lesions similar to rheumatoid nodules

iv) Secondary pulmonary hypertension (rare)

2. Systemic lupus erythematosus (SLE)
a. General
i) Common autoimmune disease characterized by production of autoantibodies
ii) More prevalent and severe in women
iii) Cardiac involvement is common

b. Cardiac manifestations
i) Valvular (common)
ii) Pericardial (common)
iii) Myocardial
iv) Pulmonary hypertension with pulmonary involvement
v) Conduction abnormalities
vi) Vascular thrombosis

c. Echocardiographic features
i) Valvular
- Libman-Sacks endocarditis (non-bacterial thrombotic)
- Valvulitis
- Diffuse leaflet thickening
- Regurgitation
- Valve stenosis (rare)

ii) Pericardial
- Effusion
- Constriction (rare)

iii) Myocardial
- Global LV dysfunction

- Regional wall motion abnormalities

iv) Pulmonary hypertension with pulmonary involvement

3. Antiphospholipid syndrome
   a. General
      i) Hypercoagulability syndrome
      ii) High titer of antiphospholipid antibodies
      iii) Clinical features
         - Recurrent vascular thrombosis
         - Pregnancy loss
         - Thrombocytopenia
         - Positive anticardiolipin test
   b. Cardiac manifestations
      i) Cardiac dysfunction with coronary thrombosis
      ii) Pulmonary hypertension with pulmonary thrombosis
      iii) Valvular (most common)
         - Nonbacterial thrombotic endocarditis
         - Diffuse thickening
   c. Echocardiographic features
      i) Intracardiac, aortic thrombi
      ii) LV systolic dysfunction
      iii) Valvular regurgitation
      iv) Pulmonary hypertension
4. Scleroderma (systemic sclerosis)
   a. General
      i) Excess connective tissue accumulates in blood vessels, skin, joints, skeletal muscle, heart
      ii) More prevalent in women
      iii) Clinical variants
         - Diffuse
         - Limited cutaneous
   b. Cardiac manifestations
      i) Clinical evidence uncommon
      ii) Conduction abnormalities
      iii) Myocarditis
      iv) Pericarditis
      v) Valvular
      vi) Pulmonary hypertension with pulmonary involvement
   c. Echocardiographic features
      i) Myocardial
         - LVH with systolic hypertension

- LV dysfunction
- Cardiomyopathy

ii) Pericardial

- Effusion, tamponade
- Constriction

iii) Pulmonary hypertension

H. Ankylosing Spondylitis

1. General
   a. Autoimmune disease
   b. Chronic systemic inflammatory disorder
   c. Primarily affects the axial skeleton (hips and shoulders)
   d. Genetic factors
   e. Men affected more than women
2. Cardiac manifestations
   a. Aortic disease common
   b. AV and MV disease
   c. Conduction abnormalities
   d. Cardiomyopathy
   e. Pericarditis
3. Echocardiographic features
   a. Dilatation of aortic annulus and sinus of Valsalva
   b. AV thickening and regurgitation
   c. MVP
   d. LV systolic dysfunction
   e. Pericardial effusion (rare)

I. Marfan Syndrome

1. General
   a. Hereditary connective tissue disorder (autosomal dominant)
   b. Defect in fibrillin
   c. Primarily affects ocular, skeletal, and cardiovascular systems
   d. Most common cause of death in adults is aortic dissection
2. Cardiac manifestations
   a. MV disease
   b. Dilated ascending aorta
   c. Aortic regurgitation
   d. Aortic dissection
3. Echocardiographic features
   a. Aortic root dilatation
   b. Dilated ascending aorta
   c. Annuloaortic ectasia

d. AR
e. Aortic dissection
f. Myxomatous mitral valve with prolapse
g. MR

J. Giant Cell Arteritis
1. General
a. Vasculitis involving large and medium-sized arteries
2. Cardiac manifestations
a. Myocardial inflammation
b. Pericardial inflammation
c. Aortitis
3. Echocardiographic features
a. Aortic aneurysm and dissection
b. Thickened AV
c. LV systolic dysfunction (myocarditis)
d. Pericardial effusion (pericarditis)

K. Takayasu Arteritis
1. General
a. Granulomatous panarteritis of large vessels
b. Unknown cause
2. Cardiac manifestations
a. Aortitis
3. Echocardiographic features
a. Aortic dilatation
b. AR
c. Stenosis and occlusion of large vessels

L. Kawasaki Disease
1. General
a. Acute systemic vasculitis of unknown origin
b. Mucocutaneous, lymph node syndrome
c. Usually seen < 5 years of age
2. Cardiac manifestations
a. Vasculitis of coronary vasa vasorum
b. Leads to coronary artery aneurysms
i) Thrombosis
ii) Stenosis
iii) Myocardial ischemia, MI
c. Conduction abnormalities
d. Myocarditis
3. Echocardiographic features

a. Coronary artery aneurysms
b. Pericardial effusion (pericarditis)
c. LV dysfunction (myocarditis)
d. MR

M. Churg-Strauss Syndrome
1. General
a. Systemic vasculitis
b. Characterized by asthma and/or allergic rhinitis, peripheral and tissue eosinophilia, extravascular granuloma formation, and vasculitis of multiple organ systems
2. Cardiac/echo findings
a. Pericardial effusion (pericarditis)
b. DCM (myocarditis)
c. Endomyocardial fibrosis

N. Wegener's Granulomatosis
1. General
a. Vasculitis of unknown origin
b. Pathology defined by the triad of small vessel vasculitis, granulomatous inflammation, and necrosis
c. Multisystem involvement
2. Cardiac manifestations
a. Pericarditis
b. Myocarditis
c. Valvulitis
d. Arteritis
e. Mass lesions (granulomas)
f. Arrhythmias
3. Echocardiographic features
a. LV RWMA
b. Global LV hypokinesis
c. Pericardial effusion
d. Valvular regurgitation

O. Endocrine Diseases
1. Hyperthyroidism
a. Sustained overproduction of thyroid hormone, usually due to enlarged thyroid gland (hypermetabolism)
b. Increased SV, CO, and LV mass
c. DCM (tachycardia-induced)
d. Diastolic dysfunction
e. Atrial fibrillation
f. Pulmonary hypertension (rare)
2. Hypothyroidism

a. Thyroid hormone deficiency
b. Decreased HR and CO
c. Diastolic dysfunction
d. DCM
e. Pericardial effusion
f. Valvular thickening
g. Accelerated atherosclerosis

3. Pheochromocytoma
   a. Rare adrenal tumor or tumor along the sympathetic chain (usually intra- abdominal) that produces excessive catecholamines
   b. Increased HR and contractility
   c. LV systolic dysfunction with catecholamine crisis
   d. LV hypertrophy (occasional)
   e. HCM with or without dynamic LVOT obstruction
4. Acromegaly
   a. Secretion of excessive growth hormone, nearly always caused by a pituitary adenoma
   b. Cardiac disease occurs in one-third of patients
   c. CHF
   d. Concentric LVH
   e. Diastolic dysfunction
   f. Global systolic dysfunction

P. Systemic Infection/Sepsis
1. General
   a. Systemic inflammatory response
   b. May have infectious or non-infectious etiology
2. Echo features
   a. Reversible dilated LV with systolic dysfunction
   b. Diastolic dysfunction
   c. Endocarditis
   d. Pericardial and pleural effusions

Q. Hereditary Hemorrhagic Telangectasia (Osler-Weber-Rendu)
1. General
   a. Autosomal dominant disorder of development of the vasculature
   b. Triad: mucocutaneous and visceral telangiectasias, recurrent epistaxis, and familial history
   c. Autosomal dominant
2. Cardiac manifestations
   a. High cardiac output
   b. Pulmonary arteriovenous malformations (AVM)
   c. Coronary arteriovenous malformations (AVM)
3. Echocardiographic features

a. Presence of pulmonary AVM indicated by late appearance of bubbles in the left atrium after agitated saline injection

Section XXI: Cardiac Transplantation

1. List primary indications for cardiac transplantation
2. Discuss surgical techniques used in cardiac transplantation
3. Describe complications of cardiac transplantation
4. Describe key echocardiographic findings associated with cardiac transplantation

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XXI. Cardiac Transplantation

- A. Indications
  - 1. DCM
  - 2. Cardiac amyloid
  - 3. Congenital heart disease
- B. Cardiac Transplantation Surgical Techniques
  - 1. Orthotopic
  - 2. Heterotopic
  - 3. Xenotransplantation
  - 4. Artificial heart
- C. Role of Echo Assessment
  - 1. LV size and function
    - a. LV systolic dysfunction may occur early
    - b. Dysfunction may be secondary to peri-operative donor heart ischemia
  - 2. RV size and function
    - a. RV systolic function may be reduced secondary to elevated PASP
    - b. Serial assessment of RVSP
  - 3. Noninvasive clues for rejection
  - 4. Post-transplant CAD
  - 5. Exclusion of pericardial effusion
- D. Complications of Cardiac Transplantation
  - 1. Side effects of immunosuppression therapy
  - 2. Coronary artery disease (graft atherosclerosis)
    - a. Accelerated coronary atherosclerosis
    - b. Full length of vessel affected (uniform, diffuse involvement)
    - c. Silent myocardial infarction (no angina due to denervation)
  - 3. Infection
  - 4. Rejection
    - a. Response of the recipient's immune system to foreign antigens
    - b. Acute rejection (mononuclear cells infiltrate myocardium and attack myocardial cells)
    - c. Confirmed by RV biopsy
  - 5. Post-transplant malignancies
  - 6. Denervation of the heart
    - a. Interruption of nerve impulse route due to excision of heart

b. Noncardiac mediators augment heart rate & contractility
c. Delayed response to exercise
d. Chest pain receptors are cut, resulting in inability to feel angina
e. Altered conduction pathways

E. Echocardiographic Features
1. 2-D
a. Normal LV size and function
b. Biatrial enlargement
c. Atrial suture lines visualized
d. Pericardial effusion
e. Paradoxical septal motion
f. Increased left ventricular wall thickness
g. Increased right ventricular dimension
h. Tricuspid regurgitation
2. Doppler findings in transplantation
a. Diastolic function assessment reflects the donor heart
b. Serial assessment of RVSP
c. Assess for increasing TR volume secondary to complications of biopsy

F. Echo Features of Rejection
1. Rejection associated with non-compliance of left ventricle due to extracellular infiltration and edema
2. Increase in LV wall thickness, LV mass
3. Decrease in LV and/or RV systolic function
4. Increase in RVSP
5. New pericardial effusion
6. Decrease in Global Longitudinal Strain
7. Restrictive filling
a. Decreased deceleration time from immediate post-op echo
b. Decreased annular TDI velocity
c. Evidence of elevated filling pressures

Section XXII: Miscellaneous Topics

1. List common cardiac abnormalities resulting from cardiac trauma
2. Describe common echocardiographic findings with cardiac trauma
3. Discuss the physiology of athlete's heart
4. Describe common echocardiographic findings of athlete's heart

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XXII. Miscellaneous Topics

- A. Cardiac Trauma
  - 1. Use of echocardiography
    - a. Early diagnosis
    - b. Diagnosis may be difficult due to injuries to other organs
    - c. TEE useful
  - 2. Blunt chest trauma – cardiac injury evaluation
    - a. May be obscured by injury to other organs
    - b. Right to left cardiac shunt
      - i) VSD
      - ii) Aneurysm of RVOT
      - iii) Pseudoaneurysm
      - iv) Ruptured septal coronary
      - v) Disruption of papillary muscle
    - c. Valve rupture
    - d. Coronary artery rupture
    - e. Chamber rupture
  - 3. Blunt chest trauma – great vessel injury evaluation
    - a. Transection of great vessels
    - b. Aortic rupture
    - c. Dissection
    - d. Evaluate
      - i) Ascending aorta
      - ii) Aortic isthmus
      - iii) Aortic hiatus
      - iv) Aortic arch and branch vessels
        - Innominate
        - Right common carotid
        - Left common carotid
        - Right subclavian
        - Left subclavian
  - 4. Penetrating trauma
    - a. Foreign objects
      - i) Bullet

ii) Nail

b. Stab wounds

i) Perforation of annulus

ii) Perforation of IVS

iii) Prolapse of aortic cusps

5. General echo features

a. Cardiac tamponade

b. Pneumothorax

B. Athlete's Heart

1. General

a. History of athletic training and performance

b. Enhanced exercise ability

c. Resting bradycardia

d. Dynamic versus static exercise

2. Echocardiographic features

a. Increased left ventricular mass

b. Increased LV end-diastolic dimension

c. Increase in LV wall thickness

d. Increased RV cavity dimensions

e. Increased LA size

Section XXIII: Radiation Safety

1. Describe the basic principles of radiation
2. Describe the basic principle of radiation safety and radiation safety techniques
3. Describe regulatory issues related to radiation safety

XXIII. Radiation Safety

A. Basic Radiation Principles

B. Basic Radiation safety principles

C. Personal Protection Techniques

D. Regulatory Issues

Section XXIV: Structural Heart Interventions

1. Describe structural heart abnormalities that could be repaired with transcatheter techniques
2. Describe the role of echocardiography in assessing transcatheter based interventions.
3. Describe transcatheter based interventions

XXIV. Structural Heart Interventions

- A. Structural Heart Abnormalities
  - 1. Native and prosthetic valvular disorders
    - a. Valvular stenosis
    - b. Valvular regurgitation
    - c. Perivalvular leaks
  - 2. Shunts
    - a. Atrial level shunts
    - b. Ventricular level shunts
  - 3. Thrombo-embolic events
    - a. Left atrial appendage thrombus
  - 4. Mitral regurgitation
- B. Role of Echo in the Assessment of Transcatheter-Based Interventions
  - 1. Pre-procedure diagnosis
  - 2. Intra-procedure guidance
  - 3. Immediate post-procedure assessment
  - 4. Long-term follow up
- C. Transcatheter-Based Interventions
  - 1. Transcatheter aortic valve replacement/intervention (TAVR/TAVI)
  - 2. Valve in valve procedures
  - 3. Mitral valve repair (E-Clip)
  - 4. Perivalvular leak closure
  - 5. Transcatheter based shunt closure
    - a. ASD
    - b. VSD
  - 6. Left atrial appendage closure

Abbreviations

A

a’	Late Diastolic Annular Velocity
AI	Aortic Insufficiency
AMVL	Anterior Mitral Valve Leaflet
Ao	Aorta
APVR	Anomalous Pulmonary Venous Return
APVS	Absent Pulmonary Valve Syndrome
A_R	Atrial Reversal Wave
AR	Aortic Regurgitation
AS	Aortic Stenosis
ASD	Atrial Septal Defect
ASH	Asymmetric Septal Hypertrophy
ASO	Arterial Switch Operation
AV	Aortic Valve
AVA	Aortic Valve Area
AVV	Atrioventricular Valve

B

BTS	Blalock-Taussig Shunt
BAV	Bicuspid Aortic Valve

C

CAD	Coronary Artery Disease
CAVC	Complete Atrioventricular Canal Defect
CCA	Common Carotid Artery
CCTGA	Congenitally Corrected Transposition of the Great Arteries
CHD	Congenital Heart Disease
CHF	Congestive Heart Failure
CO	Cardiac Output
COA	Coarctation of the Aorta
CRT	Cardiac Resynchronization Therapy
CS	Coronary Sinus
CW	Continuous Wave

D

DCM	Dilated Cardiomyopathy
D-loop	Dextrolooping
DOMV	Double Orifice Mitral Valve
DORV	Double Outlet Right Ventricle
DT	Deceleration Time
DTGA	Dextro-Transposition of the Great Arteries

E

e’	Early Diastolic Annular Velocity
ECA	External Carotid Artery
ECG	Electrocardiogram
ED	End-Diastole
EDD	End-Diastolic Dimension
EDV	End-Diastolic Volume
EF	Ejection Fraction
EPSS	E-Point Septal Separation
ESV	End-Systolic Volume

F

FS	Fractional Shortening

H

HCM	Hypertrophic Cardiomyopathy
HLHS	Hypoplastic Left Heart Syndrome
HOCM	Hypertrophic Obstructive Cardiomyopathy

I

IAA	Interrupted Aortic Arch
IAS	Interatrial Septum
ICE	Intracardiac Echocardiography
IHSS	Idiopathic Hypertrophic Subaortic Stenosis
ICA	Internal Carotid Artery
lVC	Inferior Vena Cava
IVRT	Isovolumic Relaxation Time
IVS	Interventricular Septum
IVUS	Intravascular Ultrasound

J

JVP	Jugular Venous Pressure

L

LA	Left Atrium
LAA	Left Atrial Appendage
LBBB	Left Bundle Branch Block
LCA	Left Coronary Artery
LCC	Left Coronary Cusp
L-loop	Levolooping
LPA	Left Pulmonary Artery
LSVC	Left Superior Vena Cava
LV	Left Ventricle
LVEDD	Left Ventricular End-Diastolic Diameter
LVEDP	Left Ventricular End-Diastolic Pressure
LVEDV	Left Ventricular End-Diastolic Volume
LVH	Left Ventricular Hypertrophy
LVIT	Left Ventricular Inflow Tract
LVOT	Left Ventricular Outflow Tract
LVPW	Left Ventricular Posterior Wall

M

MAC	Mitral Annular Calcification
MAPCA(s)	Major Aorto-pulmonary Collateral Artery(ies)
MI	Myocardial Infarction
MPA	Main Pulmonary Artery
MR	Mitral Regurgitation
MS	Mitral Stenosis
MV	Mitral Valve
MVA	Mitral Valve Area
MVP	Mitral Valve Prolapse
MVR	Mitral Valve Replacement

N

NCC	Non-Coronary Cusp

P

PAEDP	Pulmonary Artery End-Diastolic Pressure
PA(s)	Pulmonary Artery(ies)
PAPVR	Partial Anomalous Pulmonary Venous Return
PASP	Pulmonary Artery Systolic Pressure
PAVC	Partial Atrioventricular Canal Defect

PDA	Patent Ductus Arteriosis
PFO	Patent Foramen Ovale
PHTN	Pulmonary Hypertension
PI	Pulmonic Insufficiency
PISA	Proximal Isovelocity Surface Area
PLAX	Parasternal Long Axis
PMV	Parachute Mitral Valve
PMVL	Posterior Mitral Valve Leaflet
PS	Pulmonic Stenosis
PSAX	Parasternal Short Axis
PV	Pulmonary Valve
PW	Pulsed Wave

R

RA	Right Atrium
RAA	Right Atrial Appendage
RAP	Right Arterial Pressure
RBBB	Right Bundle Branch Block
RCC	Right Coronary Cusp
RCM	Restrictive Cardiomyopathy
RIMP	Right Ventricular Index of Myocardial Performance
RPA	Right Pulmonary Artery
RV	Right Ventricle
RVEDP	Right Ventricular End-Diastolic Pressure
RVH	Right Ventricular Hypertrophy
RVIT	Right Ventricular Inflow Tract
RVOT	Right Ventricular Outflow Tract
RVP	Right Ventricular Pressure
RVVO	Right Ventricular Volume Overload

S

s’	Systolic Annular Velocity
SAM	Systolic Anterior Motion
SPWMD	Septal to Posterior Wall Mechanical Delay
SSN	Suprasternal Notch
SV	Stroke Volume
SVC	Superior Vena Cava

T

TAPSE	Tricupsid Annular Plane Systolic Excursion
TAPVR	Total Anomalous Pulmonary Venous Return
TAVI	Transcatheter Aortic Valve Intervention
TAVR	Transcatheter Aortic valve Replacement
TDI	Tissue Doppler Imaging
TEE	Transesophageal Echocardiography
TGA	Transposition of the Great Arteries
TOF	Tetralogy of Fallot
TR	Tricuspid Regurgitation
TS	Tricuspid Stenosis
TTE	Transthoracic Echocardiography
TV	Tricuspid Valve
TVA	Tricuspid Valve Area
TVP	Tricuspid Valve Prolapse

V

Vp	Velocity Propagation
VSD	Ventricular Septal Defect
VTI	Velocity Time Integral

W

WMA	Wall Motion Abnormality
WPW	Wolff-Parkinson White

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